Clinical and Surgical Management of Pediatric Diseases of the Oral Cavity, Maxilla, and Mandible

12

Raanan Cohen-Kerem

Introduction

The oral cavity, including the jaws, is a site for numerous pathologies in the pediatric age group. Appearance of such a lesion in a child is alarming to the parents who would seek for prompt medical attention. Non-treated pathology in the oral cavity may cause functional disruption and lead to airway compromise, reduced nourishment, and speech disturbance. Moreover, neglected conditions can lead to facial disfigurement followed by psychological consequences and body dysmorphic disorder.

A broad differential diagnosis is applicable to most of the pathologies, congenital, degenerative, inflammatory, infectious, and neoplastic. Several are rare and some are esoteric that one would encounter once in a lifetime career, therefore, it is important that the pediatric otolaryngologist is acquainted with those pathologies, the workup needed, and the appropriate management that would facilitate a successful result with minimal morbidity. A successful outcome may require the cooperation of a multidisciplinary team including pediatric otolaryngologist, maxillofacial surgeon, plastic surgeon, neurosurgeon, pediatrician, intensive care physician, dentist, orthodontist, pediatric oncologist, nurses, and a child psychologist.

In the following chapter, we tried to present a variety of medical conditions in the oral cavity and the jaws accompanied by clinical examples in ordrer to assist the clinician in decision-making.

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Soft Tissue (Tongue, Mucosa, Gingivae)

Epulis of Gingiva

Definition

The Greek word "epulis" means "swelling of the gingiva," it is a rare congenital benign tumor that occurs in the anterior alveolar ridge of the jaws [1]. The tumor is usually pedunculated, variable in size with firm consistency and smooth or lobulated surface. It is also known as a congenital gingival granular cell tumor [2].

The tumor is seen solely in newborns [3] and its most common site is the anterior maxillary alveolar ridge followed by the mandible and rarely at the anterior tongue. Multiple tumors may occur and girls are affected 8–10 times more often than boys [3]. The tumor does not tend to grow further after birth [2].

Clinical Presentation

The tumor is apparent promptly after birth protruding from the newborn's mouth or with the baby first cry in case of a small size epulis. It may interfere with feeding or breathing depends on the tumor size. Thorough examination of the oral cavity is warranted to exclude multiple lesions (up to 10%) [1].

Differential Diagnosis

- Granular cell tumor
- Granular cell myoblastoma
- Congenital oral teratoma
- Hemangioma
- Fibroma
- Rhabdomyoma
- Enteric duplication cyst

Radiological Features

Prenatal diagnosis by ultrasound and MRI is possible, however, it is rarely diagnosed since its slow development is in the third trimester. Postnatal imaging is not necessary. It is hypoechoic on ultrasound with crowded branching pattern of

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vessels in the mass. On postnatal MRI the tumor is isointense to the muscle on T1-weighted imaging, T2-weighted images show heterogeneous signal intensity. Gadolinium-enhanced T1-weighted images show mild contrast enhancement of the major portion of the mass with peripheral rim strong contrast enhancement [4].

Management

Though some reports suggested a spontaneous regression, the accepted management strategy is surgical excision while various tools could be utilized. Radical excision must be avoided in order to prevent future alveolar damage. The outcome of incomplete excision is good with no recurrence or malignancy reported [2].

Clinical Examples

Case Presentation 1

A term baby girl was born with a solid mass originating on the mandibular alveolar process. The mass was resected in the NICU under sedation and infiltration of the mass base with 1% lidocaine (Fig. 12.1). Recovery was uneventful with no feeding limitation.

Case Presentation 2

A smaller epulis was seen on the maxillary alveolus of a 3-month-old baby. It was interfering with breastfeeding. Having a broader base, it was excised using bipolar cautery under general anesthetic. The pathology indicated a granular cell tumor (epulis) (Fig. 12.2).

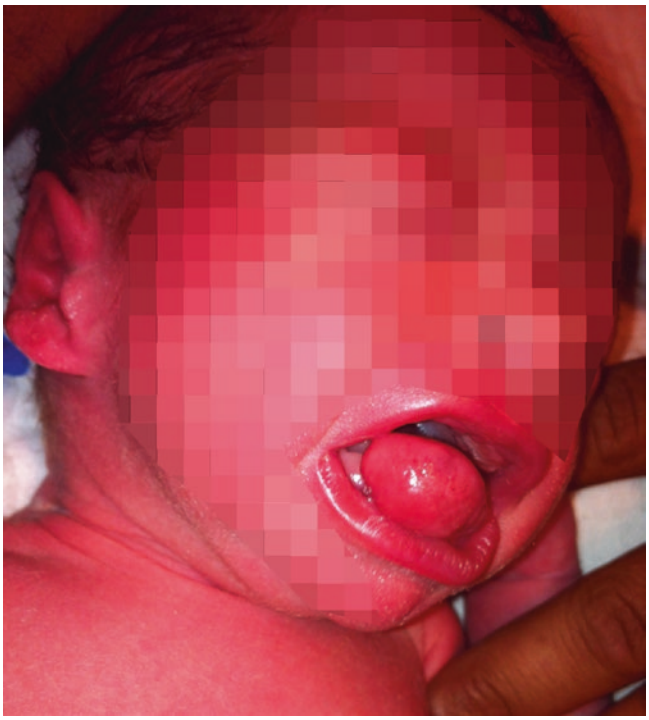

Fig. 12.1 Large epulis on lower gingiva of a newborn

Pemphigus

Definition

A group of rare autoimmune diseases characterized with blistering of the skin and mucous membranes. Pemphigus vulgaris is the commonest type and it may occur at any age, the occurrence in the pediatric age group is very rare with disease onset of 13 to18 years [5, 6]. Oral mucosa is the most commonly affected site but any area of stratified squamous epithelium may get involved [7]. Gender distribution is equal. Ethnic groups such as Ashkenazi Jews and people of Mediterranean and Indian origin are particularly susceptible and there is an association to HLA class II alleles [8].

Clinical Presentation

Erythematous patches, blisters, and erosions are visible in the oral mucosa, skin, and genital mucosa. Oral lesions precede the onset of skin lesion by days to years.

Differential Diagnosis

- Bullous pemphigoid
- Dermatitis herpetiformis
- Drug-induced pemphigus
- Erythema multiforme
- Paraneoplastic pemphigus
- IgA pemphigus [9]

Radiological Features

No radiological study is indicated.

Management

Based mainly on corticosteroids and other immunosuppressants, intravenous immunoglobulin (IVIG) is utilized as well. Earlier diagnosis is likely to result in rapid recovery [5].

Clinical Example

A 13-year-old male presented with a 3-month history of oral blisters. Lesions were identified on the buccal, gingival, and lingual mucosa. He was treated with limited success with antiviral medication and topical steroids. A biopsy of the lesion was performed (Fig. 12.3).

Cysts

Floor of Mouth Developmental Cyst (Dermoid)

Definition

A slow-growing, whitish, cystic mass composed of tissue that is not native to its location and is divided to either dermoid, epidermoid, or teratoma but is usually referred to as dermoid. The floor of the mouth location is usually at the midline anterior part but can present laterally as well [10]. The cyst is lined with keratinized squamous

epithelium and it is thought to originate from a stem cell that undergoes ectodermal differentiation. The floor of the mouth dermoids comprises about 19% of all head and neck dermoid cysts [11].

In addition to dermoids, choristoma, foregut or enteric duplication cysts and potentially bronchogenic cysts can present in the floor of the mouth. They are differentiated based on their contents as outlined in this table:

	Mature cystic teratoma	Foregut duplication cyst	Bronchogenic cyst	Choristoma
Different types of tissue	Yes Ectodermal endodermal mesodermal	No Respiratory/intestinal epithelium	No Respiratory/squamous metaplastic epithelial lining	No Respiratory/intestinal/squamous epithelium
Enteric epithelium	Yes	Yes	No	Yes
Respiratory epithelium	Yes	Yes	Yes	Yes
Cartilage within the wall	Yes	No	No	No
Distinct muscle layers	No	Yes	No	No

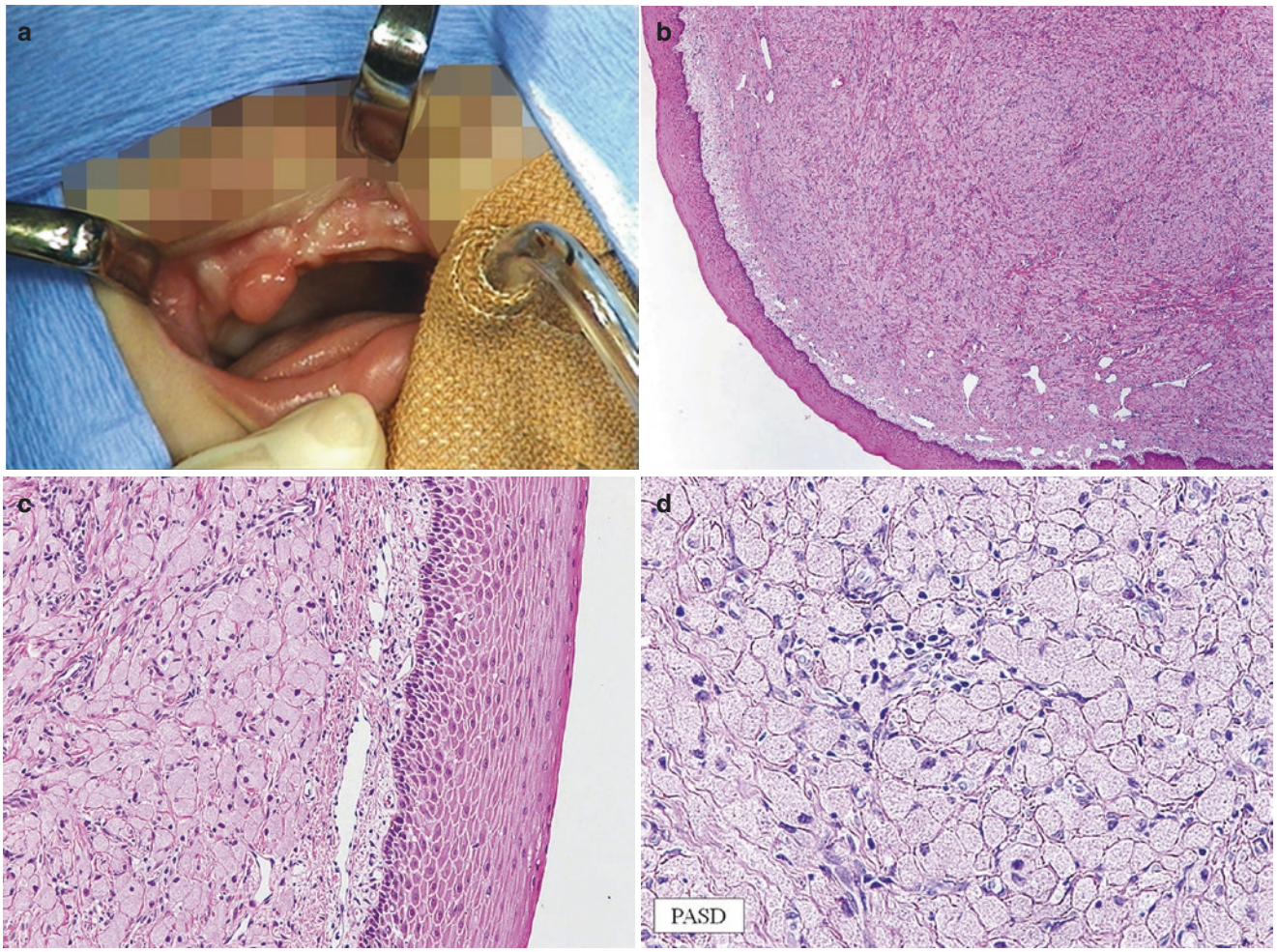

Fig. 12.2 (**a**) Epulis on upper alveolus. (**b**) H&E. Polypoid tumor mass pushing against an intact squamous epithelium of the oral mucosa. (**c**) H&E. Tumor cells were polygonal with large amounts of cytoplasm with ground glass like granular material. The nuclei are small and contain condensed chromatin. (**d**) PASD stain for glycoprotein showing weak staining of the granular cytoplasmic inclusions. (**e**) Positive immunostain for vimentin. (**f**) Negative immunostain for S100. (**g**) Negative immunostain for macrophage marker CD68. Only the stromal histiocytes around some tumor cells are stained

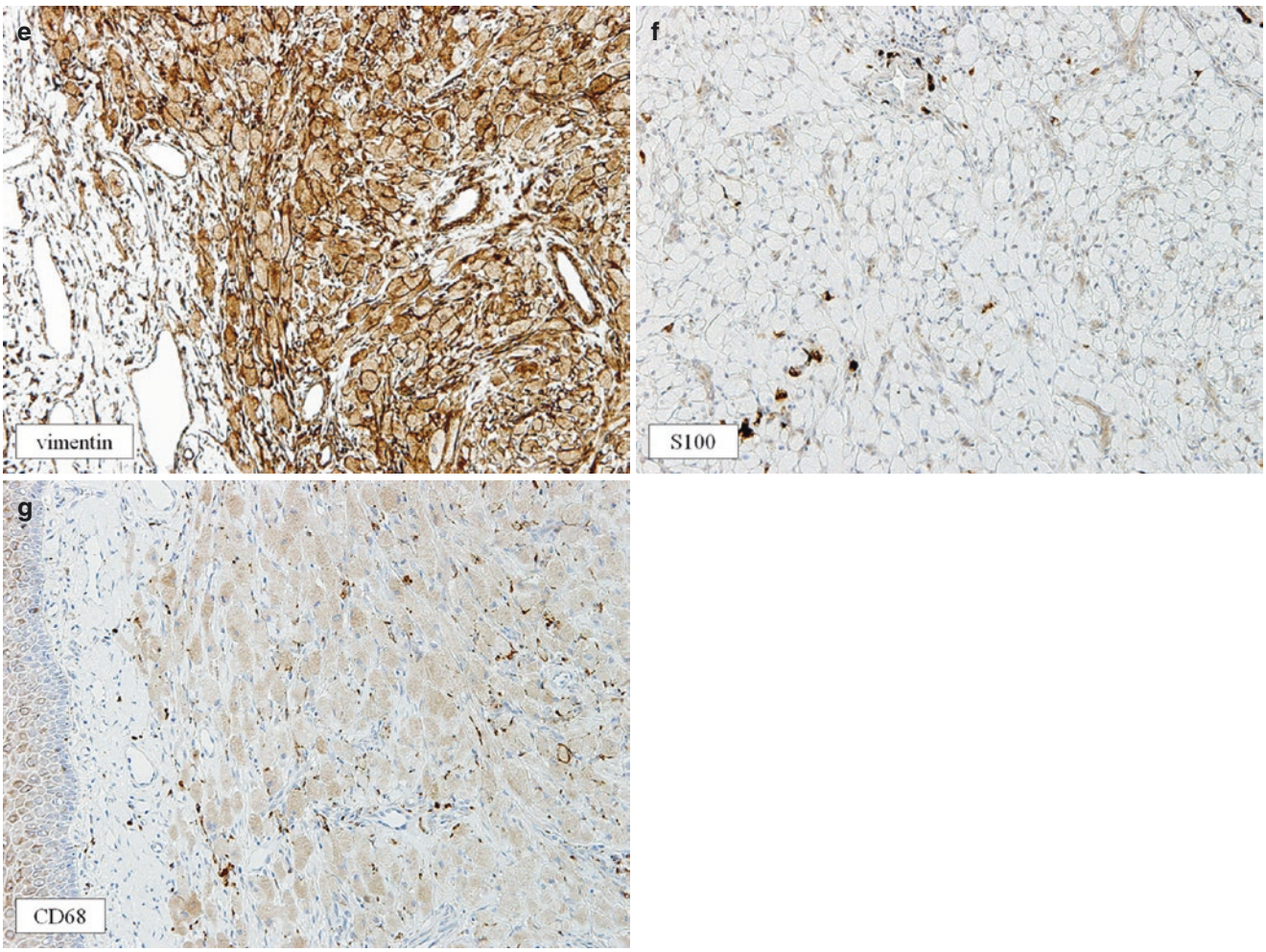

Fig. 12.2 (continued)

Clinical Presentation

Can present as a midline submental mass, usually superior to the hyoid bone or as a sublingual mass or mouth floor whitish mass. Location superior to the geniohyoid and mylohyoid muscles will result with the floor of the mouth swelling where a mass positioned between these muscles would result with a submental bulging [11]. Large cysts may appear in the submental region and in the floor of the mouth as well. Because of its slow-growing nature it is usually apparent in late childhood or adolescence and even later into adulthood. Its contents can be caseous, sebaceous, or purulent, with hair, nails, or fat. Intraoral of extraoral sinus tracts are uncommon [12].

Differential Diagnosis

- Ranula
- Blockage of the submandibular gland duct
- Choristoma
- Foregut duplication cyst
- Cystic teratoma
- Bronchogenic cyst
- Neoplasm of the sublingual or minor salivary glands
- Thyroglossal duct cyst
- Lymphatic malformation
- Benign or malignant neoplasm
- Normal fat of the submental area

Radiological Findings

Ultrasound usually reveals a cystic structure that may be anechoic, hypoechoic, or contains internal debris depending on which germ cell layers are present. CT-scan appearance can be homogeneous or heterogeneous depending on the tissues present. On MRI the T1 signal may be variable but is usually T2 hyperintense. A marble-like appearance is possible when macroscopic fat is present therefore a fat-saturation sequence can be of value when a dermoid is suspected [13].

Management

Surgical treatment would be the preferable choice. Planning of the appropriate approach, external or trans-oral, could be based on the imaging relations of the cyst with the surrounding musculature. Intraoral aspiration or decompression is not rou-

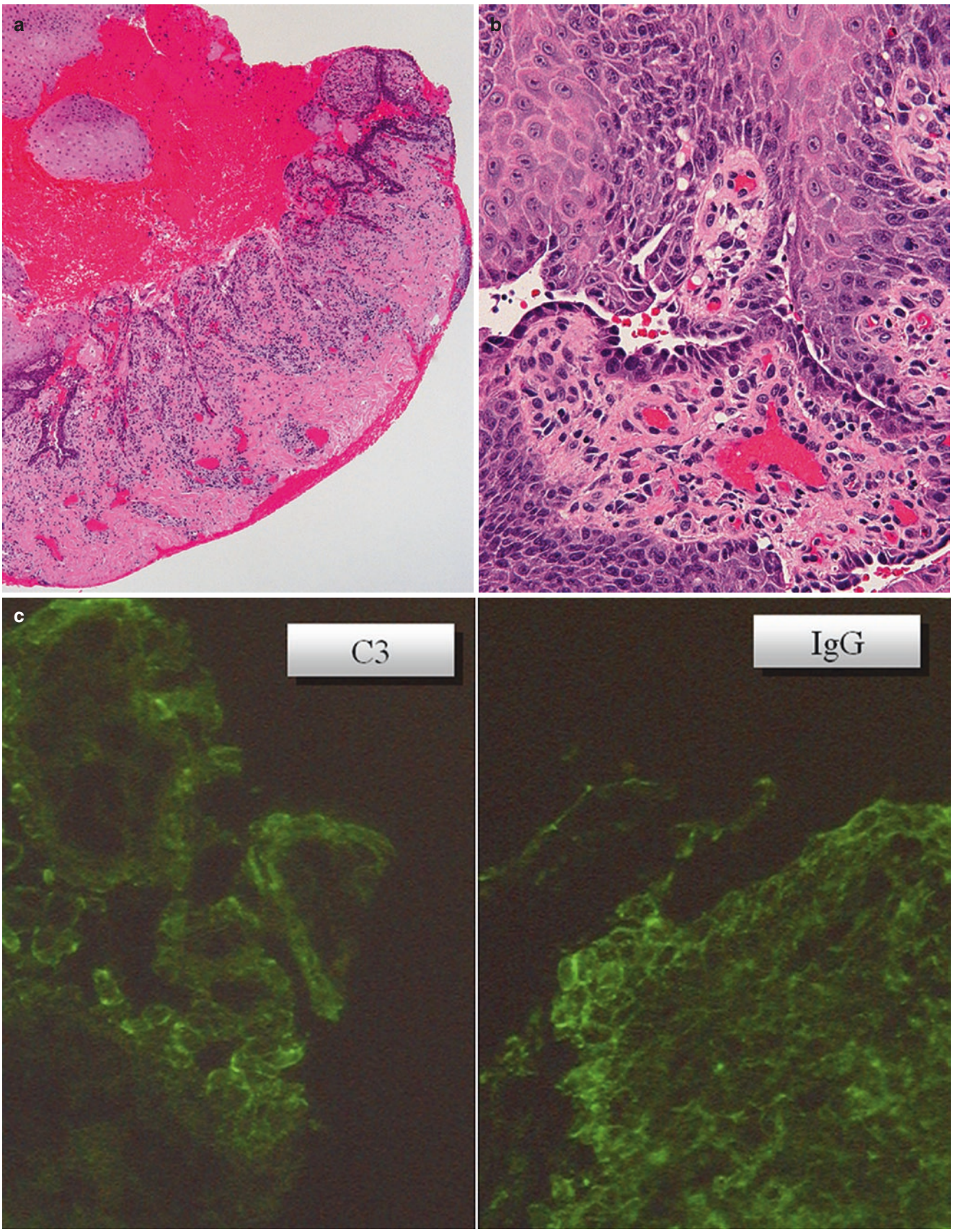

Fig. 12.3 (**a**) H&E. Showing the roof of the blister contained the full thickness of the oral squamous epithelium. Blood filled the blister cavity. (**b**) H&E. High magnification of an area within the blister where the blister appeared to have separated above the basal cell of the epithelium: a feature of pemphigus. (**c**) Immunofluorescence stain for C3 and IgG. There was 3+ granular intercellular staining of the epithelium for IgG and C3. Staining for IgA and IgM was negative (not shown). This finding was in support of pemphigus

tinely used but could be an option when airway compromise is imminent followed by a formal complete excision [11].

Clinical Examples

Case Presentation 1

A 1-year-old female presented with a cystic lesion under the tongue since birth. There was no functional impairment but recurred after a marsupialization procedure. Definitive excision was performed through a transoral approach. The pathology indicated that this lesion was choristoma due to the presence of two or more distinct types of epithelium (Fig. 12.4).

Case Presentation 2

A 1-month-old male presented with a floor of mouth mass since birth without any symptoms. It was excised through a

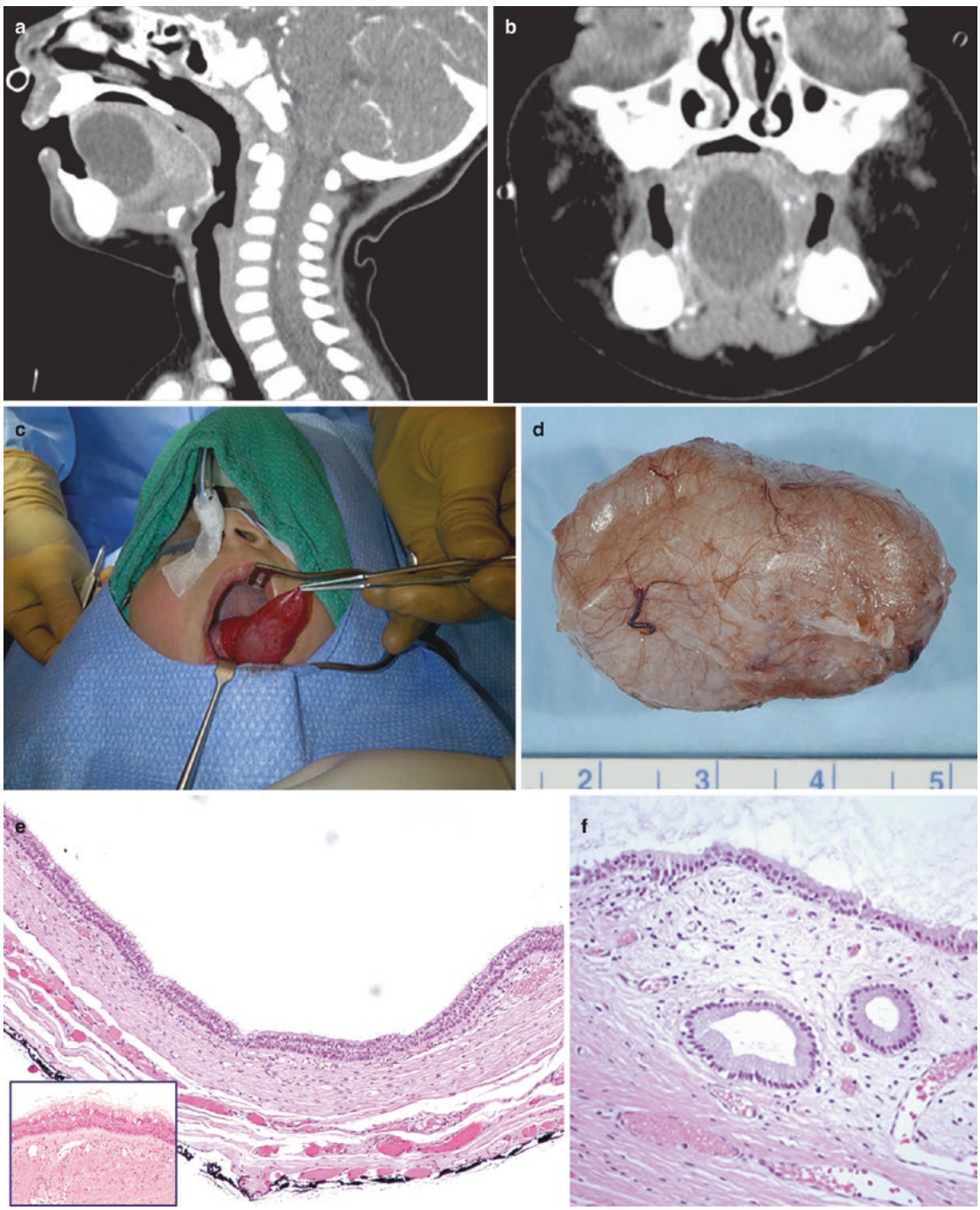

Fig. 12.4 (**a**) Sagittal T1 MRI showing cyst in tongue. (**b**) Coronal T1 MRI showing cyst in tongue. (**c**) Large cyst with well-defined capsule at surgery. (**d**) Gross appearance of cyst. (**e**) The lining consisted mainly of pseudostratified respiratory ciliated epithelium and the cyst wall of fibrovascular connective tissue. The cyst wall was partially surrounded by smooth muscle fibers but there was no distinct muscularis. Insert shows the presence of goblet cells among the ciliated epithelium. (**f**) This slide demonstrates gastric foveolar epithelium and gastric-type glands. (**g**) There is a small focus of squamous non-keratinizing epithelium

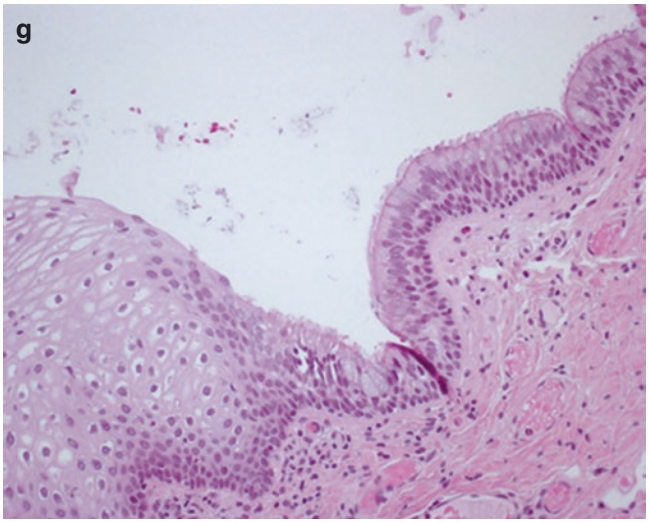

Fig. 12.4 (continued)

transoral approach with bipolar cautery. It was distinct from the surrounding salivary glands (Fig. 12.5).

Pathologically, this was a benign developmental cyst which could represent a congenital ranula or a monodermal cystic teratoma.

Case Presentation 3

A 12-year-old female presented with a submental fullness and dysphagia. Imaging indicated a well-circumscribed round mass in the floor of the mouth and neck with features suggesting a dermoid cyst. This was removed through a transoral approach. The well-formed thick capsule allows for grasping and retraction. Decompression with aspiration of the thick contents prior to removal was not necessary but can be performed if required (Fig. 12.6).

Oral Pseudocyst (Ranula)

Definition

A mucocele formed by mucus extravasation in a proper layer beneath the oral epithelium to form a pseudocyst, means that no epithelial lining exists [14]. Its source is usually the sublingual or minor salivary glands resulting in ranula formation. Ranulas are divided into a simple intraoral which are more common and cervical or plunging. The simple ranulas are located in the floor of the mouth while the plunging are located in the infra mylohyoid compartment and may extend into the neck [15]. The etiology of a plunging ranula is speculated to be either by entering the facial plane between the mylohyoid and the hyoglossus muscles or entering through a defect in the mylohyoid muscle (mylohyoid boutonniere) which may be congenital in origin [15]. Thirty-five percent of the plunging ranulas are combined with an intraoral component [16]. Ranulas may present during the first decade of life while plunging ranulas tend to present during the second and thirds decades of life [14].

Clinical Presentation

Commonly, patients will present with a non-symptomatic blue-domed unilateral cystic lesion in the floor of the mouth. In the case of a plunging ranula, the presentation will usually be of a unilateral submandibular painless fluctuating mass. Aspiration of the cyst will result with saliva rich in protein and amylase consistent with the secretions from the mucinous acini in the sublingual gland [17].

Differential Diagnosis

- Blockage of the submandibular gland duct
- Neoplasm of the sublingual or minor salivary glands
- Lymphatic malformation
- Vascular malformation
- Floor of the mouth developmental cyst
- Benign or malignant neoplasm

Radiological Features

Ultrasound often depicts an ovoid or lobulated cystic lesion, anechoic or with low-level internal echoes, none to minimal peripheral hyperemia is apparent with color Doppler activation. On CT-scan, ranulas have a unilocular homogeneous cystic lesion. MRI findings consist of a thin-walled cystic mass with low intermediate signal on T1-weighted sequence and high signal on T2-weighted images [13]. Plunging ranulas may have a tail seen in CT-scan or MRI that extends into the sublingual gland [18].

Management

Surgery is the mainstay of ranula management and different procedures were reported with different success rates. Intraoral cyst excision accompanied with removal of the sublingual gland has the lowest rates of recurrence though reports suggesting drainage of the cyst with sublingual gland excision, even in plunging ranulas, showed favorable results [19]. However, it is still common to encounter those who would prefer a simple marsupialization with cold instruments or CO_2 laser [20]. OK-432 sclerotherapy was suggested as well [21].

Clinical Examples

Case Presentation 1

A 5-year-old boy presented with a floor of the mouth cystic mass. The mass was completely excised with the adjacent sublingual salivary gland (Fig. 12.7a and b).

Case Presentation 2

A 9-year-old female presented with a 7-month history of a right neck mass and swelling extending to the right submandibular triangle. The mass was soft, compressible, and non-tender. MRI showed a large cystic mass in the floor of the mouth and upper neck (Fig. 12.8a). The mass was

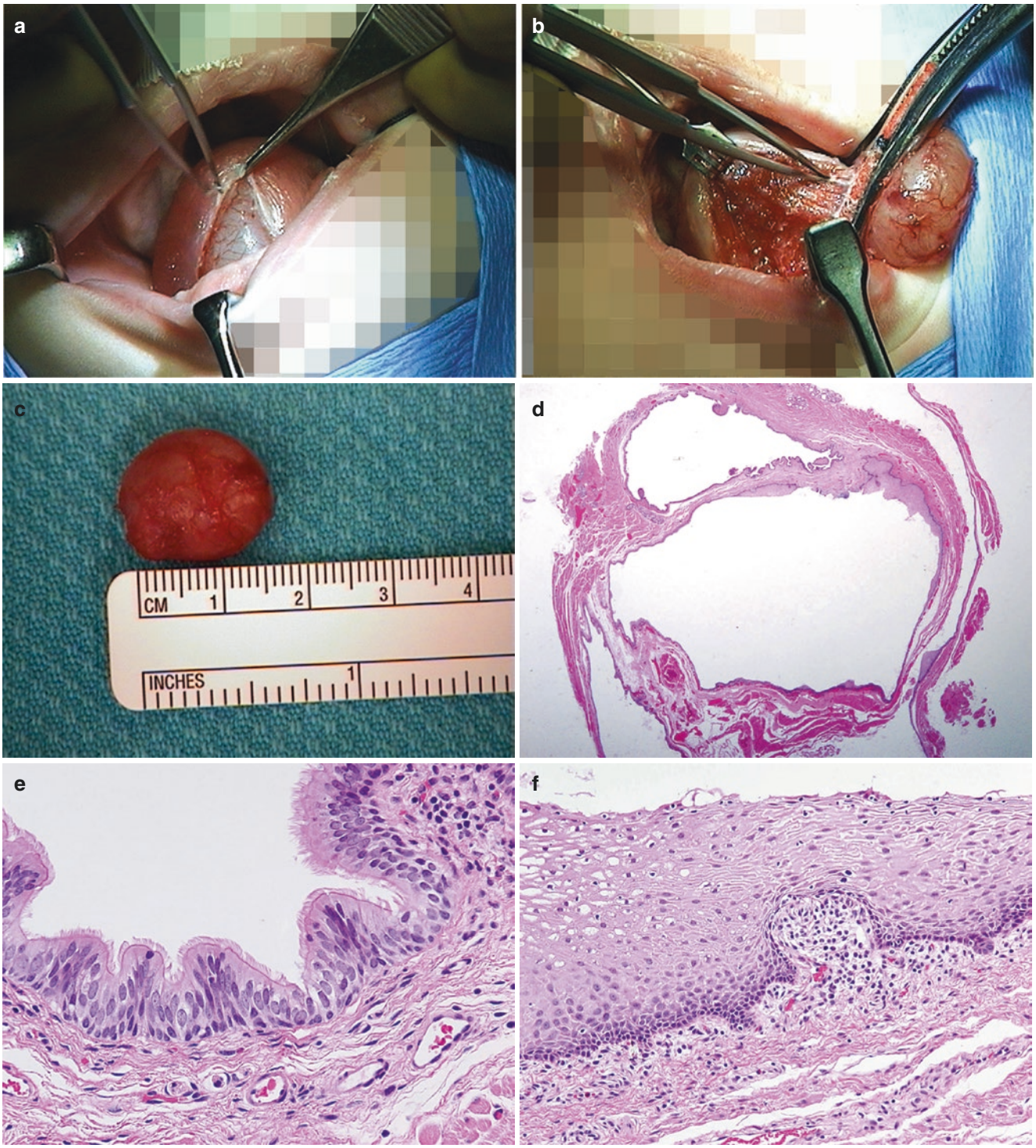

Fig. 12.5 (**a**) Cyst exposed transorally. (**b**) Cyst removed using micro bipolar dissection. (**c**) Gross appearance of cyst. When it is cut open, it contained milky white fluid. (**d**) The wall was thin. 0.1 cm thick and the internal cyst surface was smooth without papillary structures or nodules. (**e**) H&E. Internal surface was lined by ciliated columnar epithelium. (**f**) H&E. Parts of the internal cyst surface was lined by stratified squamous epithelium

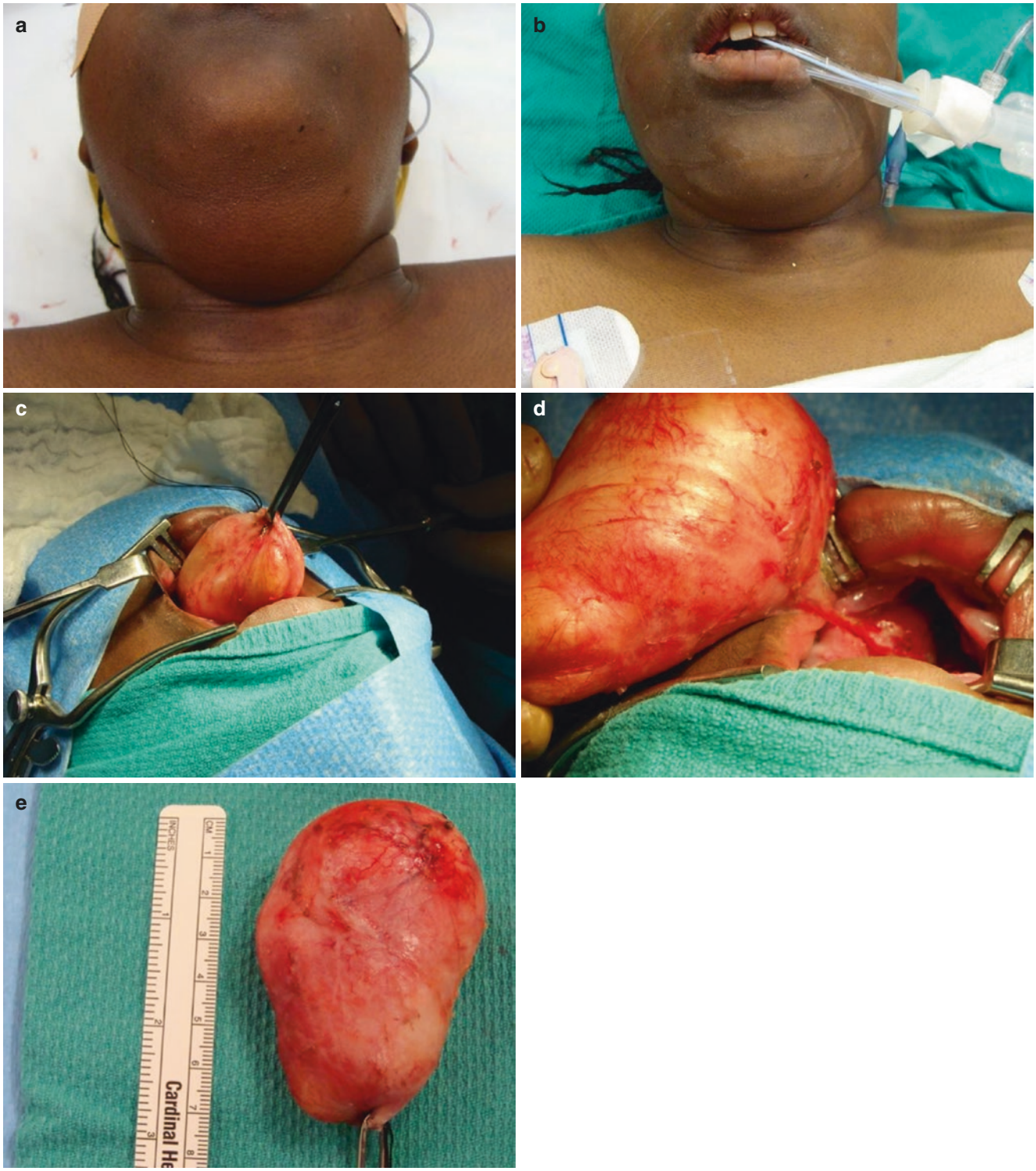

Fig. 12.6 (**a**) Preoperative photo showing large submental cystic mass. (**b**) Postoperative appearance after transoral removal of the submental mass. (**c**) A large dermoid cyst with a firm capsule, making retraction possible, was dissected through a transoral approach. (**d**) Intact cyst removed. Note color of the cyst reflecting a thick whitish "cheesy" content. (**e**) Gross appearance of cyst

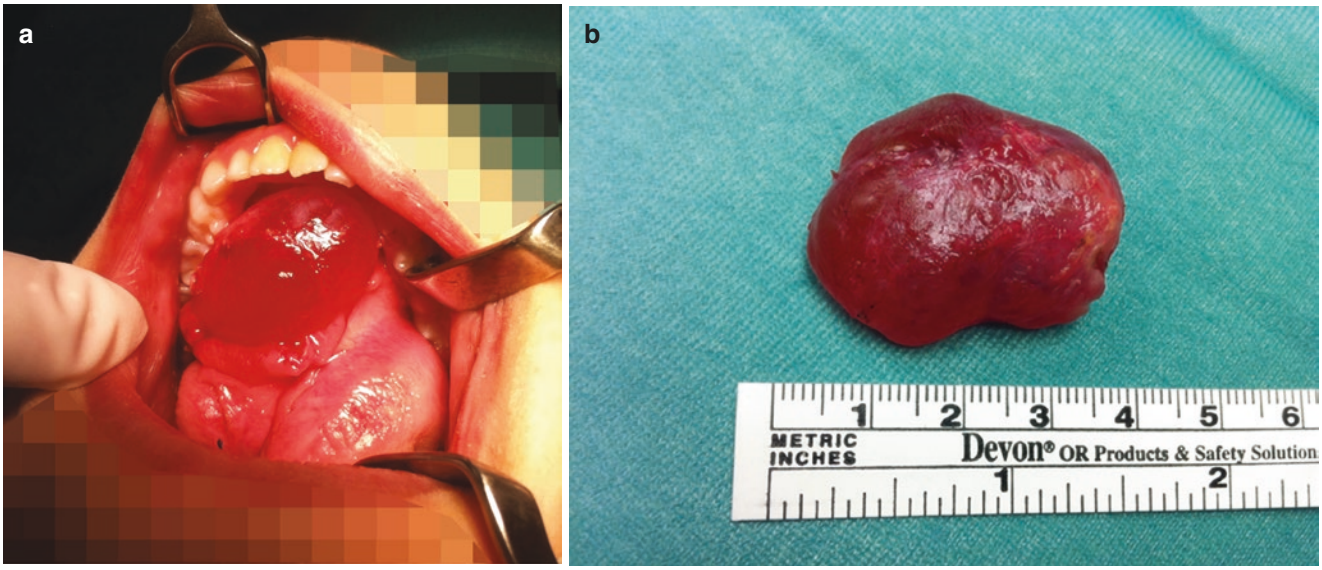

Fig. 12.7 (**a**) Ranula exposed in the floor of the mouth. (**b**) Gross appearance of intact ranula

decompressed, a biopsy taken of the cyst "wall" and the right sublingual salivary gland also excised. Pathology was consistent with a deep plunging ranula (Fig. 12.8b–d).

Microcystic Lymphatic Malformation of the Tongue

Definition

Microcystic lymphatic malformation of the tongue is a macroscopic granular appearance of the anterior two-thirds, involving the lingual dorsum [22]. These numerous granules are lymph-filled cysts with color ranging from transparent to purple in case of capillary rupture into the lymphatic spaces. Recurrent upper respiratory infection or accidental trauma commonly enlarges the lymphatic malformation and tend to worsen the swelling leading to secondary macroglossia. Weigand et al. classified the condition into four groups according to their extent into the tongue musculature [23].

Clinical Presentation

The tongue has a granular appearance; the small cysts color tends to be transparent to purple and they may occasionally bleed. Secondary macroglossia can cause airway obstruction, swallowing difficulties, malocclusion, dental caries, excessive salivation, cosmetic deformity, and speech problems. Most patients will present at birth and during the first decade of life.

Differential Diagnosis

A large intramuscular component can mimic a benign or malignant tumor of the tongue, however, when identifying the typical microcystic structures the diagnosis is quite obvious.

Radiological Features

Imaging would not be necessary in case of a superficial lesion. In case of suspected deep tongue involvement, MRI would be the preferred modality. The lesion will appear with increased signal intensity on T2-weighted images while T1-weighted images have intensity similar or slightly less than that of the surrounding muscles.

Management

Superficial non-symptomatic lesions would not require an intervention and follow-up will do. Surgical intervention in case of symptomatic lesions such as recurrent bleeding or functional interference due to macroglossia would be required. Utilization of the CO_2 laser [23] or low power radiofrequency [24] is reported. Partial resection of the tongue, either a wedge, U-shaped, Jian glossectomy, or Dingman glossectomy, is reported in severe cases [25].

Clinical Example

A two-year-old girl presented with numerous microcysts (Fig. 12.9a) on the tongue surface, extended posteriorly and covering completely the anterior two-thirds. Note the hypertrophied tongue with the bifid appearance and a few small purple hemorrhagic microcysts. An axial T2-weighted MRI demonstrates the bifid appearance with the lesion that partially involves the right part of the tongue (Fig. 12.9b). The cysts were treated by repeated ablation sessions with CO_2 laser (Fig. 12.9c) and a partial wedge resection of the tongue.

Enteric Duplication Cyst

Definition

A cyst attached to the alimentary tract and lined with some type of gastrointestinal mucosa. This duplication cyst can be found

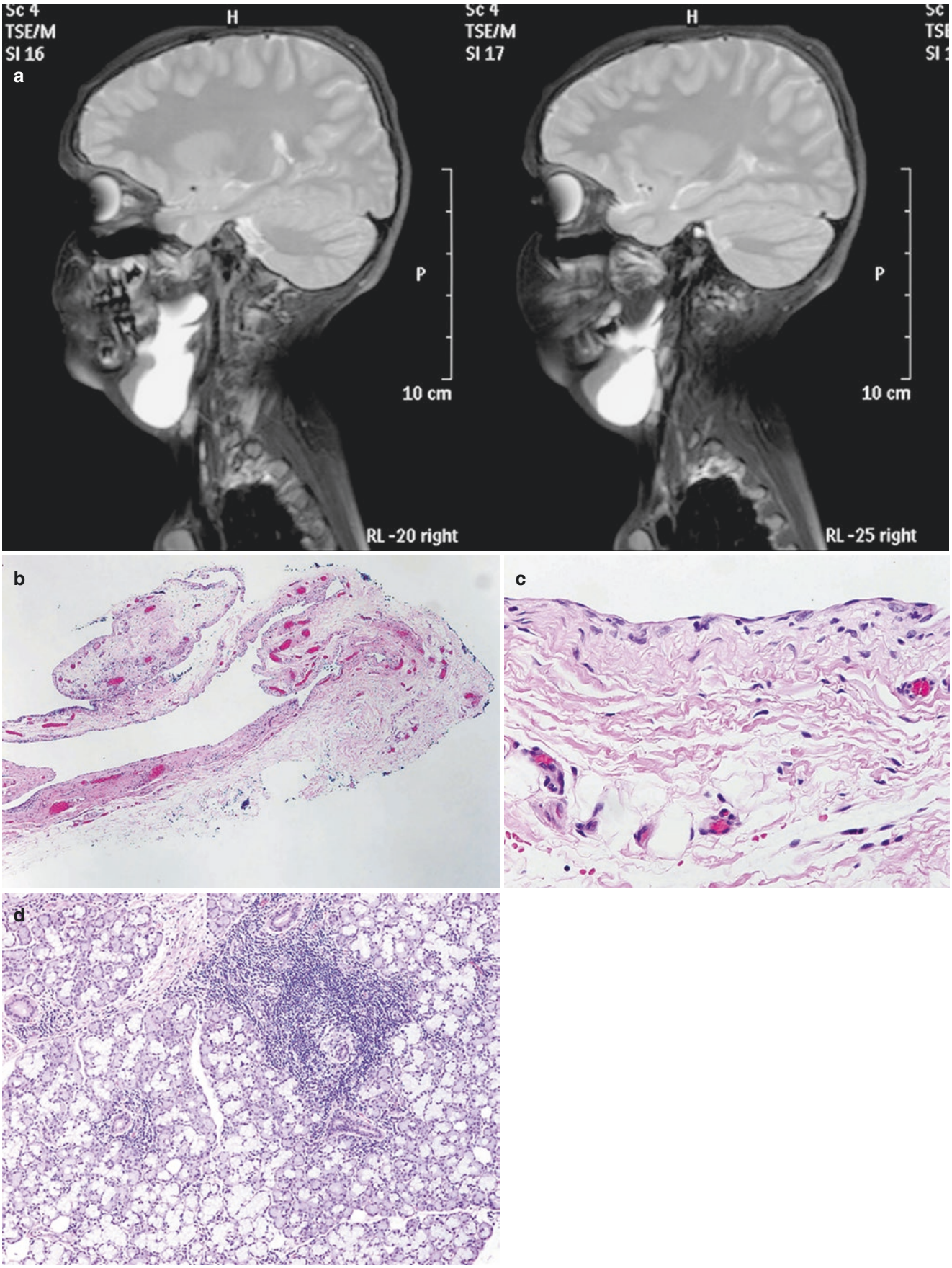

Fig. 12.8 (**a**) Sagittal T2 MRI showing a large collection of fluid in the neck. (**b**) H&E. A collapsed large thin wall cyst with a partially fibrotic wall. It can be consistent with a cyst of a ductal origin. (**c**) H&E. Simplified mucosal epithelial cells lined the remnant of a duct wall with layered structural organization. (**d**) H&E. Portion of the cyst was associated with salivary gland ducts with reactive lymphoid hyperplasia

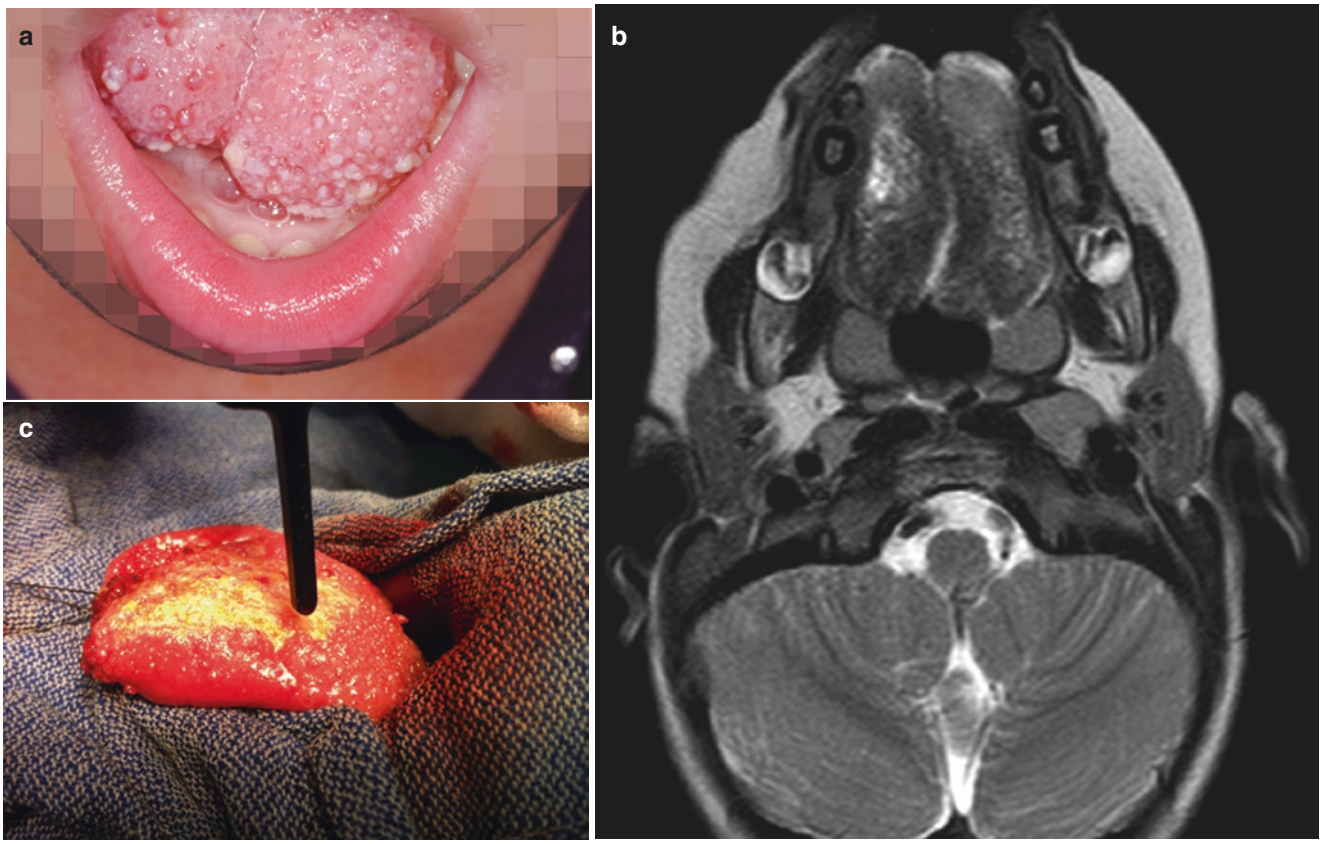

Fig. 12.9 (**a**) "Vesicular" appearance of the tongue seen with microcystic lymphatic malformation. Vesicles appear "pus," clear or "bloody" filled. (**b**) Axial T2-weighted MRI demonstrates the bifid appearance of the tongue with a bright lesion that partially involves the right side of the tongue. (**c**) Laser treatment of the mucosa of the tongue removing/unroofing the microcysts offers good relief but unlikely cure of the malformation

anywhere in the alimentary tract from the oral cavity to the rectum [26]; however, its existence in the oral cavity is very rare [27, 28] and the anterior two-thirds of the tongue is the commonest site [29]. It presents usually at birth or early childhood [27].

Clinical Presentation

Typically the neonate or young child would be asymptomatic. Concern is associated with airway compromise and feeding difficulties, which can be relevant with large volume cysts.

Differential Diagnosis

- Pseudocyst (ranula)
- Lymphatic malformation
- Developmental cyst
- Hemangioma
- Thyroglossal duct cyst

Radiological Features

MRI is the imaging modality of choice where T1-weighted images would appear non-enhanced depending on the proteinaceous component; high levels of protein would cause a hyperintense appearance. T2-weighted images would appear hyperintense.

Management

Surgical removal of the cyst would be appropriate taking care of significant anatomical structures in the surroundings.

Melanotic Macule (See Also Chap. 13)

Definition

Melanotic macule is a small, solitary well-circumscribed brown to black macule that occurs on the lips or mucous membrane and comprises the vast majority of melanocytic lesions in the mouth [30]. Hyperpigmentation is in the basal membrane or in the lamina propria or both [31].

Clinical Presentation

The lesion is asymptomatic and would be identified incidentally by the patient or his parents.

Differential Diagnosis
- Melanocytic nevus
- Atypical melanocytic hyperplasia
- Melanoacanthoma
- Epidermal choristoma
- Peutz-Jeghers syndrome
- Melanoma

Radiological Features
No radiological workup is indicated.

Management
Surgical excision if atypical or any doubt in diagnosis.

Fetal Rhabdomyoma

Definition
Fetal rhabdomyoma is a rare benign tumor that is subdivided into myxoid and cellular types. Myxoid type would mainly appear during childhood subcutaneously in the retroauricular region while the cellular type appears in the deep tissues of the head and neck region, such as the tongue, nasopharynx, larynx, orbit, and neck [32].

Clinical Presentation
Rhabdomyoma would appear as a smooth mass at the base of the tongue or at the anterior part, usually non-symptomatic dependent on size and location.

Differential Diagnosis
- Rhabdomyosarcoma

Radiological Features
MRI would be the preferable imaging study.

Management
Surgical excision is required and follow-up is recommended since it tends to recur.

Case Presentation
A 1-year-old male presented with a 2 cm midline dorsal tongue lesion. It had a multilobulated appearance but was asymptomatic (Fig. 12.10).

Hamartoma of Tongue

Definition
Hamartoma is a growth of normal tissues endogenous to the site of occurrence. Endogenous elements within the tongue that might result in a Hamartoma would include vessels, nerves, lymphatics, skeletal muscle, fat, and salivary gland tissue [33]. Gender distribution is equal [34]. Usually an isolated lesion is found, however, multiple hamartomas are reported in association with various syndromes, such as cleft palate, tuberous sclerosis, and oral-facial-digital syndrome [33, 34].

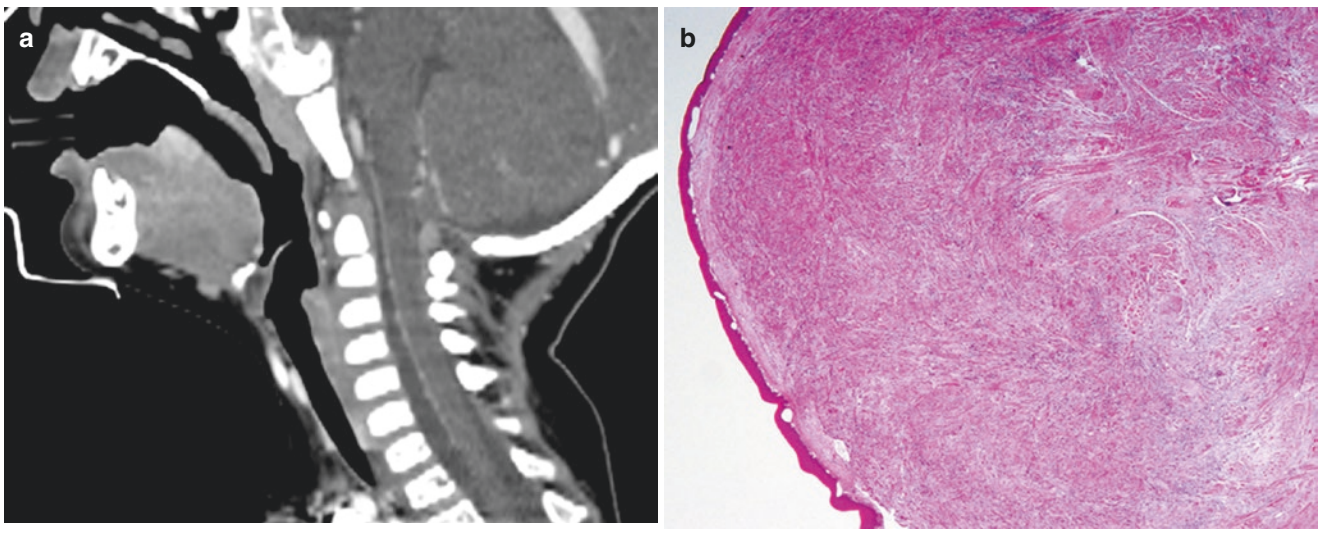

Fig. 12.10 (**a**) Sagittal MRI demonstrates "nodular" appearance of dorsal surface of the tongue without evidence of any deeper involvement. (**b**) H&E. A relatively ill circumscribed, un-encapsulated mass within the tongue. It has compact proliferations of small spindle cells. (**c**) H&E. The spindle cells infiltrated and spread the skeletal muscle cells of the tongue apart. The tumor cells have some eosinophilic fibrillar cytoplasmic contents. (**d**) Masson Trichrome connective tissue stain marking collagen as green color. Stain result showed the presence of fibrosis at the periphery of the mass (left side of figure). In the right side, high density of smaller tumor cells is present. (**e**) Immunostain shows positive cytoplasmic staining of the normal skeletal muscle cells on the left side of the figure (positive control) as well as the cytoplasm of the tumor cells (right side of the figure). These tumor cells show positive nuclear staining for expression of myogenin (a transcription factor for muscle cell differentiation). The tumor has a low nuclear proliferation index of 5% as seen in immunostain with Mib-1

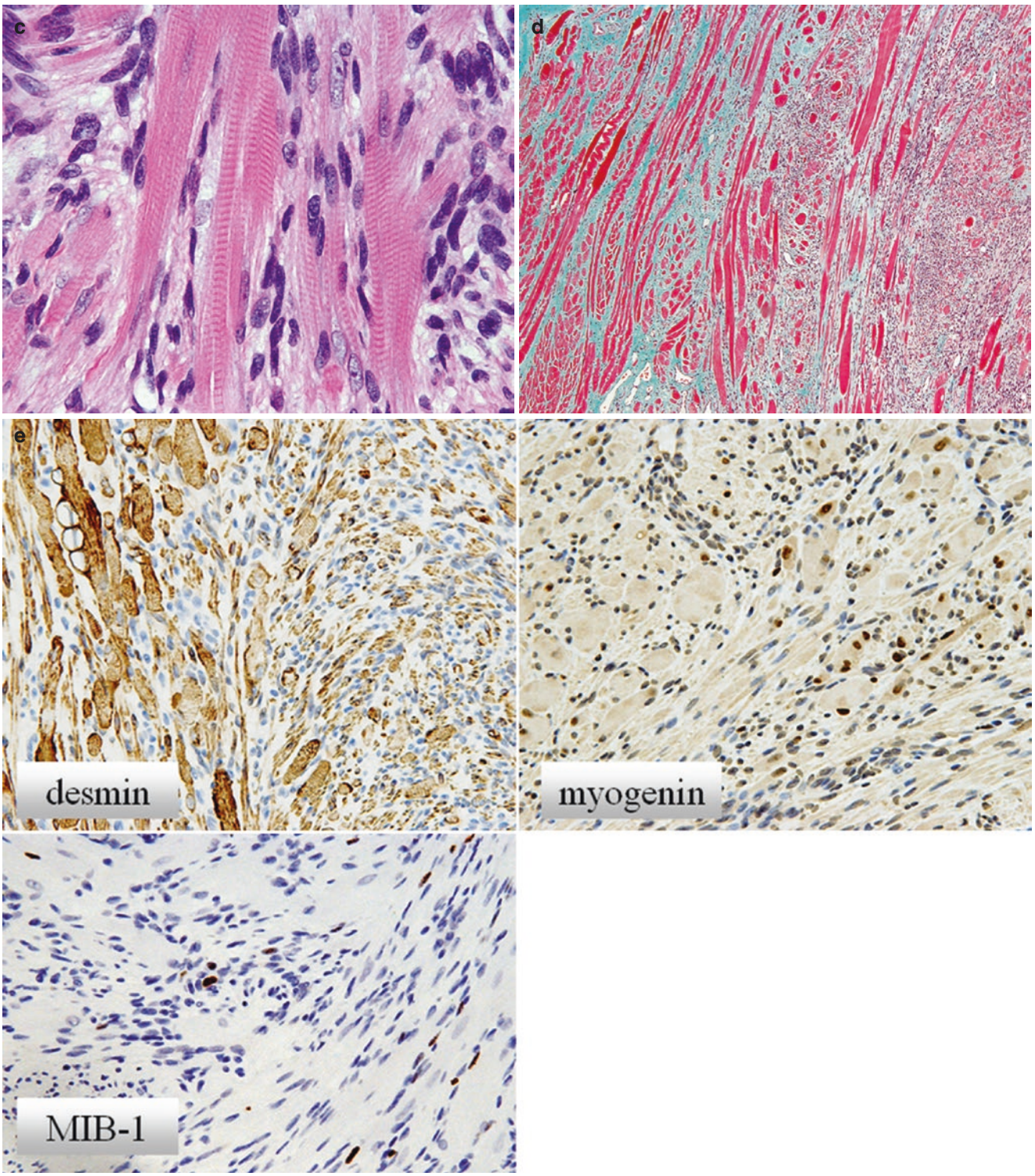

Fig. 12.10 (continued)

Clinical Presentation

Hamartoma would mostly appear as a non-symptomatic mass up to two centimeters, polypoid, and pink to fleshy color, frequently located in the midline.

Differential Diagnosis

- Rhabdomyoma
- Leiomyoma
- Developmental cyst

- Hemangioma
- Thyroglossal duct cyst
- Enteric duplication cyst

Radiological Features

CT-scan of the lesion show minimal contrast enhancement. T1-weighted MRI showed hypointense signal and heterogeneous on T2-weighted images [35].

Management

Surgical excision.

Clinical Examples

Case Presentation 1

A one-month-old baby girl presented with a medial posterior tongue mass (Fig. 12.11a), she was asymptomatic with no feeding difficulties. When she was 1 year old an MRI was

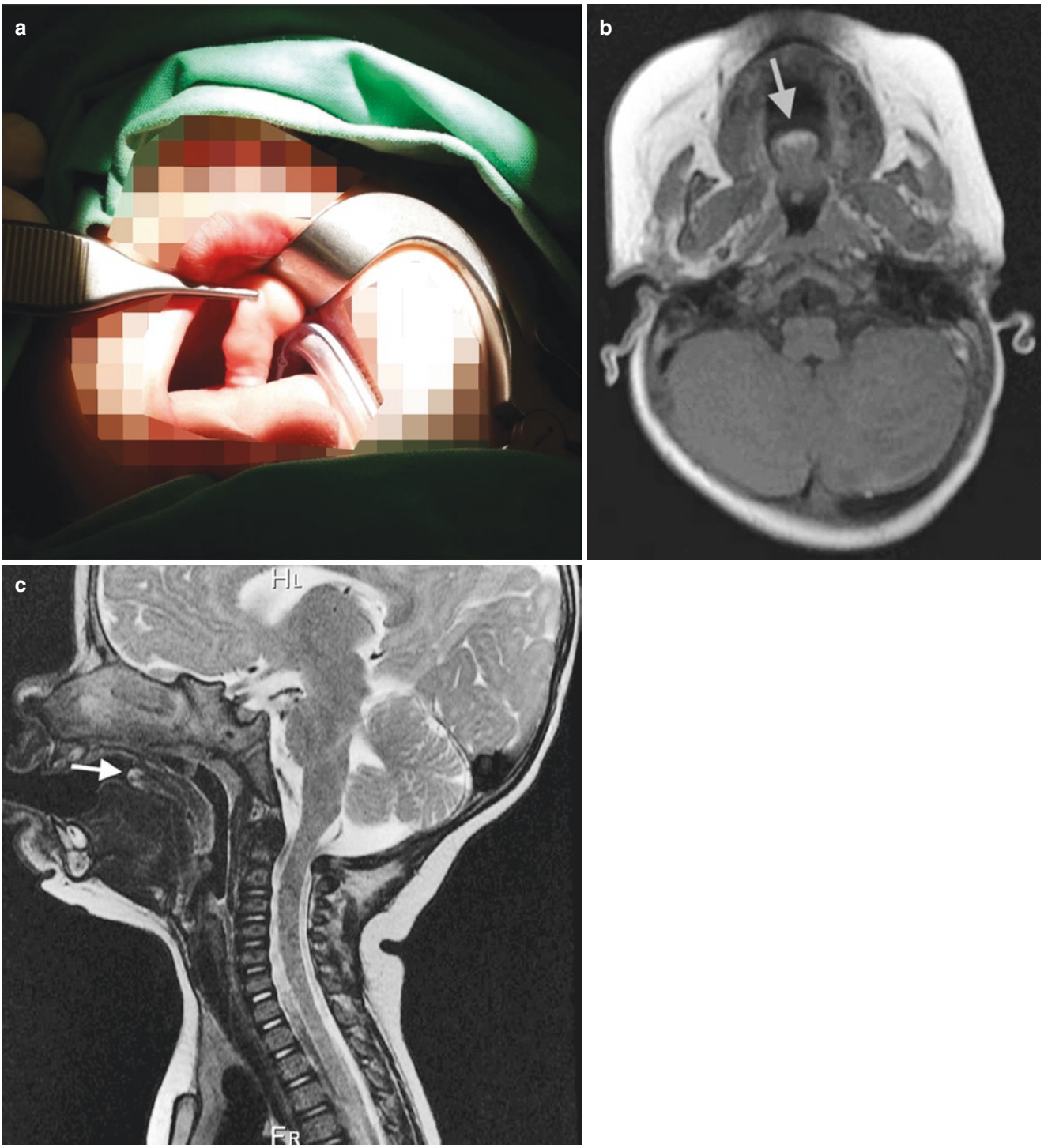

Fig. 12.11 (**a**) Well epithelialized polypoid mass from tongue. (**b**) Axial T1 MRI shows "polypoid" tongue mass (arrow) to be isointense to the normal tongue base tissue. (**c**) Sagittal T2 MRI showing no change in intensity of the mass from the T1 image in the previous figure

performed showing the mass to be of similar intensity in axial T1-weighted and sagittal T2-weighted sequences (Fig. 12.11b, gray arrow and Fig. 12.11c, white arrow). The mass was uneventfully excised.

Case Presentation 2

Newborn with a central tongue mass that caused dysraphism of the tongue, splitting it into 2 pieces (Fig. 12.12).

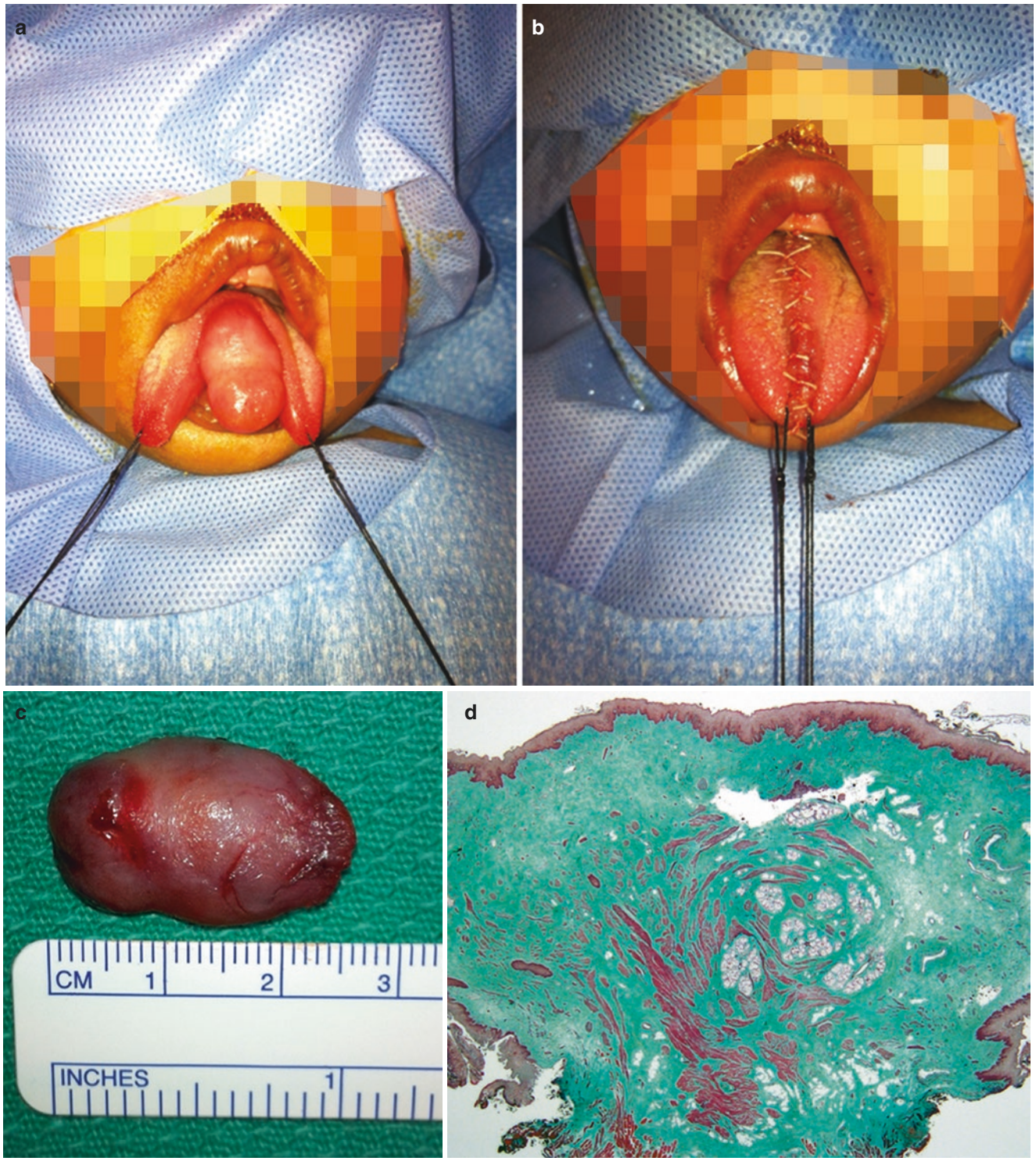

Fig. 12.12 (**a**) Mass causing dysraphism of the tongue in a newborn. (**b**) Layered closure after removal to the mass. (**c**) Gross appearance of the well epithelialized, firm mass. (**d**) Masson Trichrome stain showing an overview of tissue components within this fibrotic mass. (**e**) Masson Trichrome stain showing the presence of haphazard arrangements of skeletal and smooth muscle bundles intercalated by adipose tissues within a (green color staining of) collagenous stroma. (**f**) Masson Trichrome stain showing a fibrotic stroma containing haphazard smooth muscle bundles around minor salivary glands. (**g**) H&E. A vein with smooth muscles of irregular thickness was seen next to developmentally malformed small bundles of smooth muscle and adipose tissue

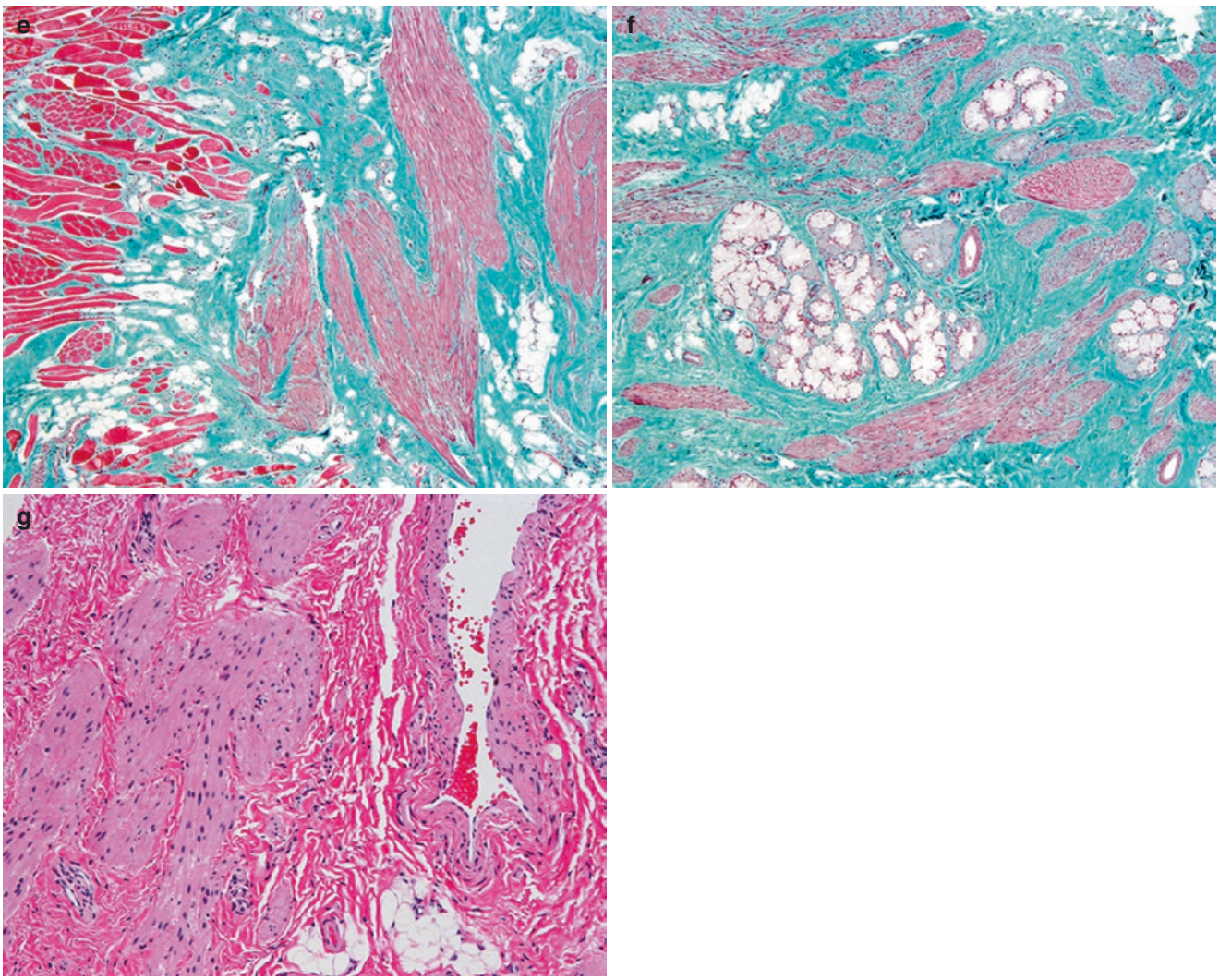

Fig. 12.12 (continued)

Bone (Mandible, Maxilla, Zygoma)

Benign

Osteomyelitis of Mandible

Definition

Osteomyelitis is defined as an infection of the cortical and cancellous bone. The mandible is the most frequently involved site in craniofacial osteomyelitis. Odontogenic bacterial infection is usually the primary site, but other etiological factors are reported [36]. Chronic bacterial osteomyelitis is defined as an infection that lasts more than four weeks and is characterized by suppuration, bone sequestration, and fistula formation. The bacterial etiology workup may remain sterile [37]. Primary chronic osteomyelitis is a similar condition with no apparent odontogenic cause characterized by painful swelling, and trismus. Recurrence is not infrequent after treatment and spontaneous remission is usually reported [38].

Clinical Presentation

Osteomyelitis secondary to an odontogenic infection in its acute phase or chronic phase may present differently than primary chronic osteomyelitis. The former may present at any age where the latter onsets during puberty, according to a case series reported by Eyrich et al. [36] The most frequent symptoms are pain, swelling, facial asymmetry, local induration, and trismus. Some patients will present with lymphadenopathy and hypoesthesia of the mandibular nerve [36]. Acute osteomyelitis secondary to an odontogenic infection is accompanied with severe form of pain.

Differential Diagnosis

- Osteosarcoma
- Fibrous dysplasia
- Non-ossifying fibroma
- Salivary gland infection
- Lymphadenitis

Radiological Features

CT-scan features of chronic osteomyelitis of the mandible include enlargement of the mandibular nerve canal, bone sclerosis, lytic lesions, ground glass pattern, and lamellate form of the periosteal bone. The surrounding soft tissues such as the masseter muscle, lymph nodes, and skin are involved as well [37].

Management

Long-term parenteral antibiotics are the mainstay of treating bacterial osteomyelitis. Resection of necrotic bone is frequently required; decortication is sufficient in milder cases. Marginal or segmental resection is required in cases with fistula formation [38].

Clinical Example

A 10-year-old female presented with a several month history of swelling of her right cheek and mandible that caused pain on mastication. There had been no history of trauma or dental treatment and she was otherwise completely healthy. The CT scan (Fig. 12.13a) shows expansion of the ramus of the right mandible. A transoral biopsy was performed that confirmed a diagnosis of chronic osteomyelitis (Fig. 12.13b–e).

Non-odontogenic Benign Tumors

Fibromas of the Maxilla and Mandible

Definition

Fibroma is a benign tumor arising in a fibrous tissue. It may arise in soft tissues or bones. Fibromas were reported as involving the mandible and the maxilla in the pediatric age

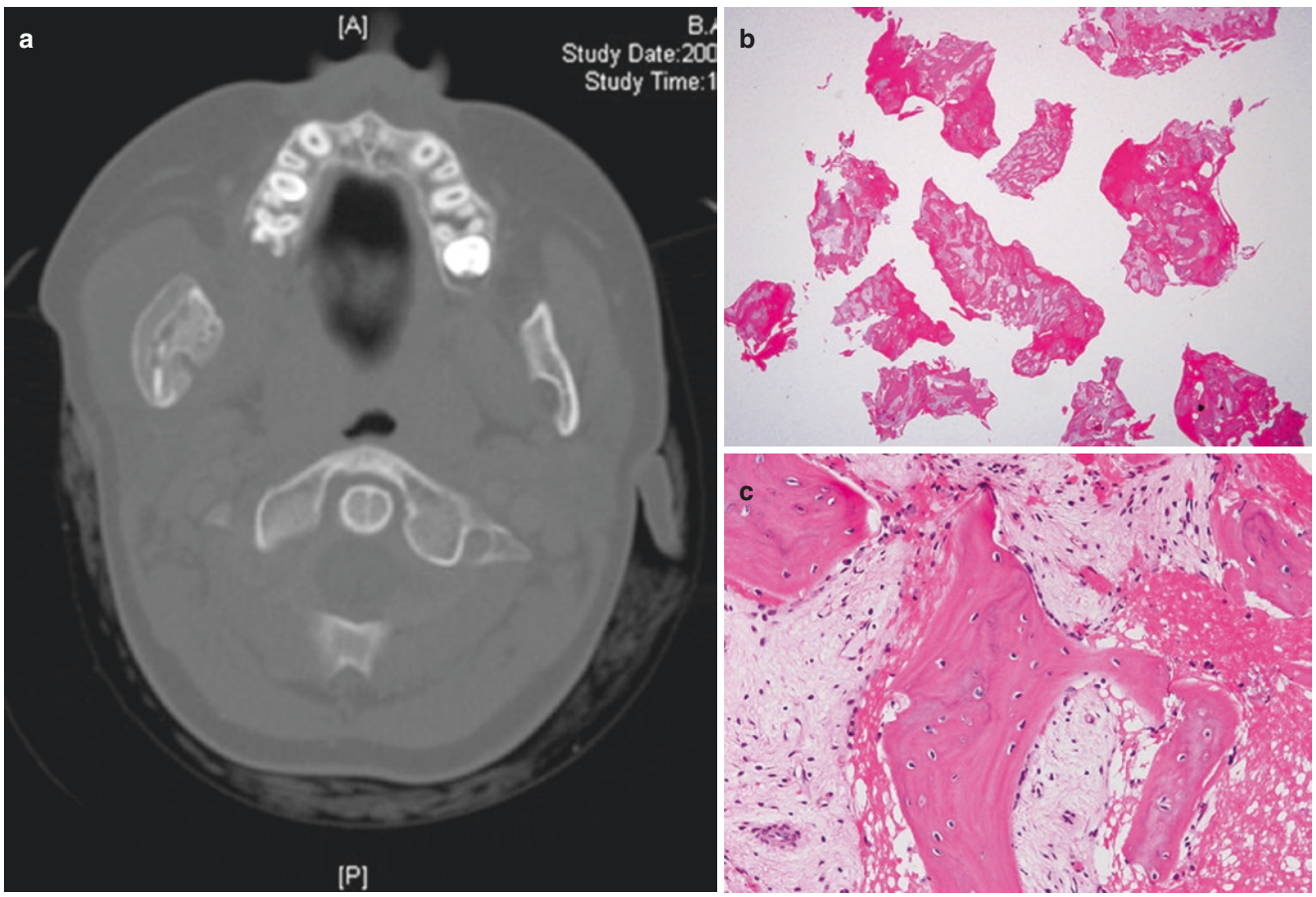

Fig. 12.13 (**a**) Axial CT without contrast demonstrated enlargement of the vertical ramus of the mandible. (**b**) H&E. Bone fragments containing uniform bone trabeculae. (**c**) H&E. Normal mature bone with focal reactive osteoblast proliferations associated with mild bone marrow fibrosis. (**d**) Composite images of H&E stain illuminated under ordinary light versus polarized light which brought permit the visualization of the collagen matrix within the bone trabeculae. The bone showed structure of woven and lamellar bone. Results confirmed normal bone formation with newly onset of reactive osteoblast proliferations. (**e**) H&E. Typical features of chronic osteomyelitis, where there are increased presence of reactive plasma cells

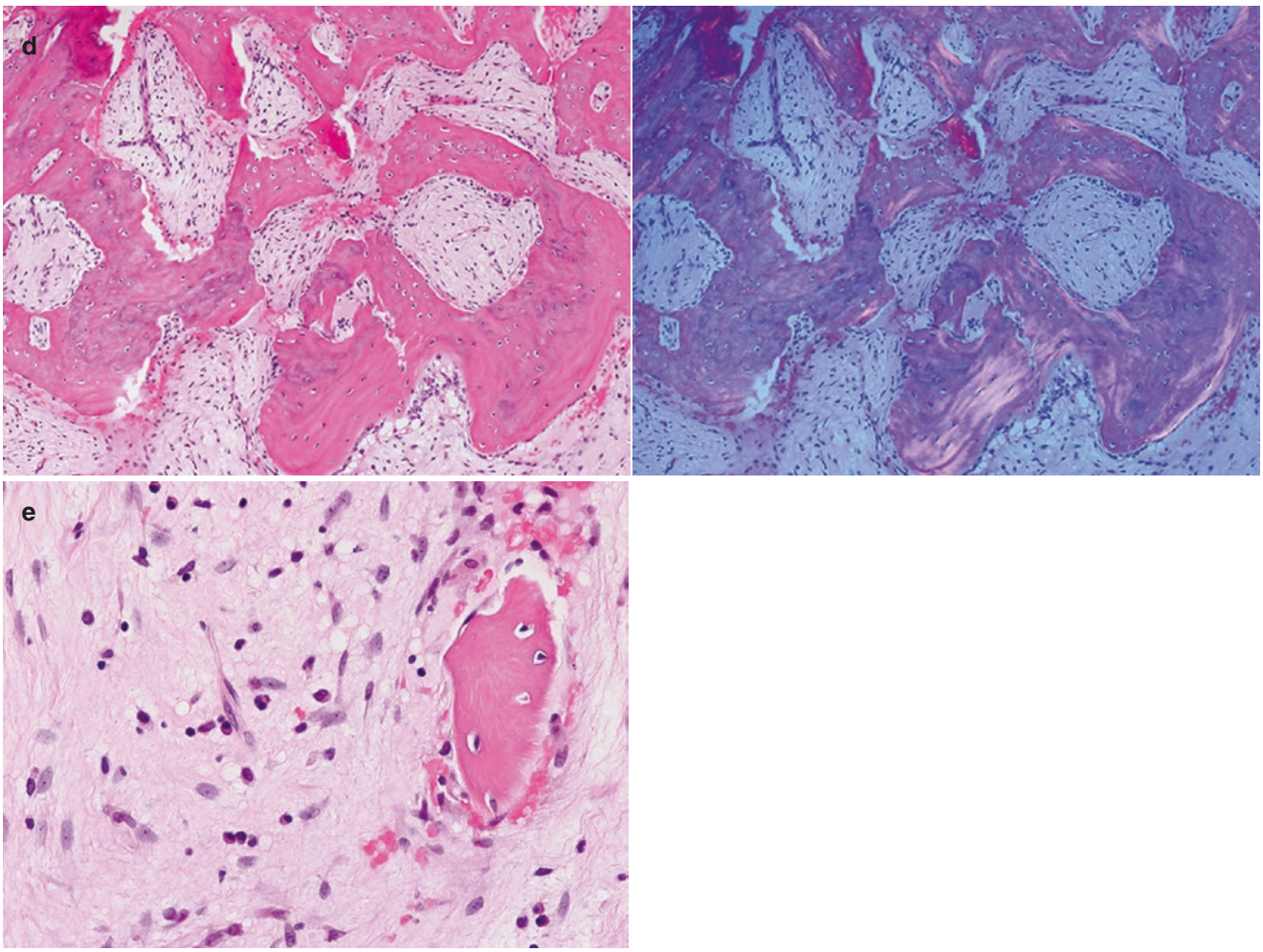

Fig. 12.13 (continued)

group and adolescence. Various types of this type of tumor were described and most are non-frequent to esoteric and may differ in their histological appearance and clinical behavior. One may encounter a fibroma, juvenile ossifying fibroma, desmoid fibroma, myofibroma, and chondromyxoid fibroma [39]. Multiple ossifying fibromas are extremely rare but may be associated with hyperparathyroidism-jaw tumor syndrome and requires further workup as serum PTH testing [40].

Clinical Presentation

Most patients present with facial swelling evolving in days to months accompanied occasionally with pain and restriction of movement [41–43].

Differential Diagnosis

- Benign or malignant non-odontogenic or odontogenic neoplasms
- Fibromatosis of the jaws
- Osteomyelitis

Radiological Features

On radiographs many lesions are radiolucent and some show a combination of radiolucency and radiopacity. Evidence of root resorption and tooth displacement is common as well [42].

CT-scan appearance of those lesions may show a lytic lesion with calcifications that invades the soft tissue with periosteal reaction [39].

Management

Enucleation or curettage of the tumor is the most common advised, occasionally in combination of the two [39]. In most of the cases the tumor does not tend to recur, however, some will, especially juvenile ossifying fibroma, which tend to be more aggressive and requires a more definitive approach.

Clinical Examples

Case Presentation 1

A 9-year-old female presented with 6-week history of a painful left mandible mass. The dentition in the area of the mass was loose. CT imaging (Fig. 12.14a–b) revealed a hypo

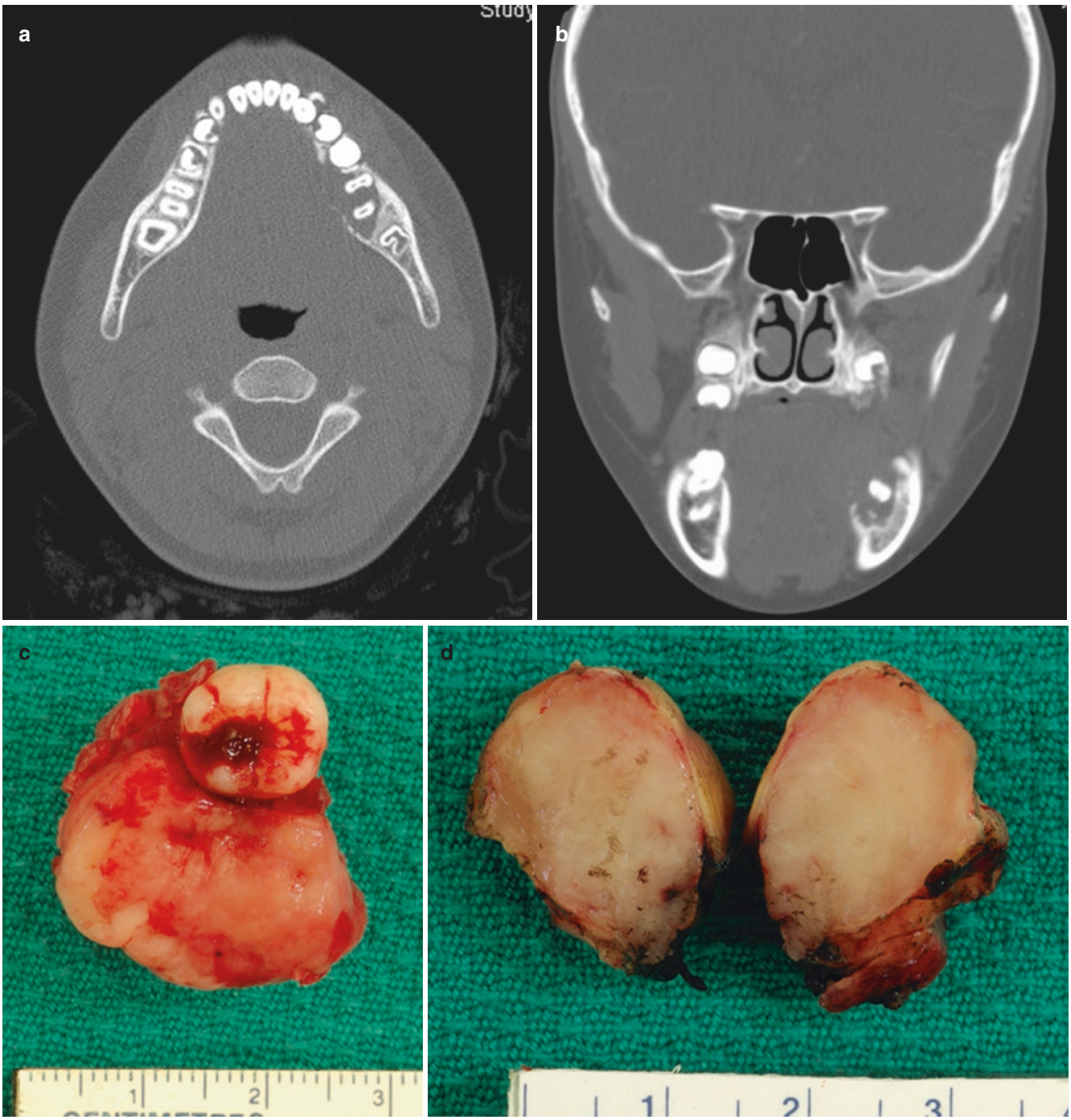

Fig. 12.14 (**a**) Axial CT revealing a hypo lucent expansile mass in the left ramus of the mandible. (**b**) Coronal CT showing hypo lucent expansile mass in the left ramus of the mandible. (**c**) Gross photo of fleshy growths of pale tan color rubbery tissue encasing a tooth. (**d**) Gross appearance of the gray tan color cut surface of the rubbery solid tissue growth beneath the oral epithelium. (**e**) Masson trichrome stain showing the presence of patchy areas of mild collagen (in light green color) deposits within the tumor parenchyma. The tumor cells were stained red and they showed a mildly whorled and streaming growth pattern. (**f**) H&E. plump-spindle shaped tumor cells growing as short interlacing fascicles. (**g**) H&E. Spindle-shaped tumor cells with fine eosinophilic cytoplasmic filaments. Mitosis (not shown) were low. (**h**) Immunostain showed only expression of smooth muscle actin in this panel of markers. (**i**) Electron micrograph. Tumor cells have fine filamentous cytoplasmic contents with moderate endoplasmic reticulum with mildly dilated cisternae. Some myofibroblastic feature such as spotty condensations of filaments on the cell surface (fibronexus-like structure) seen in the upper edge of the end of the lacuna like cavity formed by a thin cytoplasmic projection of this myofibroblast

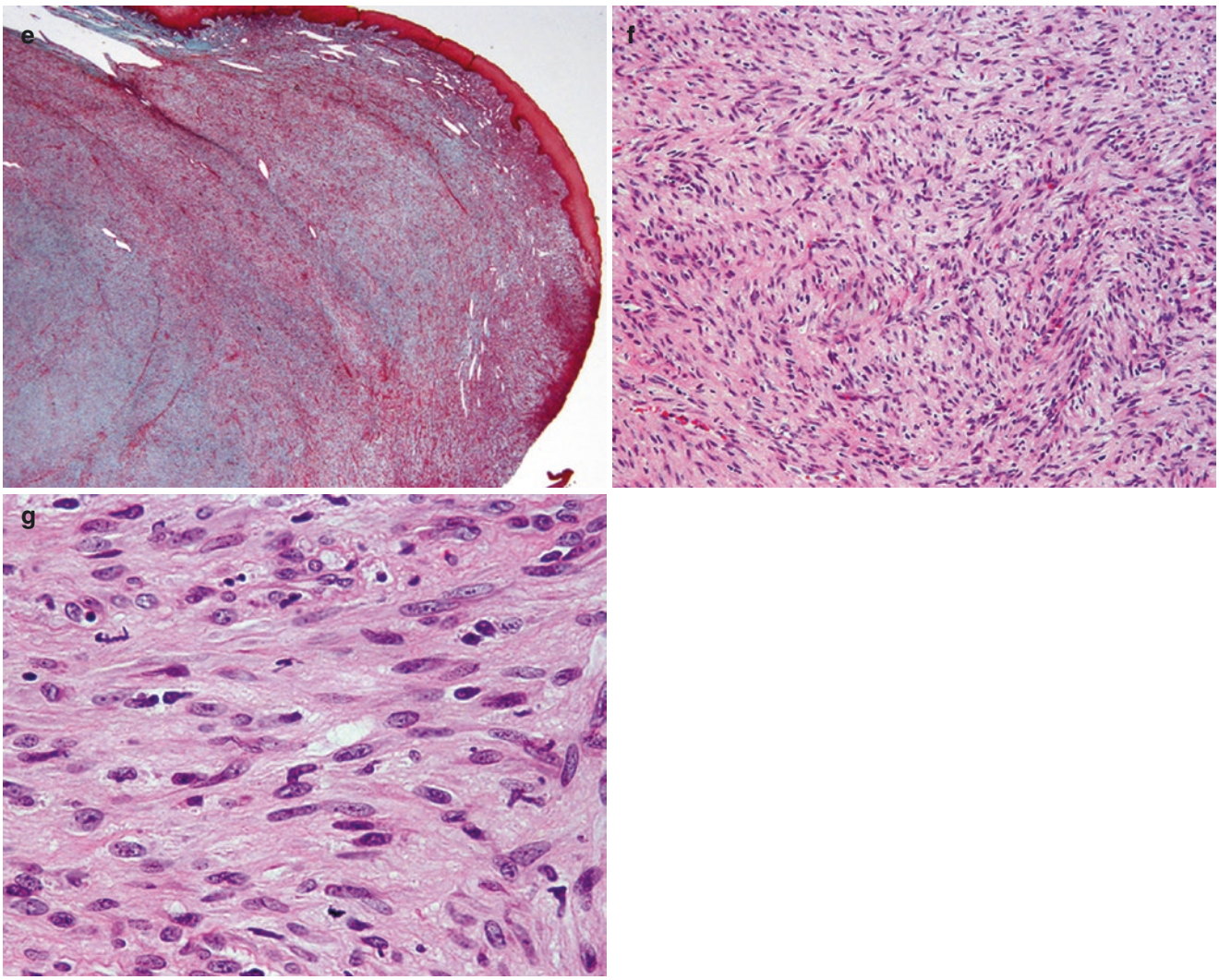

Fig. 12.14 (continued)

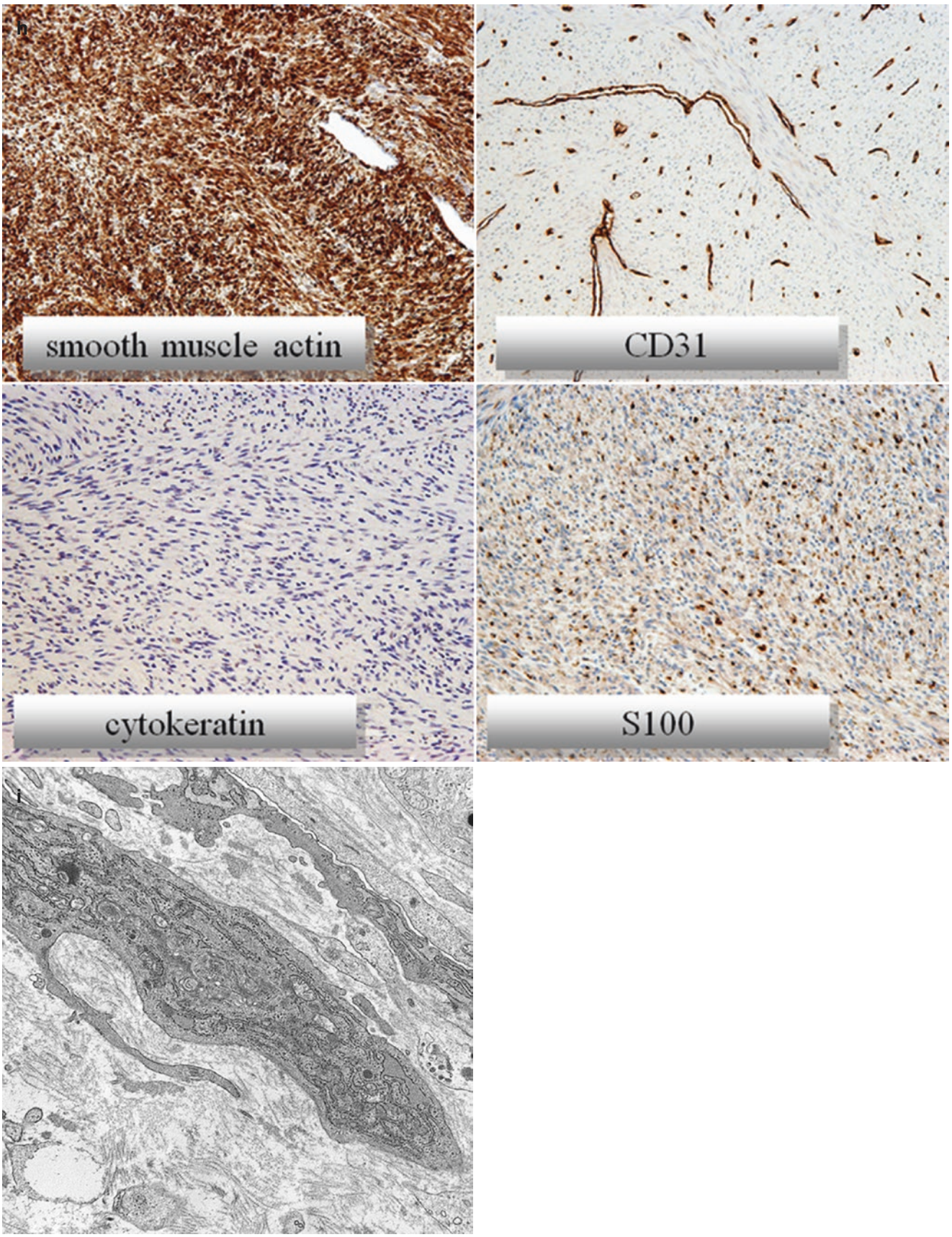

Fig. 12.14 (continued)

lucent expansile mass in the left ramus of the mandible. The lesion was excised through a transoral approach. The loose dentition was removed and the bony cavity drilled out. Final pathology revealed a myofibroma (Fig. 12.14c–i).

Case Presentation 2

An 8-year-old female presented with a 2-month history of an enlarging right mandibular mass. There was an obvious mass visible intra-orally (Fig. 12.15a). Imaging revealed a large mandibular lesion that was displacing a molar and had a smooth lytic appearance in the involved mandible (Fig. 12.15b–c). An open biopsy was performed which was consistent with a benign-appearing neoplasm. The lesion was removed through an intra-oral approach. The affected dentition was removed and the tooth socket and remaining bone polished with drill burrs (Fig. 12.15d). Final pathology showed chondromyxoid fibroma (Fig. 12.15e–l).

Case Presentation 3

A 4-year-old female presented with a right cheek swelling after a fall. A CT scan (Fig. 12.16a) revealed an expansile osteolytic lesion involving the right maxilla. The lesion was excised via a Caldwell-Luc approach. Pathology revealed a juvenile ossifying fibroma with active or aggressive features (Fig. 12.16b–f).

Fibromatosis of Mandible (see also Chap. 26)

Definition

Fibromatosis is a fibrous neoplasm that arises from a musculoaponeurotic structure. Desmoid tumors, grade I fibrosarcoma, aggressive fibromatosis, and fibromatosis of soft tissues are all terms referring to the same benign histological entity [44]. Despite the benign histological appearance and the fact that it is not metastatic its biological nature is locally aggressive and it tends to recur. The pathogenesis is not known and it is speculated that genetic factor, endocrine system effect, and physical trauma may play a role [45]. Fibromatosis of the head and neck constitutes 12–15% of all fibromatosis cases, predominantly in the mandible and tongue.

Clinical Presentation

Most cases present with a firm, non-mobile 1–15 centimeters swelling fixed to the underlying tissue and not affecting the overlying skin.[45] The tumor tends to expand quickly and it is usually not painful. Functional impairment may occur infrequently [46].

Differential Diagnosis

- Albright's syndrome
- Cherubism
- Desmoplastic fibroma
- Neurofibroma
- Proliferative myositis
- Fibrosarcoma [46]

Radiological Features

Radiological findings may vary. Infiltration of soft tissue and bone boundaries are seen on CT-scan and MRI and no radiological feature can help in diagnosis but are crucial before surgical intervention.

Management

Surgery is the treatment of choice. Extensive lesions may require mandibulectomy. Adjuvant radiotherapy and chemotherapy may apply in certain cases when excision is difficult or in a recurrent tumor [44].

Aneurysmal Bone Cyst (ABC)

Definition

Aneurysmal bone cysts are benign, blood-filled, cystic osseous tumors, which can be rapidly expanding and locally destructive. They typically involve the long tubular bones and spine. Approximately 3% of these tumors are found in the head and neck region with the mandible being the most common site [47]. The lesion is considered to be neoplastic and generally arises in isolation [48]. About 30% of ABCs considered to be secondary to another bone lesion [47]. There is a female predominance and most patients are younger than 20 years, the tumor tends to be more aggressive in older age [49].

Clinical Presentation

ABCs in the head and neck typically present with a rapid onset of pain and swelling dependent on its location. Headache, diplopia, proptosis, tooth loosening, facial nerve palsy, and lip paresthesias may occur [49].

Differential Diagnosis

- Giant cell tumor
- Myxoma
- Unicameral bone cyst
- Fibroma (ossifying, non-ossifying)
- Reparative giant cell granuloma of hyperparathyroidism
- Enchondroma
- Fibrous dysplasia
- Osteosarcoma
- Bone fracture
- Osteomyelitis

Radiological Features

In panoramic radiography of the jaws, ABC will appear as an eccentric, ballooned cystic expansion surrounded by a rim of sclerosis [49]. Most cysts will have a radiolucent appearance (87%).

CT-scan shows a cystic mass with bone destruction and stromal cavities enhanced with intravenous contrast. MRI will show the cystic content at T1- and T2-weighted images with internal fluid-fluid levels. A persistent venous circula-

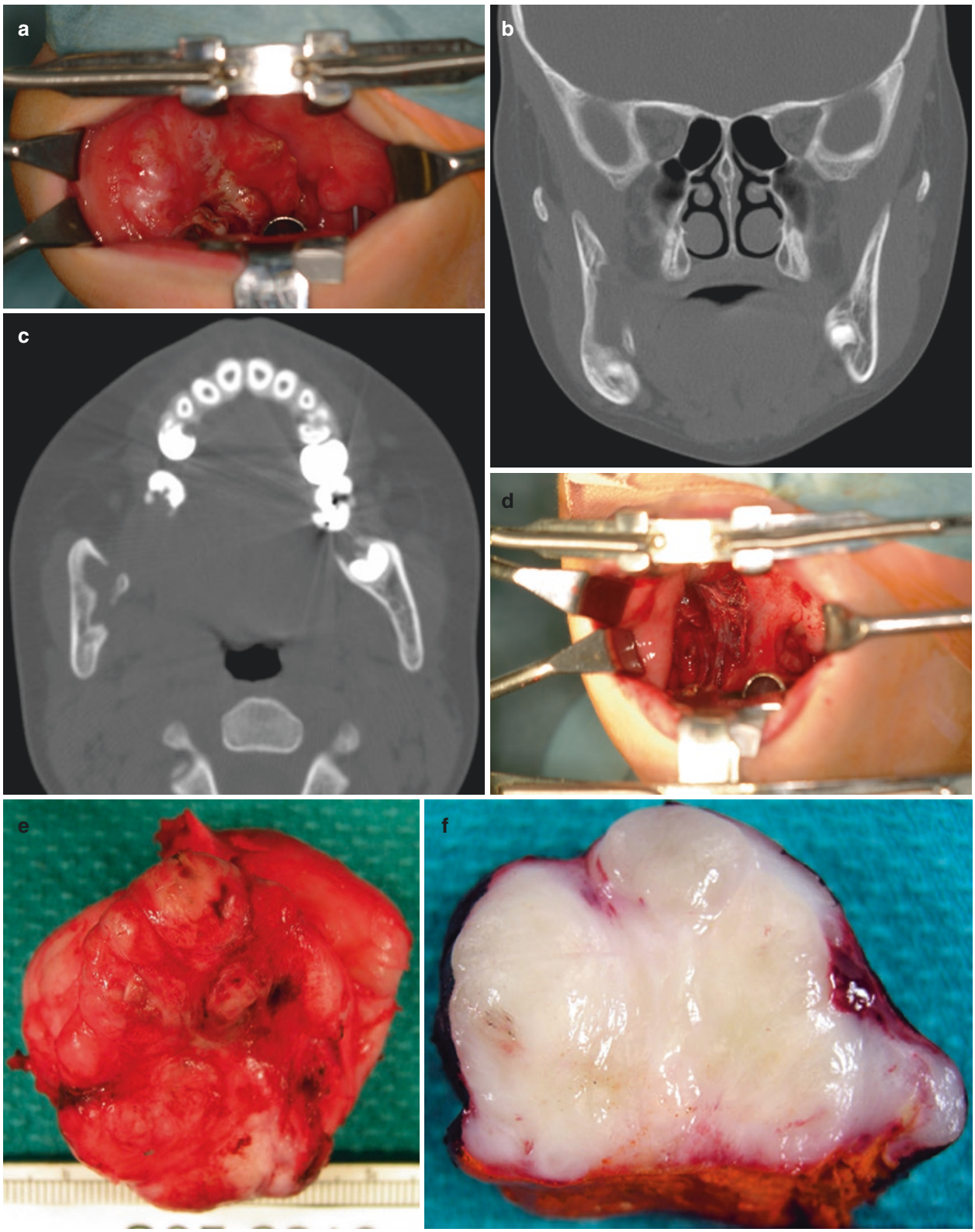

Fig. 12.15 (**a**) Large mass seen intraorally. (**b**) Coronal CT showing smooth lytic changes in the right ramus of the mandible suggestive of benign pathology. (**c**) Axial CT showing large mass displacing a molar with smooth lytic changes in the mandible. (**d**) Surgical bed after removal of the tumor and polishing all bony surfaces with a drill burr. (**e**) Gross appearance of tumor. (**f**) Cut surface revealed a pale gray glistening fleshy stroma with gelatinous consistency. Patchy areas had pale yellow discolorations. (**g**) H&E. Within the hypocellular areas the tumor cells have a myxoma-like appearance. (**h**) H&E. There were distinct myxoma-like areas (hypocellular and pale staining) which could be mistaken as tumor degeneration. More intensely stained eosinophilic areas (right upper corner) represented the fibrotic domains. (**i**) H&E. Numerous short plump spindle cells with minimal cytological atypia were present within an abundant fibrous/fibrillar matrix. (**j**) Immunostain showed expression of vimentin. (**k**) Immunostain for S100 was negative. (**l**) Immunostain for CD34 showed weak reactivity

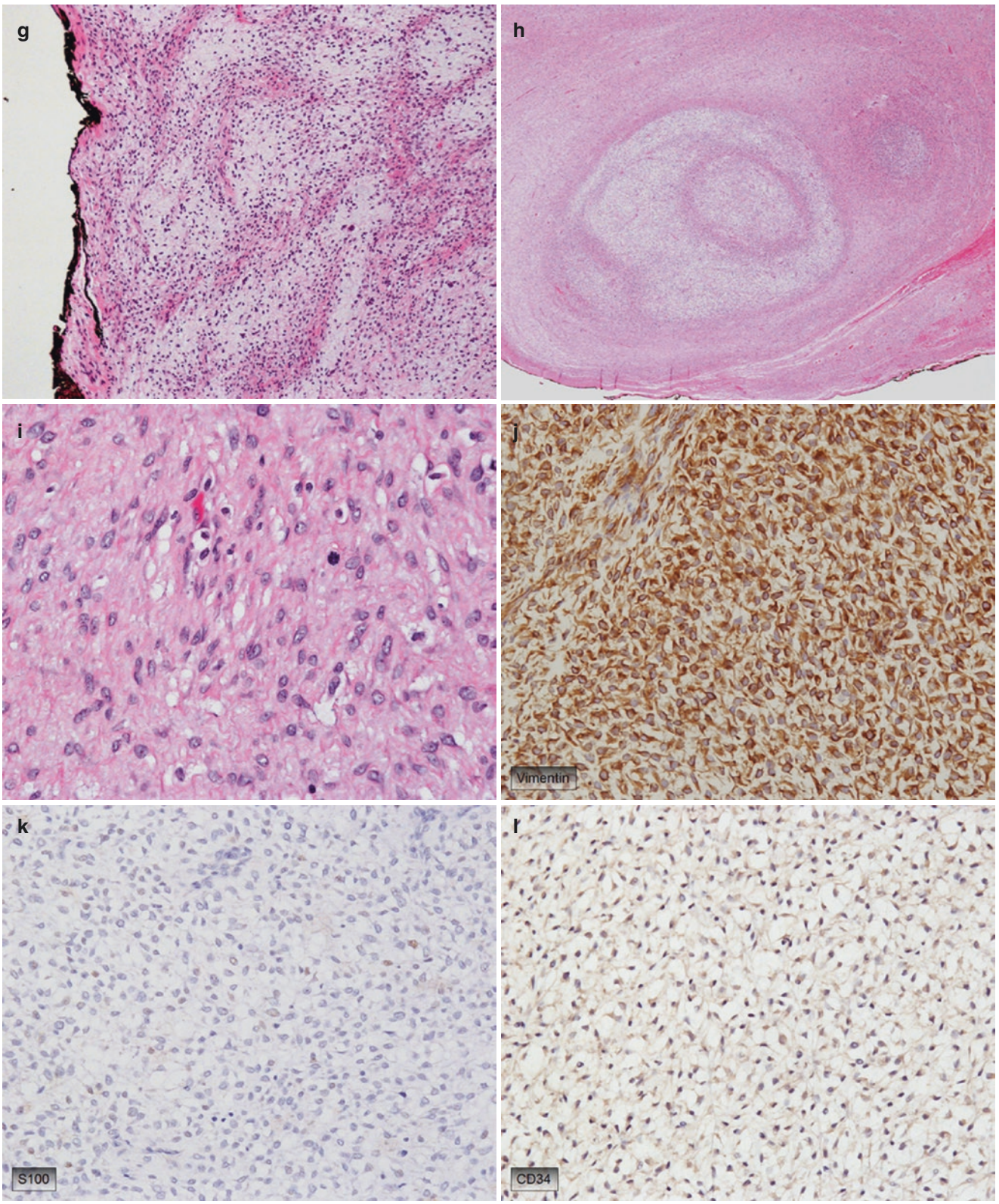

Fig. 12.15 (continued)

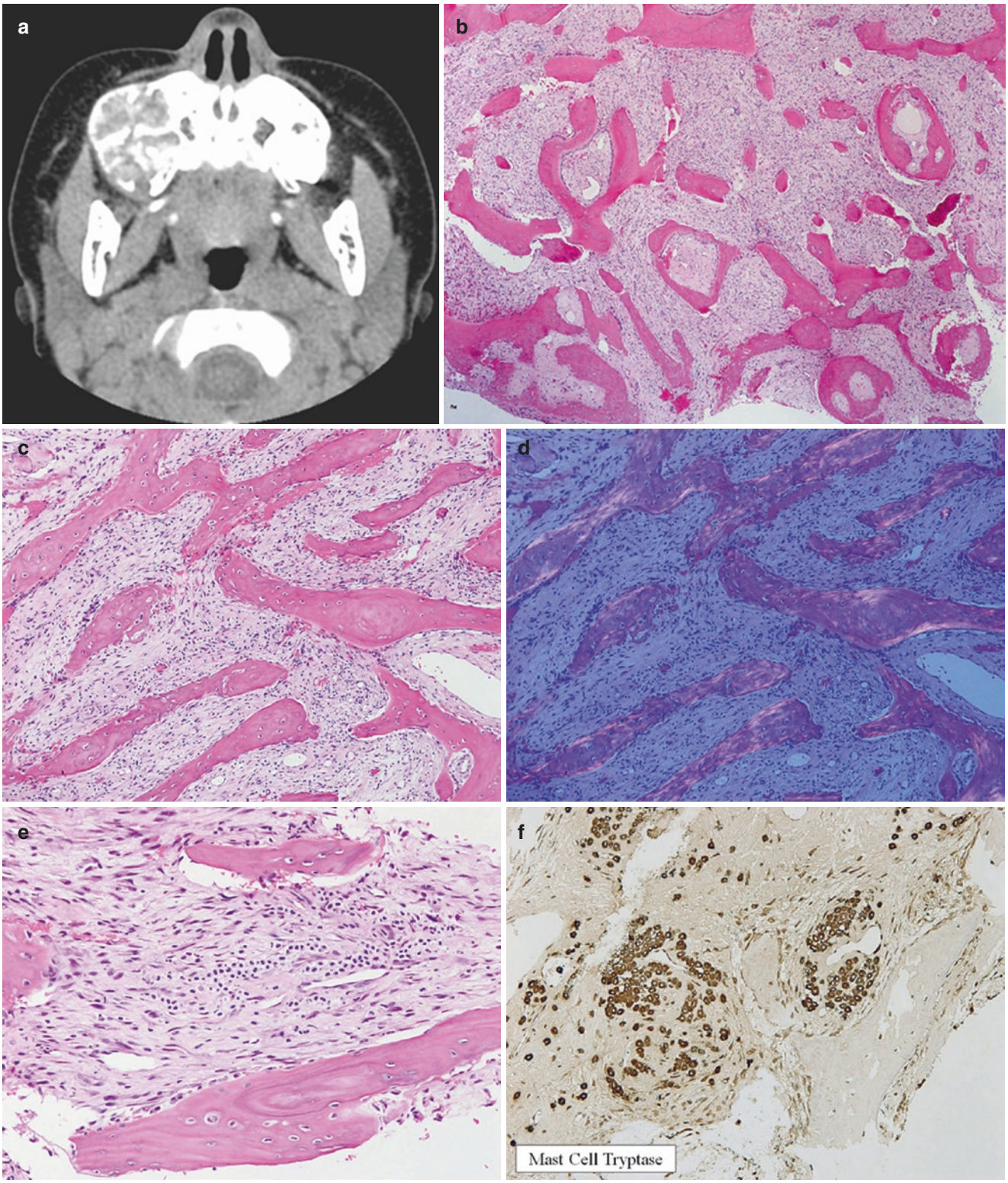

Fig. 12.16 (**a**) Axial CT showing expansile mass in left maxilla. (**b**) H&E. presence of haphazard arranged thin misshapen bone trabeculae with focal osteoblastic rimming. (**c**) H&E. Presence of a spindle cellular stroma in the marrow space. These cells have mild pleomorphism with hyperchromatic nuclei. (**d**) H&E under polarized light illumination: Mostly woven bone was present. (**e**) Small clusters of mononuclear cells within this mildly hypercellular marrow stroma. (**f**) Immunostain showed the mononuclear cells are mast cells

tion will be demonstrated on angiography with occasional arterial-venous shunting [50].

Management

En-bloc resection of the cyst remains the most effective treatment with the lowest rate of recurrence. However, other methods such as selective arterial embolization, sclerosing agent injection, and radiation have been utilized as well.

Clinical Example

A 16-year-old female presented with a left zygoma mass. The mass was tender and fluctuated in size over the past year. There was no history of trauma to the area. CT showed multicystic expansion of the left maxilla consisted with an aneurysmal bone cyst (Fig. 12.17a–b). The lesion was removed via a sublabial approach. The pathology confirmed a suspected diagnosis of aneurysmal bone cyst (Fig. 12.17c–e).

Melanotic Neuroectodermal Tumor of Infancy (See also Chap. 6)

Definition

Melanotic neuroectodermal tumor of infancy is a rare pigmented tumor arising most frequently in the maxilla (68%) during the first year of life. It is believed to originate in the neural crest cells and the pigmented epithelium resembles the retinal-pigmented epithelium. These facts led to different theories regarding the origin of the tumor and various names were suggested along the years [51–53]. The tumor appears more frequently in female babies [53] and is quickly growing and in some cases is associated with high level of urinary vanillylmandelic acid [51] reflecting on the possibility of a neuroectodermal origin.

Clinical Presentation

The tumor is apparent in the first ear of life, usually at 6–12 months, as a bluish lump bulging under the upper lip or cheek with a loss of the maleo-labial fold.

The lesion is not tender or pulsating but can cause feeding disturbances, no breathing difficulties were reported. The overlying mucosa or skin is usually intact.

Differential Diagnosis

- Ewing sarcoma
- Neuroblastoma
- Peripheral neuroectodermal tumor
- Askin tumor
- Rhabdomyosarcoma
- Lymphoma

Radiological Features

Either on plain radiography, CT-scan, or MRI a large osteolytic mass is shown but none has a diagnostic ability.

Management

The mainstay of managing this tumor is surgical excision. A substantial recurrence rate of 15–20% was reported; however, one should take into account future consequences of future facial growth deformity [53].

Odontogenic Benign Tumors (See Also Chap. 13)

Odontogenic Cysts

Definition

Odontogenic cysts are cysts that arise from odontogenic epithelium and occur in the tooth-bearing regions of the jaws [54]. These cysts are common in the pediatric age group and many are non-symptomatic and found incidentally on radiographs; however, they can lead to bone destruction and teeth displacement [55]. Most of the pediatric odontogenic cysts are developmental in origin and few are inflammatory [54, 56].

Dentigerous Cyst

A dentigerous cyst is a developmental cyst that arises from a crown of an unerupted tooth, usually from the third molars or maxillary canines. The cyst is commonly encountered in the second decade of life.

Eruption Cyst

An eruption cyst is a developmental cyst as well originating in a separation of dental follicle during eruption, usually from the incisors or first molars during the first decade of life.

Periapical Cyst

A periapical cyst is an inflammatory cyst origination from dental pulp necrosis and can be associated with a non-vital tooth, it may occur at any age and in any tooth-bearing site [38].

Odontogenic Keratocyst

Odontogenic keratocyst is termed as a keratocystic tumor characterized by a unicystic or multicystic, intra-osseous growth of odontogenic origin with a parakeratinized squamous epithelial lining, usually 4–8 cell layers in thickness and palisaded basal cell layer [57]. It has an aggressive clinical behavior and may originate in either the maxilla or mandible (involved in 60% of pediatric cases) with a predilection for the posterior body and ascending ramus [38]. Odontogenic keratocysts are commonly discovered at the second decade of life. Children with multiple or recurrent keratocysts should be investigated for nevoid basal cell carcinoma syndrome (Gorlin syndrome) which is an autosomal dominant syndrome caused by mutations in the Patched (PTCH) tumor suppressor gene [58].

Clinical Presentation

Many of the cysts are non-symptomatic and may be encountered incidentally at the dentist's office or in dental radio-

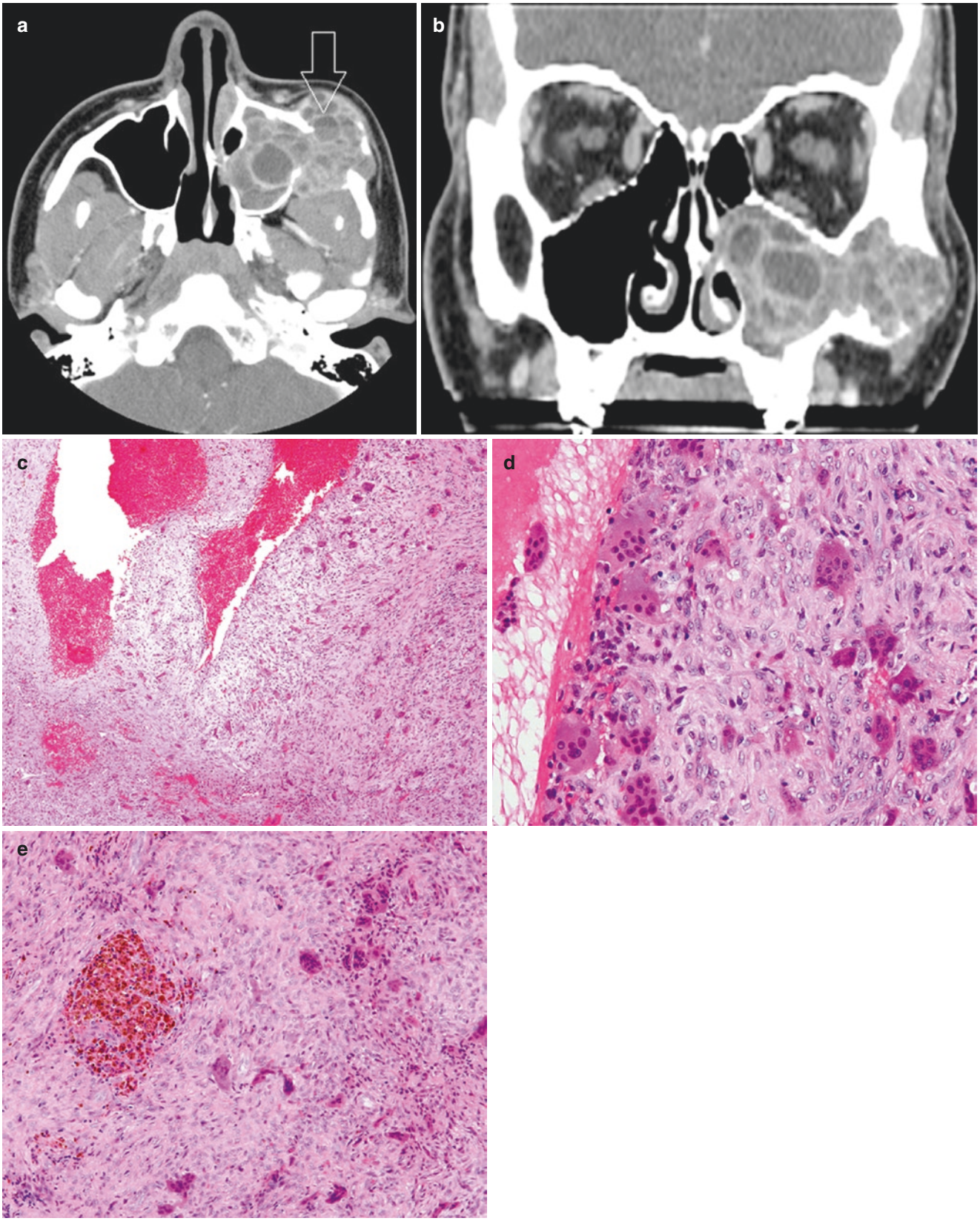

Fig. 12.17 (**a**) Axial CT showing multicystic expansion of the left maxilla. Fluid-fluid levels can be seen (arrow) suggesting recent hemorrhage most likely responsible for increase in size and pain. (**b**) Coronal CT of ABC of the maxilla. (**c**) H&E. Fleshy membranous tissue in the cyst wall often abutted onto the bone interface. These tissues had a fibrotic granulation tissue-like appearance. Sprinkling of osteoclastic giant cells within these tissues was often found. (**d**) H&E. Infiltrates of osteoclasts within the fibrous cyst wall tissue. (**e**) H&E. Collections of hemosiderin containing histiocytes were present

graph. Odontogenic keratocyst can grow to be extremely large and cause pain, swelling, and drainage.

Differential Diagnosis
- Odontogenic benign neoplasm
- Odontogenic malignant neoplasm

Radiological Features
Plain radiography may be sufficient for the initial workup of cysts limited to the jaws; however, extension into adjacent anatomical structures such as the nasal cavity, sinuses, orbits, and pterygopalatine fossa will require a CT-scan or MRI for a precise anatomical evaluation prior to surgical intervention.

Management
The mainstay of managing those cysts is surgical intervention either by marsupialization, curettage, or enucleation.

Clinical Examples
Case Presentation 1 (Fig. 12.18)
Case Presentation 2 (Fig. 12.19)
Case Presentation 3 (Fig. 12.20)
Case Presentation 4
This 12-year-old male presented with a 2-week history of left facial swelling and left nasal obstruction. He had a left dental extraction 7 weeks earlier. A CT scan revealed a large left maxillary cyst that was excised via a sublabial approach (Fig. 12.21).

Cemento Ossifying Fibroma

Definition
Cemento ossifying fibroma is a benign fibro-osseous neoplasm of the jaws, affecting either the mandible or the maxilla, it is slowly progressing and could be unilocular to multilocular [59]. There is a controversy with respect to its origin, bony, or odontogenic. The tumor may contain cement-like calcifications along with bony material [60].

Clinical Presentation
Most patients would be non-symptomatic with lesions present as a slow, painless, expanding swelling, usually more of the mandible than of the maxilla [61].

Differential Diagnosis
- Fibrous dysplasia
- Cemento osseous dysplasia
- Ossifying fibroma
- Gigantiform Cementoma

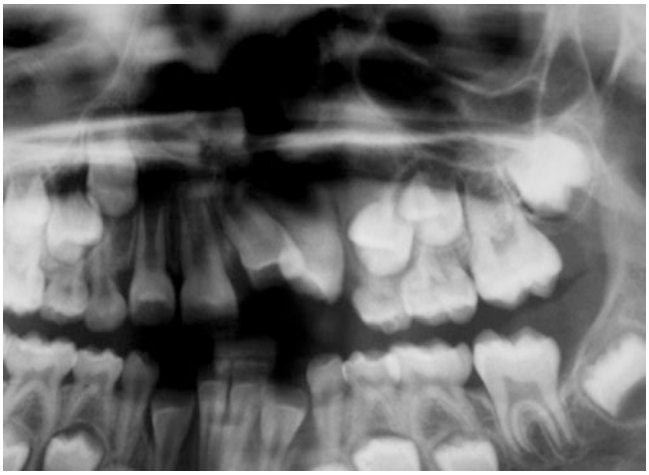

Fig. 12.18 This panorex of a 6-year-old female with a dentigerous cyst involving the anterior maxilla superior to tooth 2.2

Radiological Features
The tumor presents as corticated radiolucency with variable radio-opacities arising in the alveolar process. The ramus or the inferior cortex of the mandible may be involved in case of a large lesion.

Management
Curettage and enucleation.

Clinical Example
Clinical Case
A 13-year-old male presented with a stable right maxillary swelling present for several years. Prior imaging was consistent with a diagnosis of fibrous dysplasia (Fig. 12.22a–b). However, there was a recent increase in size which prompted the surgical resection via a Caldwell-Luc approach. Pathology was consistent with cemento ossifying fibroma (Fig. 12.22c–f).

Malignant Tumors

Malignancies of the oral cavity are very rare in the pediatric age group. Soft tissue malignancies such as lymphoma and rhabdomyosarcoma are discussed in more detail elsewhere in this book. Mucosal malignancies such as squamous cell carcinoma or even melanoma can occur in the oral cavity of children and should be included in the differential diagnosis of unusual erosive or pigmented lesions, especially in the immunocompromised or post solid organ transplanted patient. This section will focus on odontogenic and bone malignancies.

Odontogenic Malignancies

Definition

Malignant odontogenic tumors are rare. Malignant transformation from a benign tumor is rare as well and may occur in less than 1%. Malignant ameloblastoma is such a tumor that originates in a recurrent ameloblastoma and may metastasize primarily to the lungs. Metastases may occur as far as to 20 years since the primary tumor was initially diagnosed. The prognosis is poor and the tumor is usually encountered during adolescence [62].

Another odontogenic malignancy is ameloblastic fibrosarcoma. The tumor usually involves the mandible and originated in ameloblastic fibroma or ameloblastic odontoma [38]. It is locally destructive and metastases are uncommon.

Clinical Presentation

A rapidly progressive swelling and pain of the involved jaw are most commonly encountered.

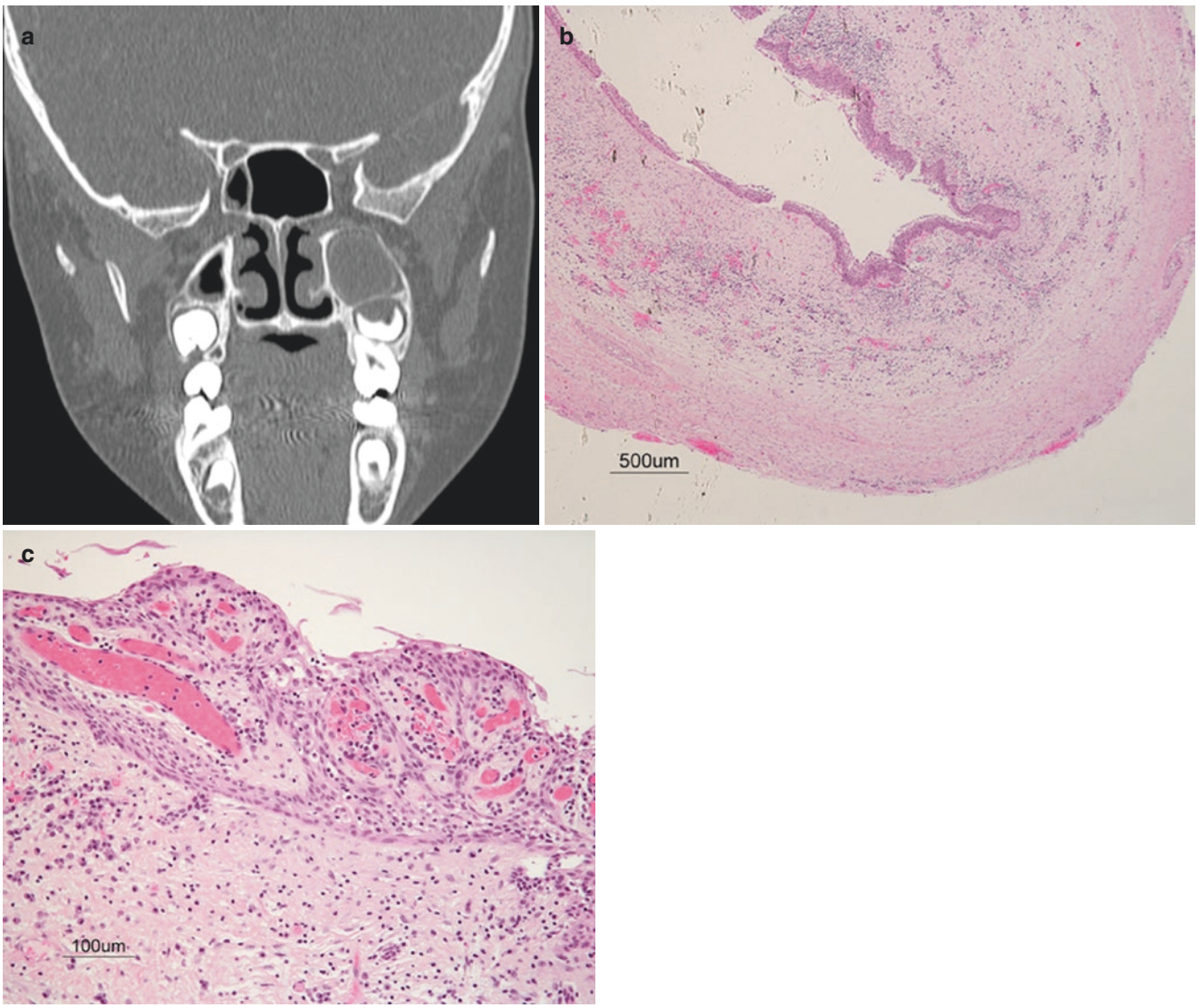

Fig. 12.19 (**a**) The CT scan is of an 8-year-old male with a 9-month history of a left malar mass and left-sided nasal obstruction. The lesion was excised through a Caldwell-Luc approach. (**b**) H&E. Fibrous tissue cyst wall with preserved stratified squamous epithelial lining cells. (**c**) H&E: Higher magnification showing squamous epithelium. As distinguished from odontogenic keratocyst, the basal cell nuclei of dentigerous cysts are flat not tall, and the nuclear long axis is not perpendicular to the basement membrane. (**d**) H&E. Several residual dentigerous rests are found adjacent to the cyst

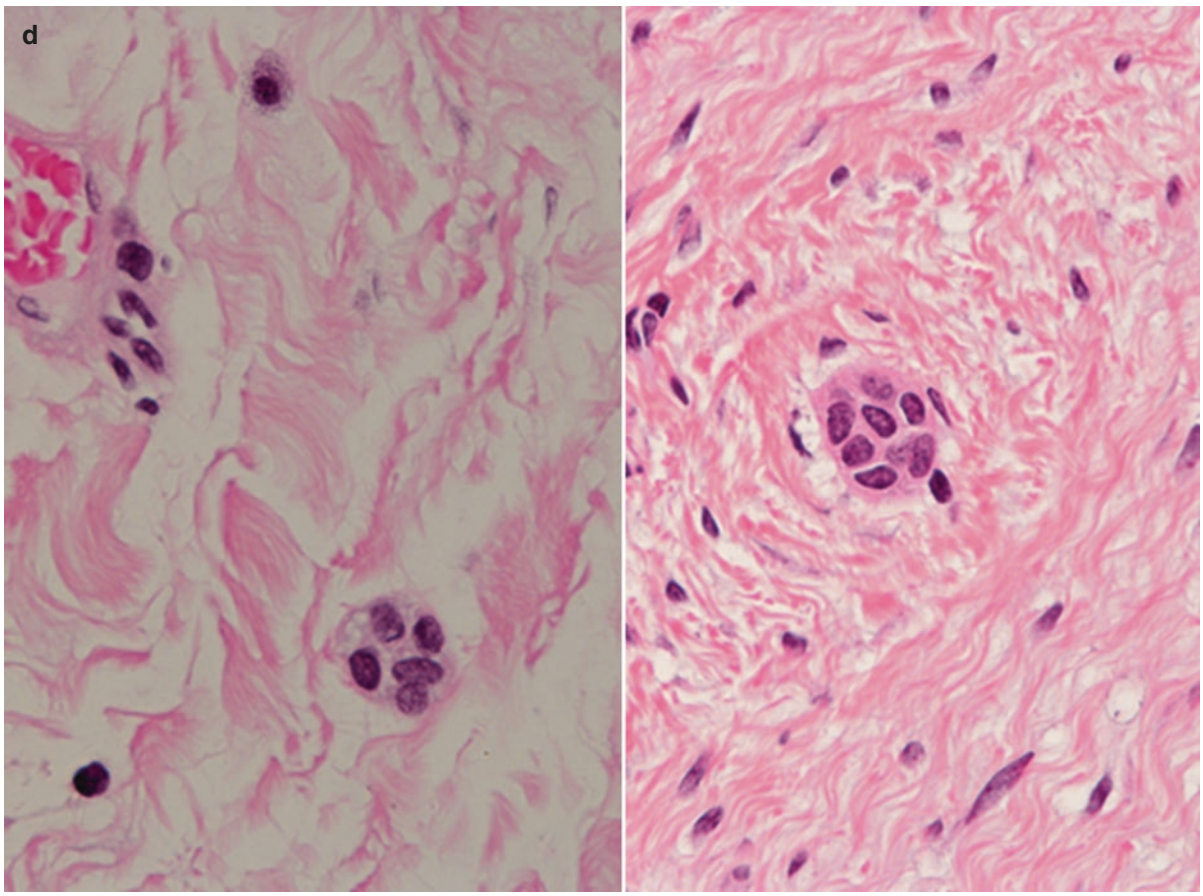

Fig. 12.19 (continued)

Differential Diagnosis
Any locally aggressive benign or malignant neoplasm of the jaws, either odontogenic or non-odontogenic.

Radiological Features
Malignant odontogenic tumor may appear as a radiolucent lesion with ill-defined margins.

Management
The treatment of choice is a wide local excision but radiotherapy and chemotherapy are used as well after incomplete excision [63].

Bone Malignancies

Osteosarcoma

Definition
Osteosarcoma is the most common primary malignancy of bone in children; however, its appearance in the jaws is uncommon in comparison to long bones [64]. Osteosarcoma may appear de novo or in a previously irradiated site with an incidence of 0.03–0.8% [65, 66].

Clinical Presentation
Osteosarcoma, like other neoplasms of the jaws, will appear as localized painful swelling, occasionally with paresthesias, teeth loosening, and ocular involvement.

Differential Diagnosis
Odontogenic or non-odontogenic aggressive bone neoplasms.

Radiological Features
The radiographic appearance would be as radiopaque or radiolucent lesion with ill-defined borders. A classical sunburst appearance is dependent on the amount of osteophytes in the bone; however, it is seen infrequently [38].

Management
Wide surgical excision with free tissue transfer combined with neoadjuvant therapy offer best chances for cure.

Ewing Sarcoma (See also Chap. 27)

Definition
Ewing sarcoma is the second most common malignant bone neoplasm in the pediatric age group; its occurrence in the jaws is very rare and it is most frequently seen in long

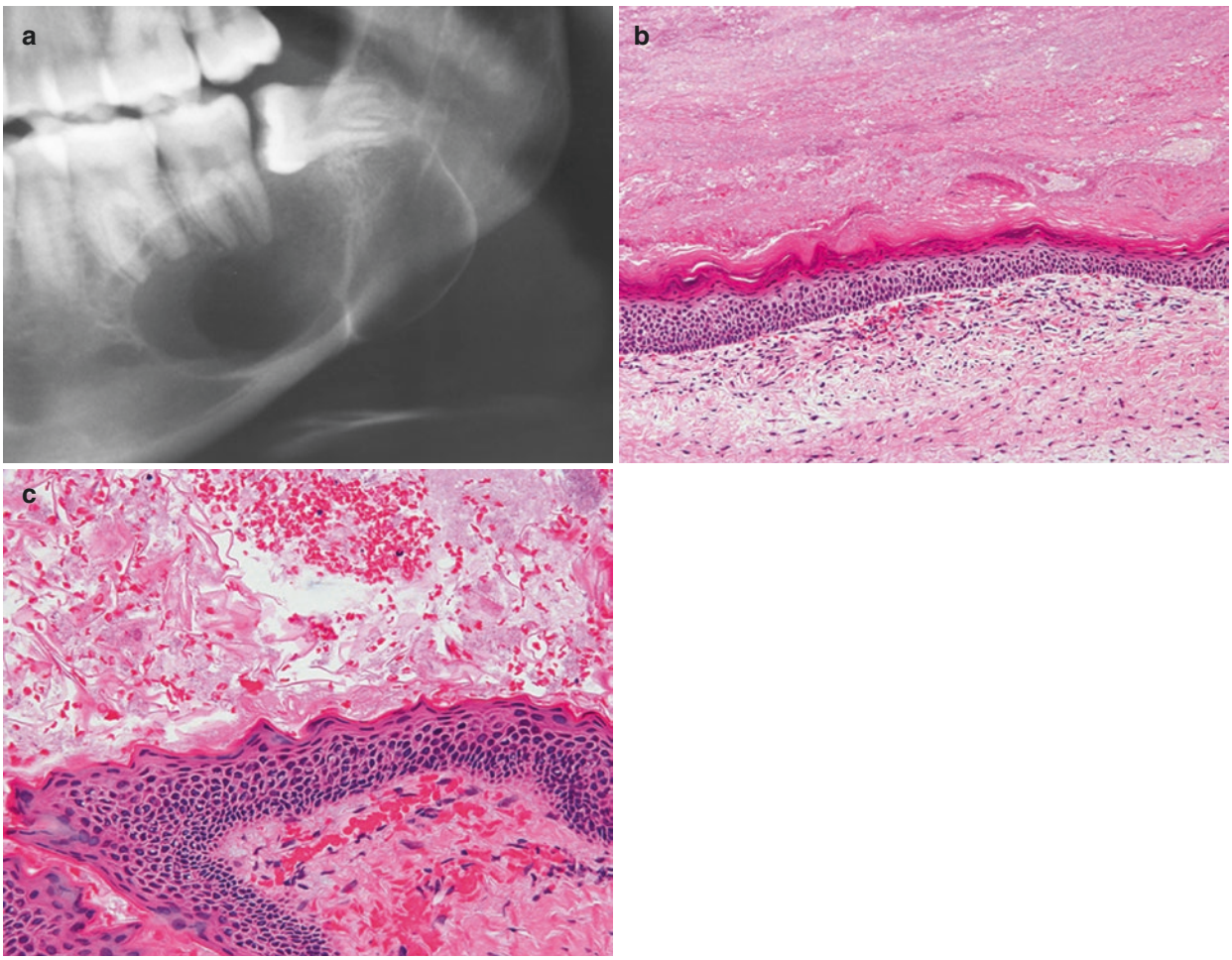

Fig. 12.20 (**a**) This is a panorex of a 16-year-old female with a slowly expansile lesion of the left mandible. This was treated with a partial mandibulectomy and extraction of several teeth. The pathology indicated that this was an odontogenic keratocyst. (**b**) H&E. The squamous epithelium here was several cell layers thick and the nuclei in the basal cell layer stood perpendicular to the basement membrane. (**c**) Numerous desquamated keratinocytes are seen within the cyst cavity

bones. The mandibular ramus is involved more frequently than other sites. In 85–90% of cases the tumor cells show a translocation between chromosomes 11 and 22 [t(11;22)(q24;q12)] [67].

Clinical Presentation

The tumor usually appears as a local painful swelling.

Differential Diagnosis

Other small blue cell tumors:

- Metastatic neuroblastoma
- Osteosarcoma
- Rhabdomyosarcoma

Radiological Features

The lesion appears with irregular bone destruction with ill-defined margins. The onion-skin typical appearance is common in long bones and unusually seen in the jaws.

Management

Wide surgical excision combined with neoadjuvant therapy.

Clinical Example

Clinical Case

A 13-year-old female presented with a 9-week history of dental pain, loose right upper maxillary dentition, and an enlarging right maxillary mass. The CT scan demonstrated a destructive right maxillary mass with invasion of the orbital floor and sinus walls. A transnasal biopsy was performed as the mass occupied the nasal cavity. Pathology was consistent with a diagnosis of Ewing sarcoma, with t(11;22)(q24;q12) EWS-FLI1 positive (Fig. 12.23a–f).

Burkitt Lymphoma (See Also Chap. 25)

Definition

Lymphomas of the head and neck are primarily seen involving cervical lymph nodes, however the endemic form of Burkitt lymphoma characteristically affects the facial skele-

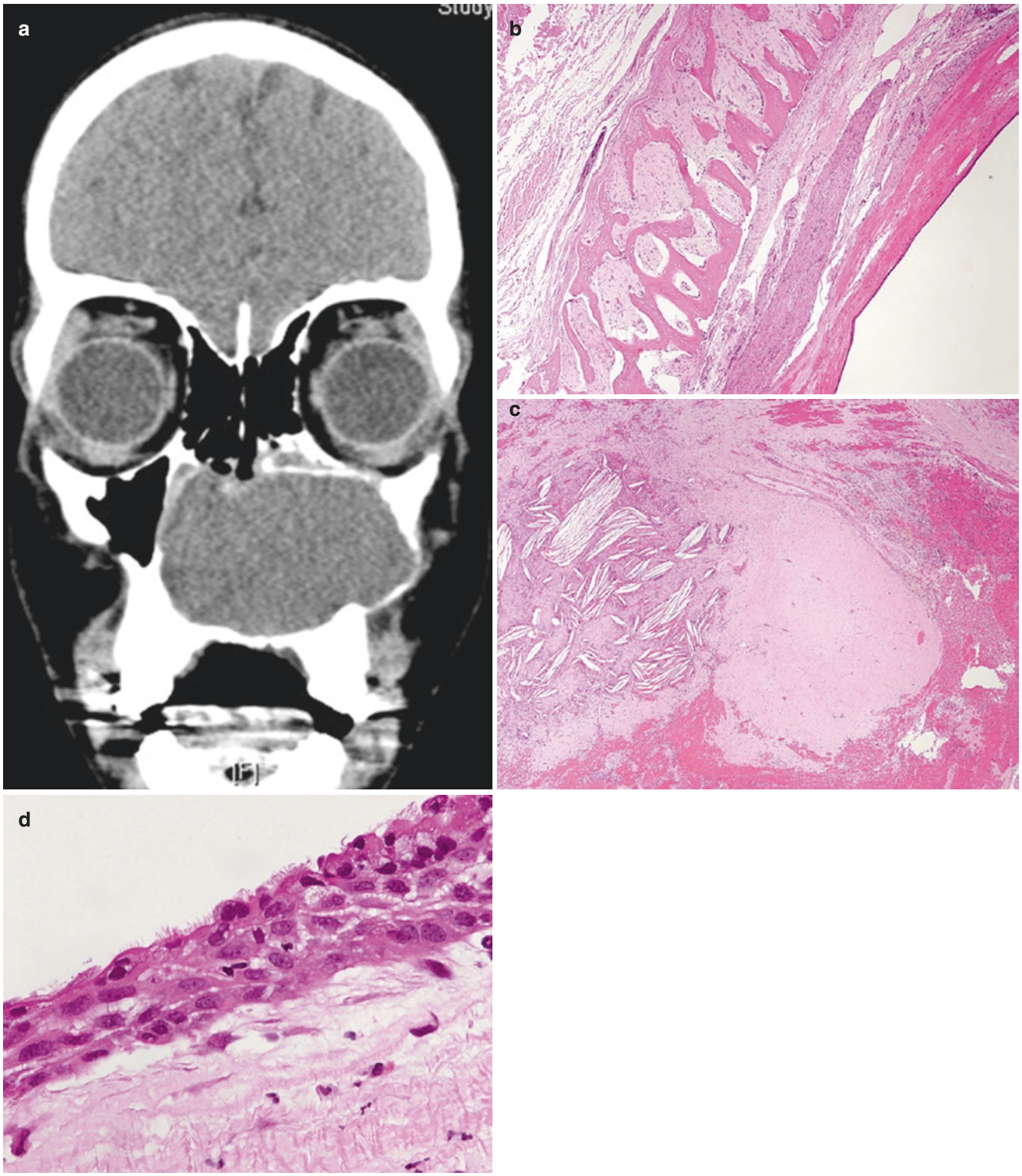

Fig. 12.21 (**a**) Coronal CT shows a large left expansile maxillary cyst thinning both medial and lateral walls. The mass has displaced the nasal septum to completely fill the nasal cavity. (**b**) H&E: Cyst wall was lined by a single layer of epithelial cells. There was a retention of a fibrotic mucosa. The sinus bone was non-reactive and normal. (**c**) H&E. Portion of the cyst contains degenerated cell membrane lipids with collections of cholesterol crystals. (**d**) H&E. Remnants of ciliated epithelial cells of respiratory type epithelium were present

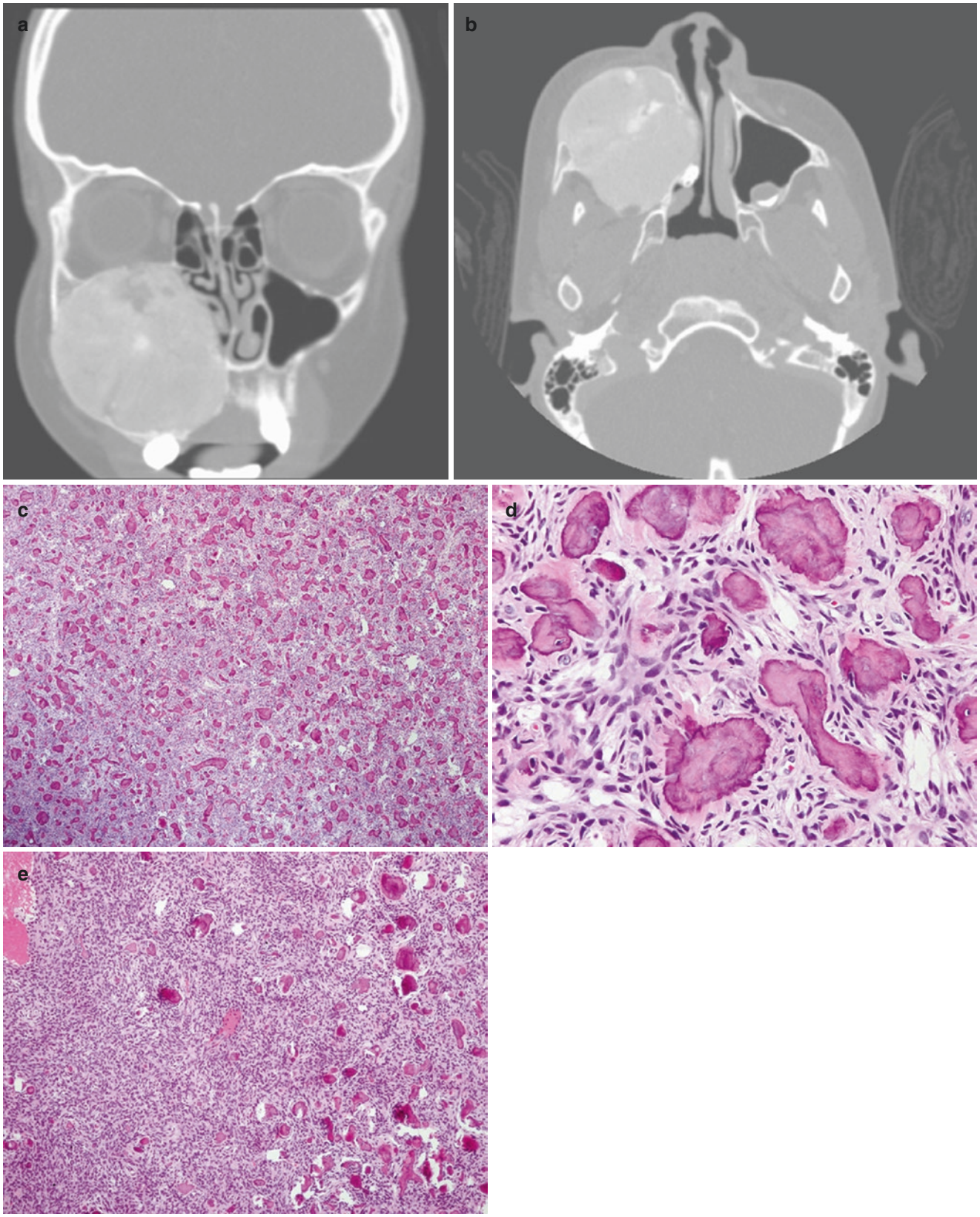

Fig. 12.22 (**a**) Coronal CT shows an expansile fibro-osseous mass of the maxilla. Note smooth margins of the tumor and displacement of canine tooth. (**b**) Axial CT of expansile fibro-osseous mass of the maxilla with deformity of the right hemi face. (**c**) H&E. Cementum-like tissues dominated the intertrabecular spaces. (**d**) H&E higher magnification of Fig. 12.22c. Detailed appearances of the cementum bodies within a bland spindle cells stroma. (**e**) H&E. A hypercellular area is present in the marrow space

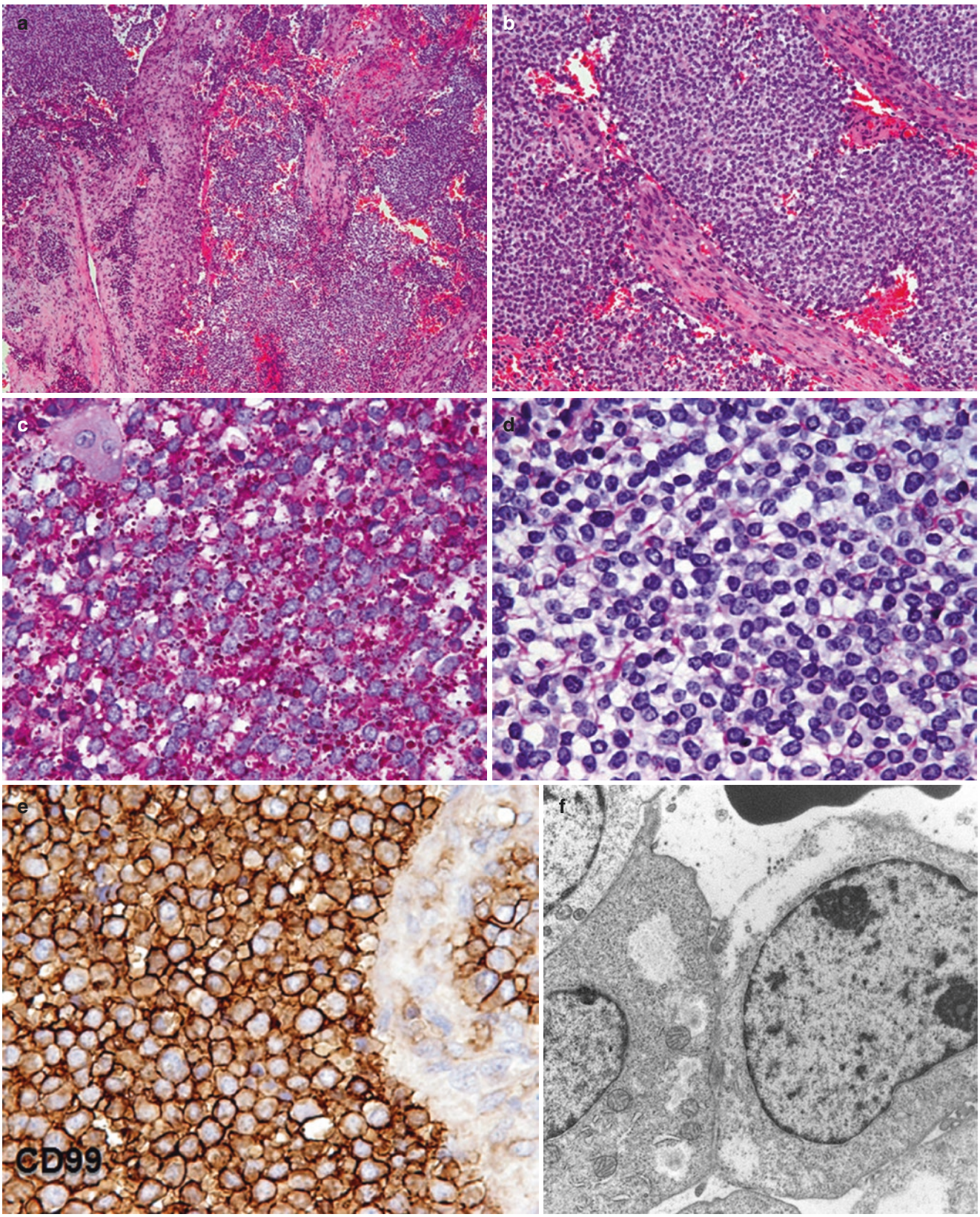

Fig. 12.23 (**a**) H&E. Characteristic tumor infiltrates which had a small round blue cell appearance. (**b**) H&E. Al hypercellular infiltrate where all cells had a spherical hyperchromatic nucleus. (**c**) PAS stain demonstrating the abundant glycogen within the cytoplasm of these tumor cells. (**d**) H&E. Predominant component shown here are the so-called pale cells with a few dark cells (in the left upper corner). This pattern is sometimes referred to a salt and pepper pattern. (**e**) Immunostain for CD99 showed strong membranous staining. (**f**) Electron micrograph showed round cells with coiled string like aggregates within the nucleoli and an abundance of glycogen granules in the cytoplasm. Between the two tumor cells only primitive cell junctional complexes are present

ton. This form of lymphoma is typically encountered in eastern Africa countries in children aged 2 to 9 years [68]. Nearly all-endemic cases are associated with Epstein–Barr virus, which is hypothesized to deregulate the c-MYC proto-oncogene, leading to chromosomal translocation [38]. Boys are affected almost twice as girls and the mandible is most frequently involved site [68].

Clinical Presentation
The tumor usually appears as a local painful swelling.

Differential Diagnosis
Odontogenic of non-odontogenic aggressive bone neoplasm.

Radiological Features
Signs of generalized destruction of tooth crypts and diffuse disruption of jaw trabeculation. Loosening teeth [69].

Management
Chemotherapy.

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