# 6 Clinical and Surgical Management of Pediatric Diseases of the Nasal Cavity, Paranasal Sinus, and Skull Base

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This chapter covers common and rare pathologic entities that are present in the nasal cavity and sinuses of children, from infancy to adolescence. The pathological entities discussed are summarized in Table 6.1, together with the typical age and location.

**Table 6.1** Pediatric nasal pathological diagnoses, anatomic locations, and typical age of presentation

| Diagnosis | Typical location | Typical childhood age at presentation |
|---|---|---|
| Nasal dermoid cyst | Midline nasal dorsum, deep to nasal bones extending up to anterior skull base | Infant |
| Meningoencephalocele | Unilateral, intranasal | Infant |
| Nasal glioma | Unilateral, intranasal | Infant |
| Antrochoanal polyp | Unilateral, maxillary sinus, intranasal and choana | Older child or adolescent |
| Inverted papilloma | Unilateral, ethmoid sinus, and intranasal | Older child or adolescent |
| Allergic fungal sinusitis | Bilateral, intranasal, and sinuses | Older child or adolescent |
| Invasive fungal sinusitis | Bilateral, intranasal, and sinuses | Older child or adolescent |
| Pyogenic granuloma septum | Unilateral, nasal septum | Not specific |
| Fibrous dysplasia | Maxilla or other craniofacial bones | Not specific |
| Melanotic neuroectodermal tumor of infancy | Anterior maxillary alveolar ridge | Young infant |
| Nasal xanthogranuloma | Cutaneous nasal | Not specific |
| Inflammatory polyps | Bilateral, associated conditions such as cystic fibrosis, dyskinetic cilia (PCD—Primary Ciliary Dyskinesia), chronic sinusitis | Older child or adolescent |

## Nasal Dermoid Cyst

### Definition

A dermoid cyst, or dermoid, is the most common congenital midline nasal lesion. It consists of a sac-like growth containing elements of both ectoderm (stratified squamous epithelium) and mesoderm (adnexal structure including hair follicles and sebaceous glands). Unlike other craniofacial dermoids, the nasal dermoid may present as a cyst, a sinus, or a fistula and may have an intracranial extension. The incidence is estimated at 1:20000 to 1:40000 births [1, 2], and pathogenesis involves the incomplete obliteration of neuroectoderm in the developing frontonasal region. During the eighth and ninth weeks of embryogenesis, the nasal and frontal bones develop by means of intramembranous ossification in the mesoderm but remain separated by the fonticulus nasofrontalis. At this time, a space is formed between the nasal bone and the deeper cartilaginous capsule known as the prenasal space. A small projection of dura extends to the skin. As the nasal process of the frontal bone grows, the skin and dura become separated, and the projection of the dura becomes encircled by the foramen cecum. The dura normally obliterates, thereby severing the neuroectodermal connection. After obliteration of the neuroectodermal connection, the fonticulus nasofrontalis and foramen cecum fuse and the cribriform plates form. The most widely accepted developmental theory of a nasal dermoid is based on the finding that as the neuroectodermal tract recedes, dermal attachments can follow its course. Therefore, as the dura mater recedes from the prenasal space, it may pull the nasal ectoderm upward and inward to form a sinus or a cyst, depending on whether there is a continued connection to the nasal dorsal skin [1, 3].

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## Clinical Presentation

Children typically present at birth or in infancy either with a small punctum over the bridge of the nose, and/or with a midline nasal mass, most commonly along the dorsum. The cystic swelling may be present anywhere between the forehead and nasal tip. Hair protruding from a nasal dorsum punctum is pathognomonic for nasal dermoid. There may be intermittent discharge of sebaceous material. Progressive enlargement of a nasal dermoid can cause soft tissue and skeletal deformity. Local infection is common, and as a tract can extend intracranially through the foramen cecum or cribriform plate to the base of the frontal fossa, it can rarely be associated with serious infectious complications such as meningitis or brain abscess.

## Differential Diagnosis

The presence of the nasal dorsal swelling, particularly when accompanied by redness and inflammation from a local infection, can be confused with an inflammatory abscess, traumatic deformity, or a hemangioma. Where there is a significant intranasal component, the differential would include benign intranasal midline neoplasia (e.g., nasal polyps, juvenile nasal angiofibroma), malignant neoplasms (e.g., rhabdomyosarcoma), and congenital intranasal masses (e.g., glioma, meningoencephalocele). The range of differential diagnoses and the possibility of intracranial extension necessitates a thorough preoperative imaging evaluation.

## Radiological Features

CT and MRI are complementary and have become the gold standard in radiographic evaluation of nasal dermoid, eliminating the need for plain radiography. Preoperative imaging should assess the anatomy of the sinonasal and cranial base with proper evaluation for intracranial extension. High-resolution computed tomography (CT) has the advantage of clearly delineating bony anatomy and contrast-enhanced images allow differentiation of enhancing cartilage from a skull base defect, and enhancing nasal mucosa from nonenhancing dermoid. Images should extend from the tip of the nose through the anterior cranial fossa. The absence of bony skull base abnormalities include widening of the foramen cecum, osseous defect in the cribriform plate, or a bifid or eroded crista galli makes intracranial extension very unlikely, although their presence is not confirmatory (Fig. 6.1).

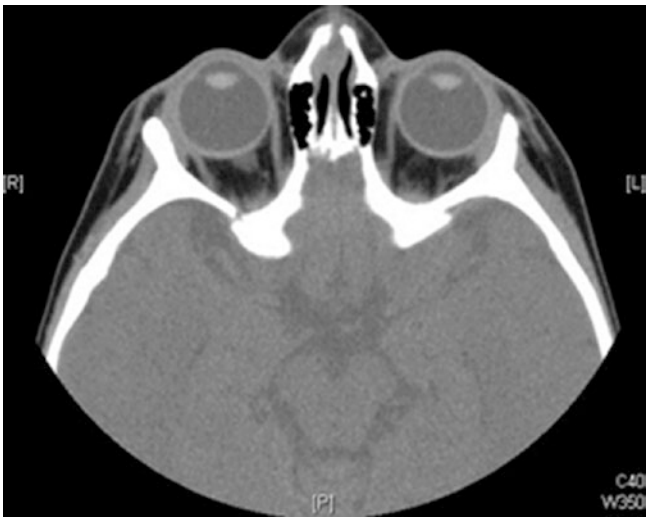


**Fig. 6.1** Axial CT demonstrating defect in central nasal bones with soft tissue density in subcutaneous tissues and extending deep to the nasal bones

Magnetic resonance imaging (MRI) can be used where intracranial extension is suggested by CT or in some cases as the primary investigation. Multiplanar (axial, coronal, and sagittal) high resolution, thin-section MRI with T1-weighted images and fat-suppressed T2-weighted or fast spin-echo sequences may allow clear delineation of a tract. Gadolinium-enhanced, fat-suppressed T1-weighted images can be used to depict the anatomy of the enhancing cartilage of the anterior skull base in infants, and use of contrast permits differentiation between nonenhancing dermoids and other enhancing masses such as hemangioma or teratoma. It is important to recognize that in infants, the crista galli is not fully ossified and does not contain marrow fat. As a result, a high-intensity signal on T1-weighted images in the vicinity of the crista galli in the newborn should suggest the presence of intracranial dermoid (Figs. 6.2 and 6.3).

## Management

Appropriate treatment of any infection and a thorough preoperative evaluation are essential before planning resection of a nasal dermoid. Many different approaches have been advocated for the removal of nasal dermoids, ranging from a simple external excision to complex procedures in which a combined external nasal and neurosurgical craniotomy approach are required. Incision and drainage, aspiration, curettage, or subtotal excision will

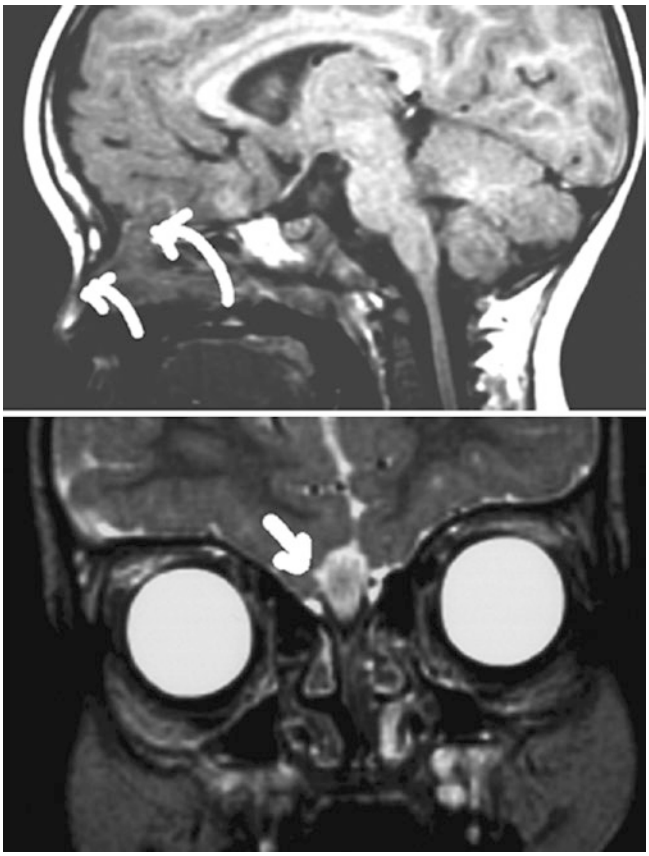

**Fig. 6.2** T1 sagittal and T2 coronal MRI clearly demonstrate tract with intracranial extension

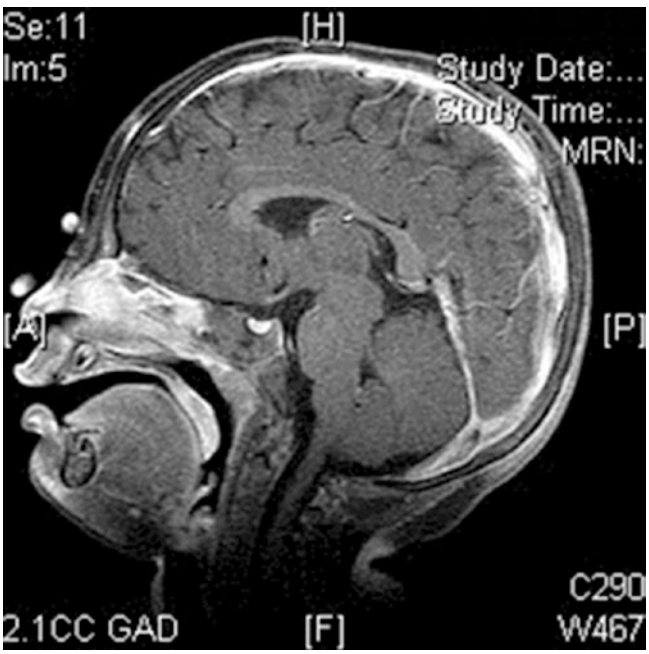


**Fig. 6.3** Gadolinium-enhanced, fat-suppressed T1-weighted images can be used to depict the anatomy of the enhancing cartilage of the anterior skull base in infants

fail to eradicate the cyst, resulting in recurrence in the vast majority of cases. A midline vertical incision remains the most common approach, and the cutaneous punctum can be removed in continuity with the tract and/or the cyst by making an elliptical incision around the sinus opening. A number of modified external incisions have been proposed aiming for either a better cosmetic outcome or to allow increased access, including a transverse incision, lateral rhinotomy, external rhinoplasty, inverted-U incision, and midfacial degloving procedure. Where the lesion is intranasal with minimal or no cutaneous involvement, an endoscopic transnasal approach excision with or without an intercartilaginous incision may allow excision of the lesion and its tract to the dura. The most common extent of the sinus tract is immediately deep to the nasal bones, and a limited nasal osteotomy may improve exposure. Here the nasal bones are fractured and separated vertically over the dorsum of the nose at the nasofrontal suture, allowing the tract to be followed. Where preoperative radiographic imaging has demonstrated a bifid and/or widened crista galli, but without an intracranial mass, a tract that appears to extend intracranially may often be fibrous without epithelium and not need formal excision by craniotomy. Frozen-section analysis may assist in differentiating fibrous tissue from a true epithelial tract, but there is a risk of missing epithelium and adnexal elements if a single biopsy site is chosen. A combined endoscopic and open nasal approach, introducing an endoscope between the displaced nasal bones, can be very effective for following intracranial extension to the skull base. With appropriate expertise, an endoscopic-assisted approach can be considered for extensive intracranial lesions, although the patient should be appropriately prepared for conversion to a craniotomy approach, particularly if inadequate access or an extensive cerebrospinal fluid leak occurs.

The decision-making for timing of resection for an isolated nasal dermoid without intracranial complication can be controversial, with early intervention having the advantage of reducing the potential risk for infection and the possible need for a more extensive procedure. Although the recurrence rate of nasal dermoid excision is low, this may occur several years after the initial surgery, so long-term follow-up is advised.

## Clinical Example 1

A newborn was noted to have a nasal tip mass that was soft and had a “whitish” hue visible beneath the skin.

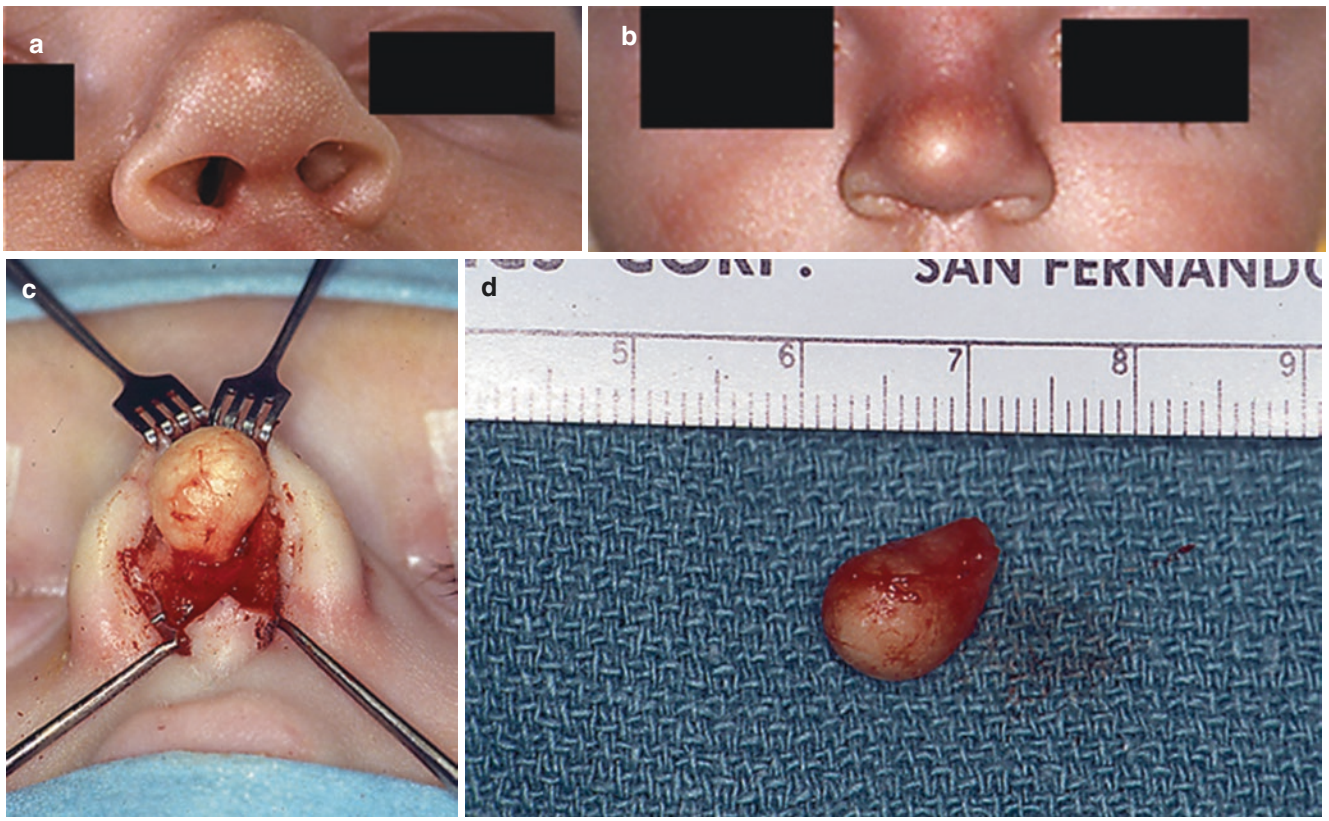


**Fig. 6.4** (**a**) Soft mass in nasal tip. No pit is seen. (**b**) "Whitish" hue of skin suggestive of keratinous contents of cyst. (**c**) Cyst exposed through transcolumellar and rim incision external approach. (**d**) Complete excision without disruption of spillage of keratin suggested no tract

There were no pits or sinus openings on the skin and the intranasal exam was normal. CT and MRI showed the mass consistent with a nasal dermoid without evidence of any intracranial extension.

Early local excision was performed through a transverse columellar external rhinoplasty incision (Fig. 6.4a–d).

## Clinical Example 2

A 7-year-old boy with a nasal tip soft tissue mass. Again a "whitish" hue is present beneath the skin suggestive of a cyst with keratinous content. No dimple or sinus opening was seen. CT and MRI showed a tract extending to the skull base without evidence of any intracranial component. Complete extension of the cyst and tract was performed through a transcolumellar external approach providing good access to remove the tract to the skull base (Fig. 6.5a–d).

## Clinical Example 3

A 3-year-old boy presented with an asymptomatic enlarging soft tissue mass on the dorsum of the nose. A nondraining nasal pit (sinus) was present in the midline close to the tip.

CT demonstrated a nasal dermoid cyst and sinus to the crista galli without any breach of bone or evidence of intracranial extension. Excision was performed through a horizontal dorsal incision with good access for complete excision (Fig. 6.6a–f).

## Clinical Example 4

A 2-year-old male presented with pain and erythema over the nasal dorsum. A sinus draining purulent material was identified at the nasal tip. The dorsum was widened. The patient was treated with antibiotics and drainage in preparation for

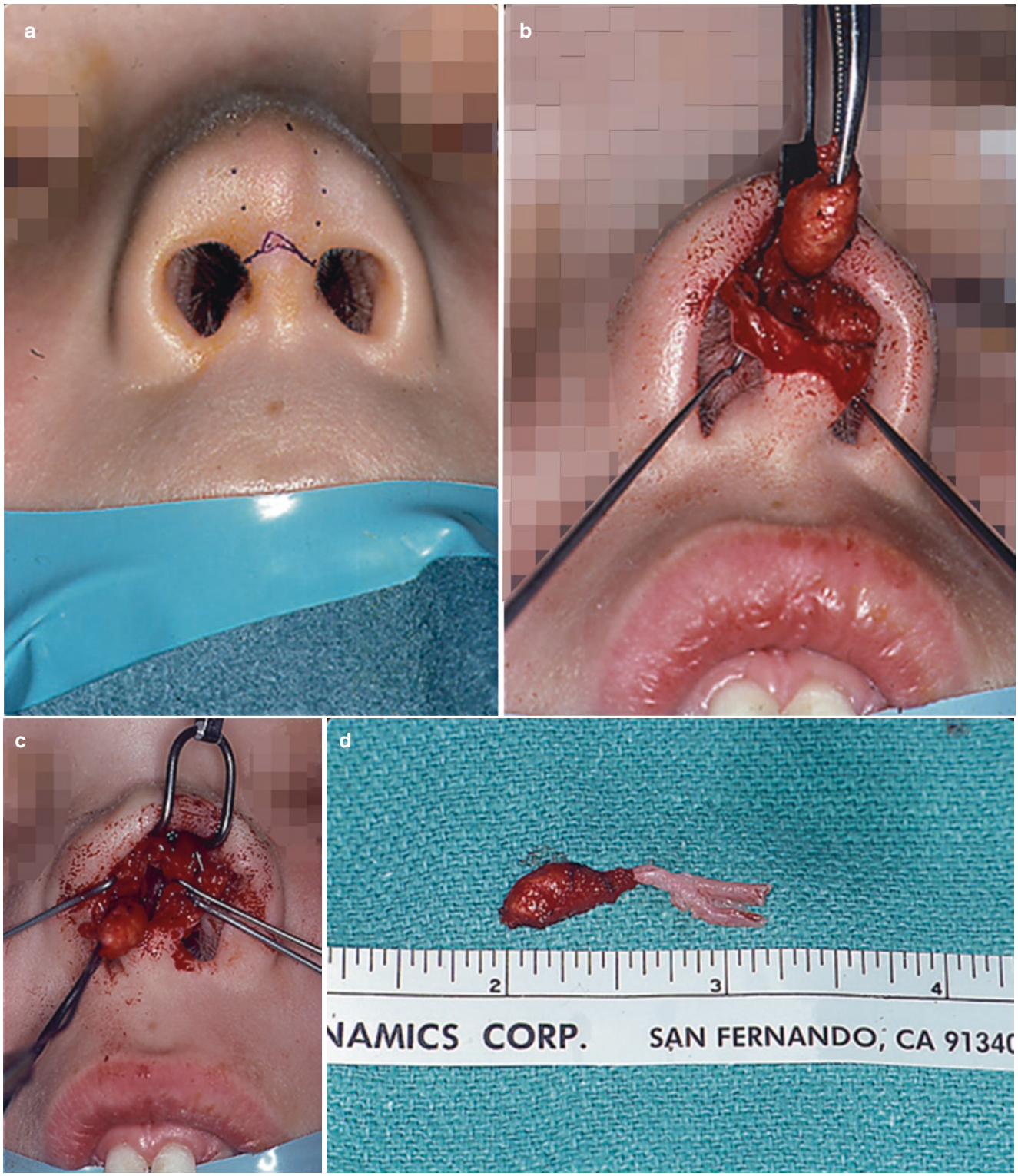


**Fig. 6.5** (**a**) Soft mass in nasal tip with planned incision. (**b**) Cyst and tract exposed. (**c**) Tract followed up the midline to the skull base through natural pathway created by the dermoid. The nasal bones can be gently displaced (splayed or spread open through the midline fissure) or osteotomized to create more exposure as needed. (**d**) Complete excision of cyst and tract to skull base. No evidence of keratin or any lumen of the tract at the proximal end ensuring complete excision. Interestingly the proximal (skull-base) end seemed bifurcated

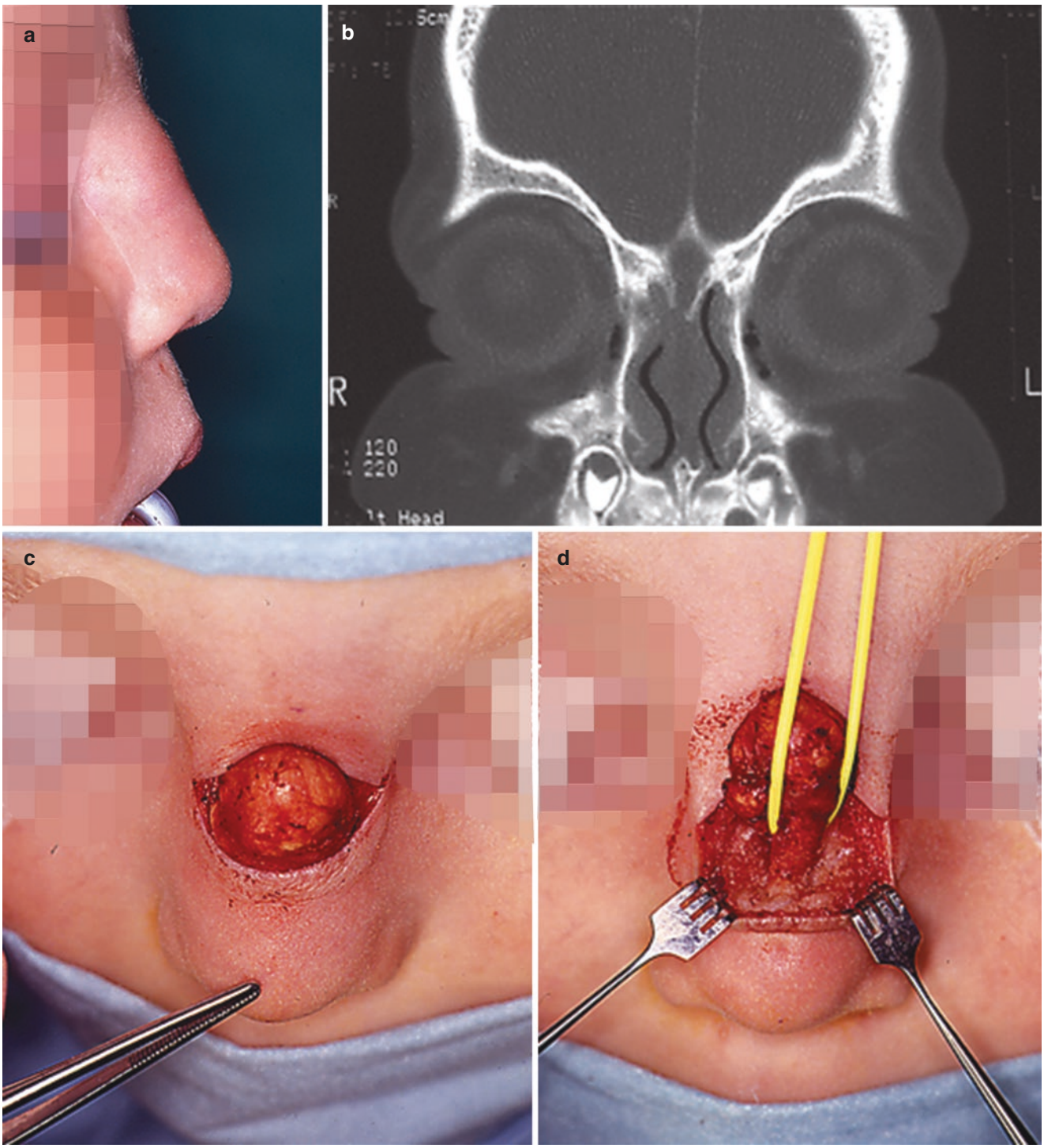


**Fig. 6.6** (**a**) Mass on nasal dorsum. (**b**) Coronal CT showing cavitation of skull base by cyst at crista galli without any breach of bone or intracranial extension. (**c**) Cyst exposed through horizontal incision on dorsum of nose. A midline sinus pit can be seen close to the nasal tip. (**d**) Tract toward tip pit dissected and removed. (**e**) Large tract followed through nasal bone defect to the skull base and complete excision performed with excellent exposure. (**f**) Excellent cosmesis 1 year post excision

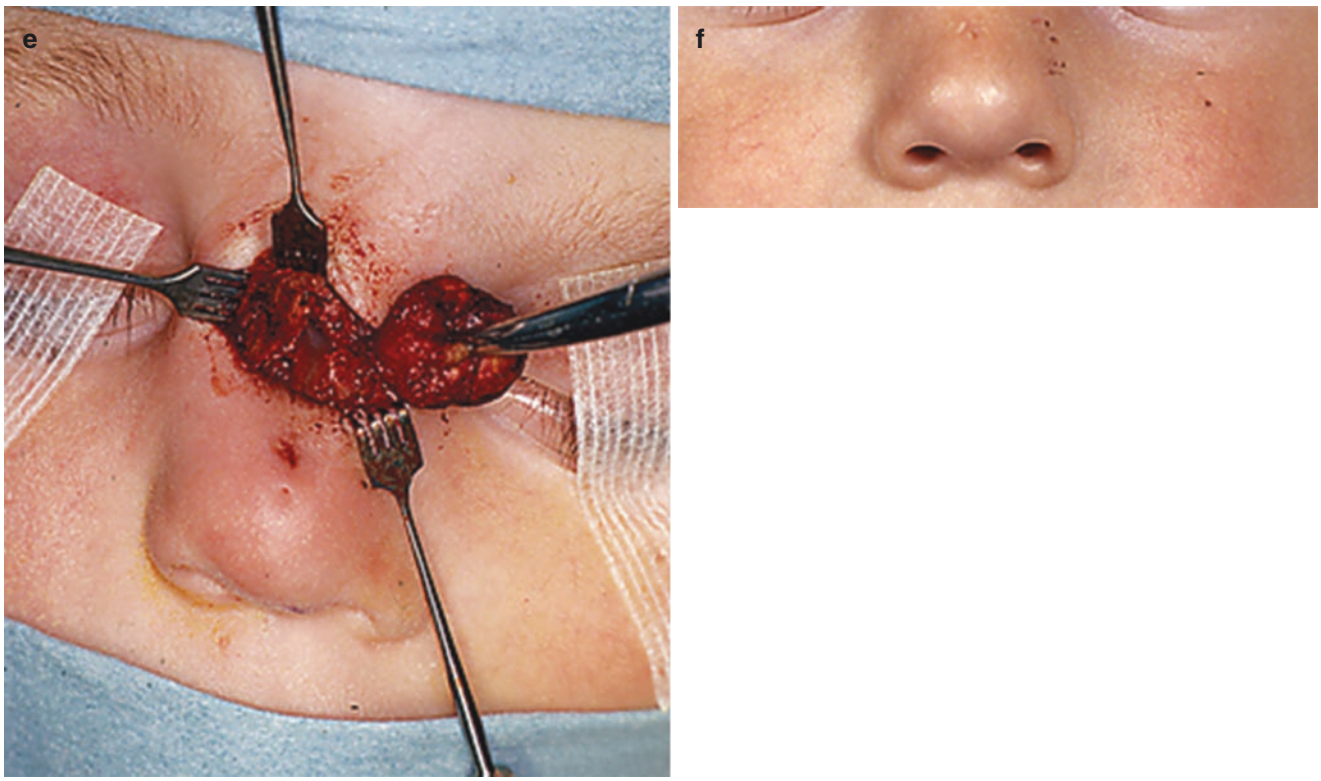


**Fig. 6.6** (continued)

definitive surgical management. A CT scan identified the sinus tract that extended deep to the nasal bones. There was no intracranial extension.

The lesion was excised via a midline vertical incision and division of the nasal bones. The skull base was intact. This approach typically has a very good cosmetic result as shown below two years after surgery (Fig. 6.7a–f).

## Meningoencephalocele

### Definition

A meningoencephalocele is the extracranial herniation of brain tissue and meninges through a defect in the skull base. Nasal meningoencephaloceles develop from a defect in the anterior neuropore in the region of the foramen cecum. The degree of herniation can vary from a meningocele (meninges alone) to a meningoencephalocele (brain and meninges).

### Clinical Presentation

Nasal meningoencephaloceles can usually be seen hanging in the nasal cavity where it may appear as nasal polyps, potentially leading to misdiagnosis and "nasal polyp removal," with potentially serious complications (e.g., persistent cerebrospinal fluid leakage, meningitis). Even without a preceding operation, spontaneous cerebrospinal leakage can occur, or the meningoencephalocele can present with meningitis. With crying, straining or compression of the jugular vein, encephaloceles may expand or pulsate due to their connection with the brain. This is called Furstenberg's sign and is used to differentiate encephaloceles from other pathologies such as gliomas and polyps.

### Differential Diagnosis

Nasal dermoids and gliomas, the other congenital midline nasal masses, are the main differential diagnosis of a meningoencephalocele. When the mass extends into the nasal cavity, it can be mistaken for a benign nasal polyp. Although rare in young children, a neoplastic lesion may be considered, such as a rhabdomyosarcoma. A hemangioma should also be included in the differential diagnosis of a pediatric nasal mass, although these are not necessarily midline.

### Radiological Features

Before a surgical intervention, cross-sectional imaging studies are required to assess the location, size, and contents of

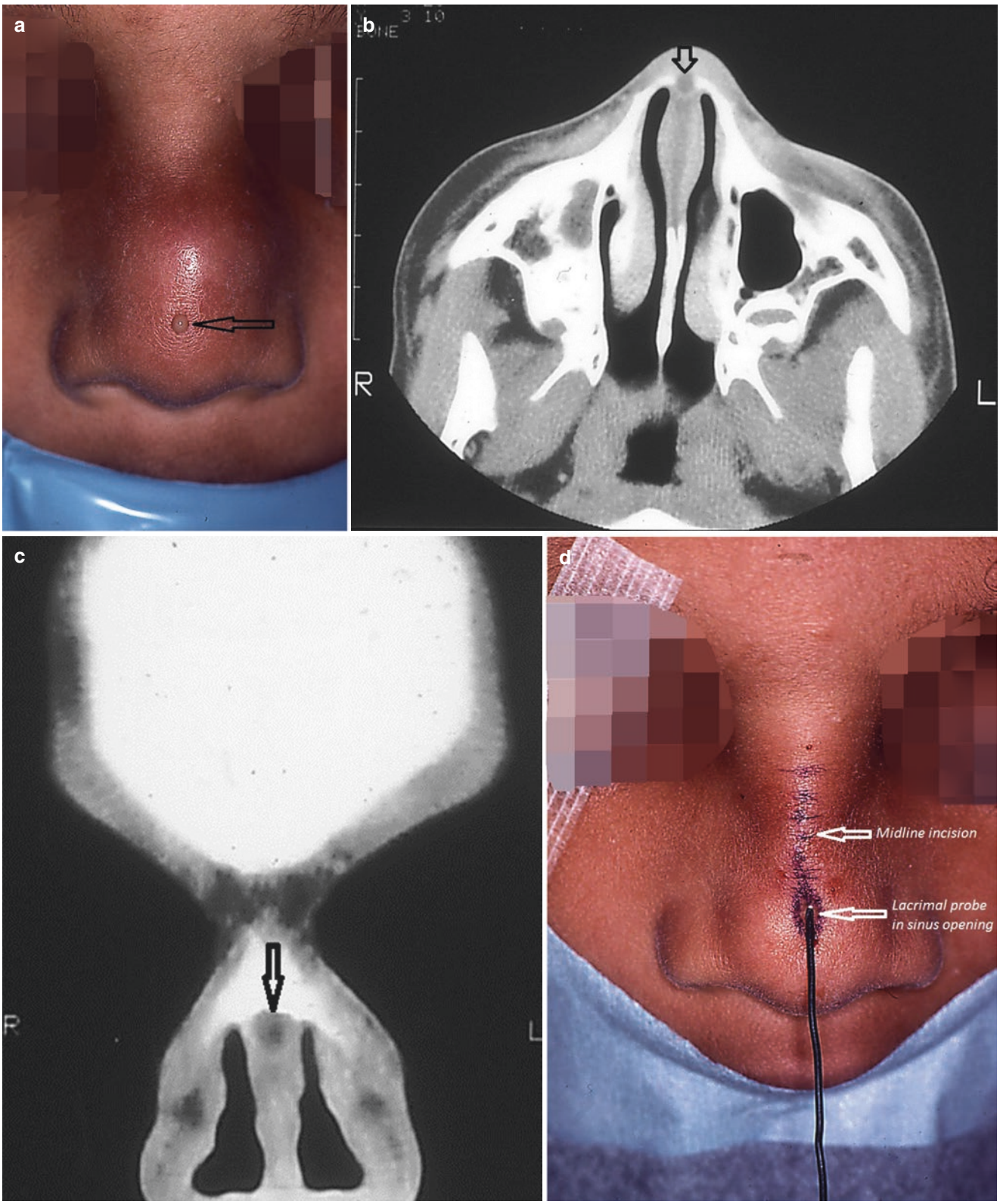


**Fig. 6.7** (**a**) Even after aggressive treatment with antibiotics, the sinus is still draining purulent material. The dorsum was widened and the skin was no longer infected. (**b**) Axial CT showing tract through midline between nasal bones. (**c**) Coronal CT demonstrates tract deep to nasal bones heading toward skull base. (**d**) Probe in sinus and excision planned through vertical midline incision. (**e**) Excellent exposure to skull base achieved through vertical approach. (**f**) Excellent cosmesis 2 years after excision

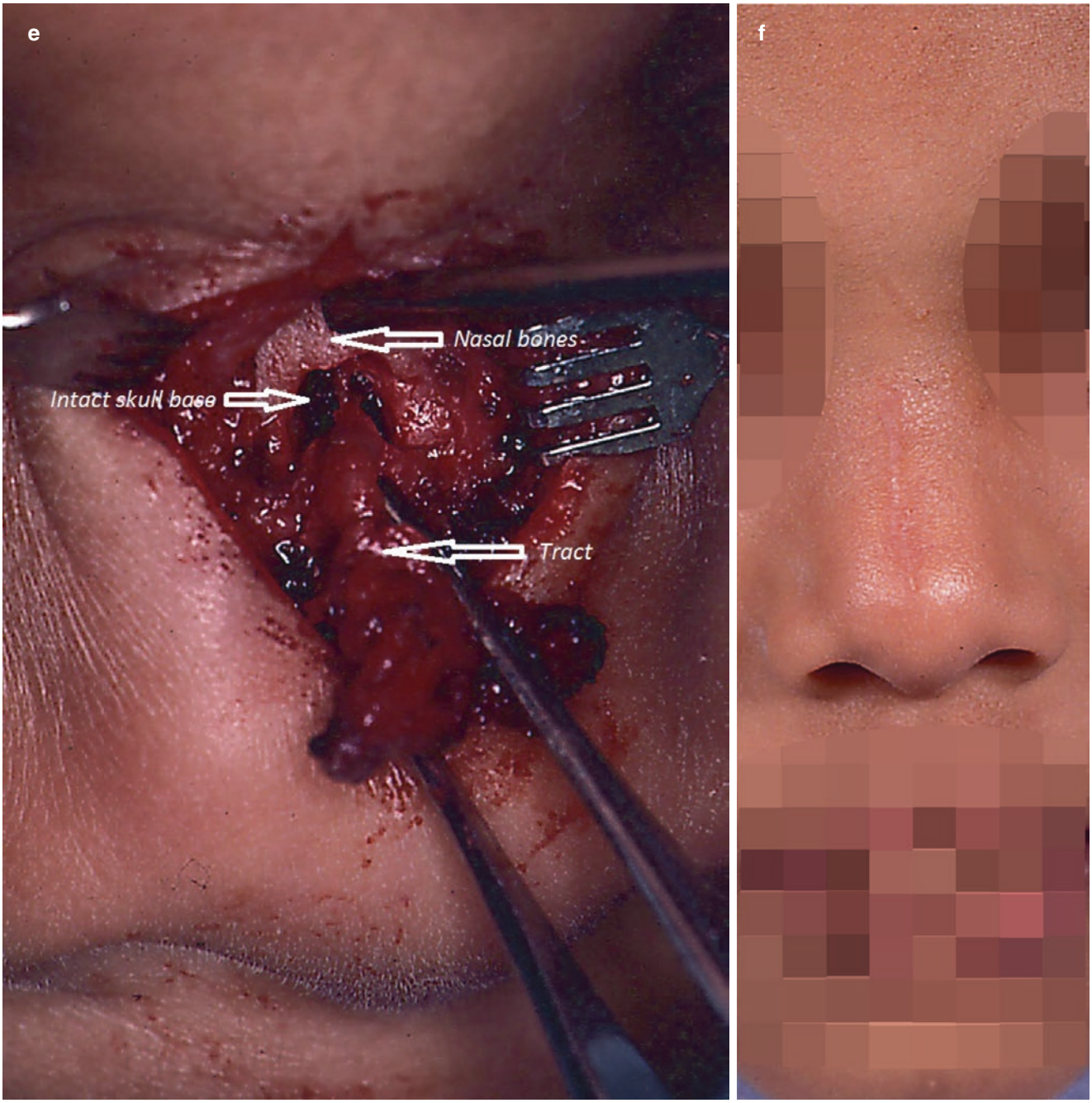


**Fig. 6.7** (continued)

the lesion. Specifically, the relationship with the skull base bone is easily obtained by high-resolution axial and coronal CT imaging. In young infants, the anterior skull base may be largely non-ossified and IV contrast may help used to delineate the cartilaginous skull base. High-resolution thin-slice multiplanar MRI provides complementary information to CT regarding the relationship with the brain, fluid, or soft tissue characteristics of the mass, and the degree of herniation (Fig. 6.8).

## Management

Meningoencephaloceles require surgical excision, and early treatment is advocated to reduce the risk of brain infection and progressively severe deformity. A transnasal endoscopic approach is ideal for excision of the extracranial-intranasal component of the encephalocele and can provide excellent exposure, avoiding the need for a classic transfacial or combined craniotomy approach. Where available, an intraopera-

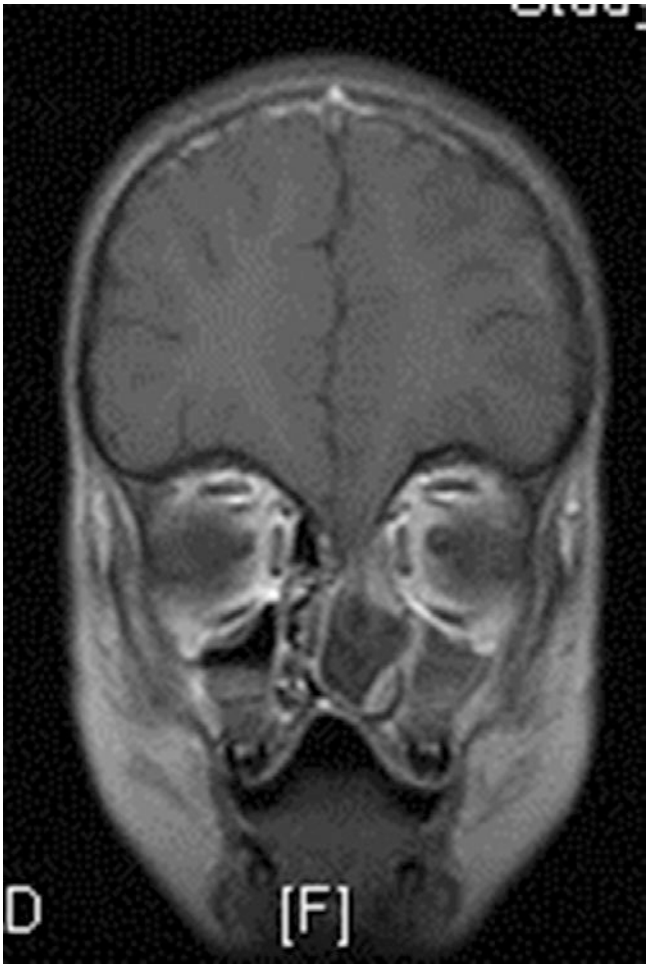


**Fig. 6.8** Coronal T1 MRI showing defect in anterior skull base with herniation of brain into left nasal cavity

tive image guidance system enhances the appreciation of the superior extension toward the skull base and may assist in ensuring complete resection of the mass. Insulated endoscopic bipolar forceps can be used to shrink and ablate the sac, making it easier to delineate and free the sac from the nasal septum and lateral nasal wall. The stalk of the sac can be transected and the main bulk of the sac amputated, using the bipolar cautery. This will allow visualization of the skull base and meningoencephalocele origin. Although the sac is fluid-filled, generally no CSF leak is encountered with transection as CSF may not be in free continuity. The stalk can then be further shrunk using the bipolar cautery until it regresses to the skull base. Mucosa around the skull base defect can then be elevated, and the size of the defect measured. The small bony defect in the skull base generally does not require closure, but a small piece of harvested turbinate or ethmoid perpendicular plate bone can be used. If a CSF leak was encountered, this can be repaired by using a synthetic dural replacement material. The defect site is typically covered using harvested mucosa from a turbinate or a nasal septal free mucosal graft. A pedicled vascularized septal flap is not helpful as it will not reach the skull base defect in a young child.

### Clinical Example

A 10-year-old boy presented with meningitis. During hospitalization and investigation was found to have a nasal encephalocele. He had a long standing history of nasal obstruction with a watery nasal discharge and had been treated for allergies.

Examination revealed a mass in the right nasal cavity. After treating his meningitis, he was successfully treated surgically with an anterior craniotomy (Fig. 6.9a–d).

## Nasal Glioma

### Definition

A nasal glioma is a congenital lesion that results from the herniation of intracranial tissues with faulty closure of the anterior neuropore, as in formation of a nasal meningoencephalocele. The difference in the development of the glioma is that the encephalocele becomes sequestrated from the brain and cranial vault early in gestation by closure of the skull base and the stalk degenerates, thereby obliterating any connection to brain parenchyma.

### Clinical Presentation

A nasal glioma is a firm, smooth, noncompressible mass. An intranasal glioma often manifests as a pale mass within the nasal cavity, possibly visible at the nostril and usually causing nasal obstruction. The base of the intranasal glioma commonly arises from the lateral nasal wall close to the middle turbinate and less commonly from the nasal septum [4]. There may be a component extending into the external nose anywhere between the nasal tip and glabella, and the nasal bridge may be widened. One rare occasions a nasal glioma may extend into the orbit, oral cavity, or nasopharynx. Gliomas do not demonstrate a positive Furstenberg's sign.

### Differential Diagnosis

The other congenital nasal masses, nasal dermoids, and meningoencephaloceles form the main differential diagnosis of a nasal glioma. In the nasal cavity, it could be mistaken for a nasal polyp or a rare malignant neoplasm such as a rhabdomyosarcoma.

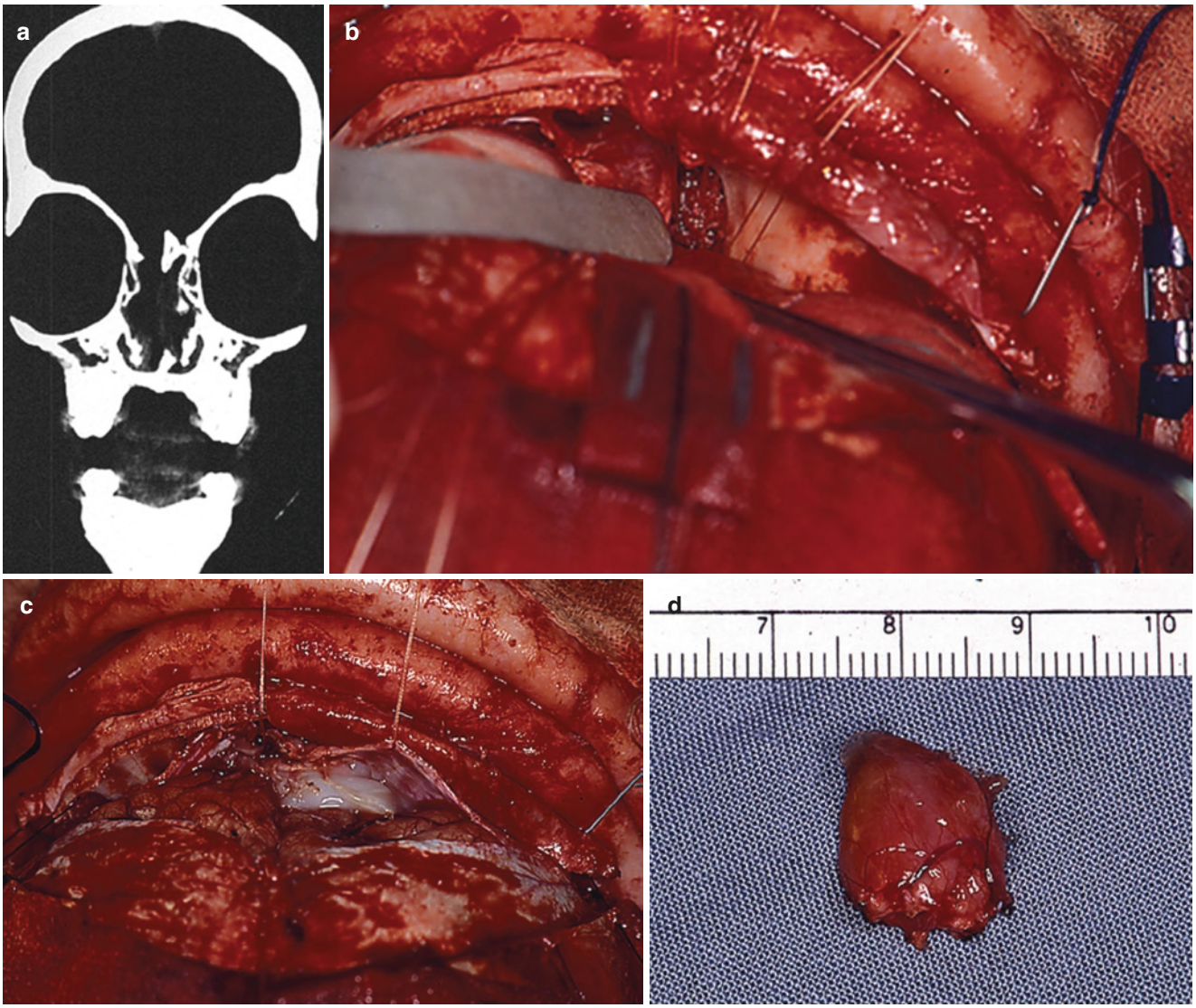


**Fig. 6.9** (**a**) Coronal CT showing bony defect in anterior skull base. (**b**) Defect of skull base exposed at craniotomy after separation of nasal portion of the encephalocele. Defect repaired with a hemi-pericranial flap. (**c**) Fibrin tissue glue used for extra seal followed by closure of the dura. (**d**) Nasal contents easily removed through the nose after separation from the brain

## Radiological Features

Nasal gliomas are usually isointense relative to normal brain at MRI, which is useful to explore the presence of any intracranial stalk. If there is suspicion of extension through the skull base or deformity of the nasal bones, CT imaging will allow characterization of the bony anatomy.

## Management

Treatment of a nasal glioma requires surgical excision, and a delay in removal may result in infection and distortion of the growth of the septum and nasal bones. Despite the lack of a true intracranial connection, some nasal gliomas may have a fibrous stalk extending toward the base of skull and even an underlying bony defect [5]. Excision of an intranasal glioma can be undertaken by a transnasal endoscopic approach, possibly assisted by image guidance. If the glioma has an extranasal component, it may require an external incision, such as an external rhinoplasty approach [5]. The approach should be based on the location and size of the mass, superior extension, and the presence of an associated deformity of the bone or cartilage. Nasal osteotomy may be required to follow a fibrous stalk to the skull base, and the surgeon should be prepared to manage an intracranial extension, with neurosurgical assistance if needed.

## Inflammatory Nasal Polyps

### Definition

Nasal polyps are "sac-like" noncancerous growths that occur in the nasal cavity or sinuses. The exact cause is unclear. They may be related to chronic inflammation of the lining of the sinuses among people who have allergies including fungal, cystic fibrosis, aspirin sensitivity, or certain infections. Allergies are very common in children but polyp formation is not so. The polyp itself represents an overgrowth of the mucous membranes.

### Clinical Presentation

Inflammatory polyp appears as a light bluish or pale, nonsensate, "boggy" sac-like mass in the nasal cavity (Fig. 6.10).

Symptoms of polyps in children include nasal congestion, loss of smell, mucoid nasal discharge, nasal speech, and mouth breathing. Recurrent sinusitis can also result from polyps but is rare in children. Long-standing large nasal polyps can cause destruction of the nasal bones and widening of the nose or even hypertelorism seen more commonly in patients with cystic fibrosis (Fig. 6.11).

A history of colic, eczema, asthma, hay fever, perennial rhinitis, paroxysmal sneezing may be present.

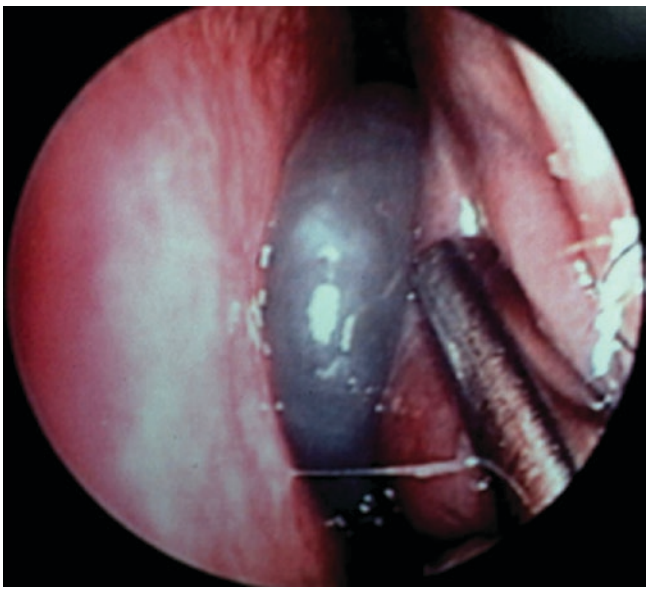

**Fig. 6.10** Inflammatory polyp can appear as a light bluish or pale, nonsensate, "boggy" sac-like mass in the nasal cavity

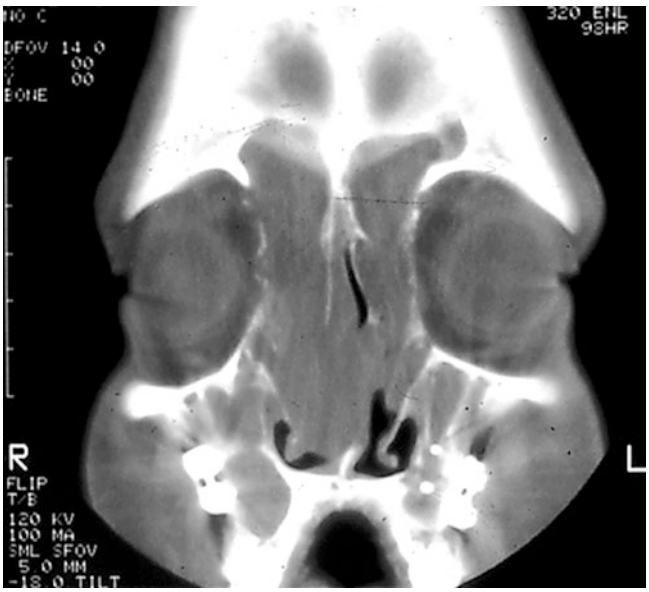


**Fig. 6.11** Coronal CT of teenage child with cystic fibrosis with chronic sinusitis and polyposis. The maxillary/frontal sinus are hypoplastic, soft tissue density fills the sinuses, and nasal cavity displacing the orbits

### Differential Diagnosis

The other benign congenital nasal masses, nasal meningoencephaloceles, and nasal glioma or rarely a malignant neoplasm such as a rhabdomyosarcoma.

### Radiological Features (Fig. 6.12)

### Management

Treatment for inflammatory polyps is typically with steroids, most often in the form of a nasal spray. If this is not effective, surgery may be considered. Despite removal or endoscopic sinus surgery, treatment may be refractory requiring the continued use of steroid nasal sprays. Antibiotics are not indicated for treatment unless an infection occurs.

## Antrochoanal Polyp

### Definition

Antrochoanal polyp is a slow-growing, benign, polypoid lesion arising from the mucosa of the maxillary sinus, which at presentation extends through the maxillary sinus ostium into the choana. Although bilateral presentation has been reported, antrochoanal polyps are almost always unilateral.

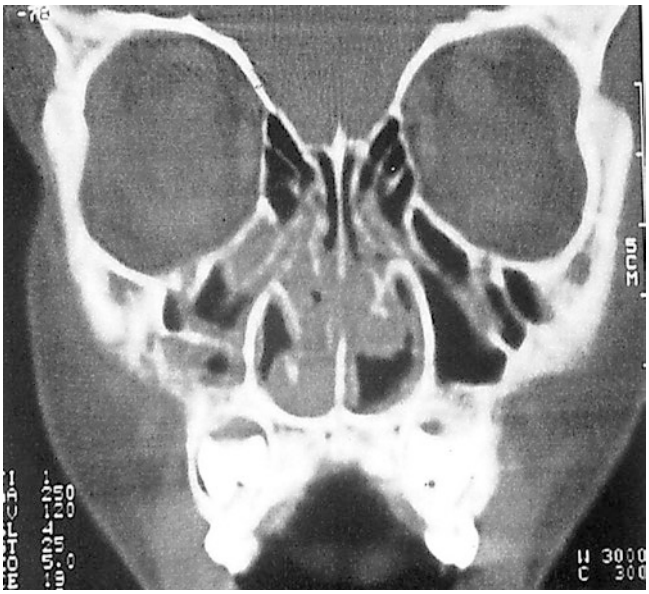

**Fig. 6.12** Coronal CT showing small bilateral polyps in child with chronic sinusitis

The pathogenesis and etiology of antrochoanal polyp remains unclear. The mass is smooth and does not cause bony destruction.

## Clinical Presentation

Antrochoanal polyp typically presents in a child or young adult with unilateral nasal obstruction and anterior nasal discharge. Endoscopic assessment of the nasal cavity demonstrates a smooth nasal polyp, usually filling the nasal cavity on the affected side. The posterior extent of the polyp may even be visible in the oropharynx if it extends through the choana and nasopharynx below the level of the soft palate.

## Differential Diagnosis

The differential diagnosis of antrochoanal polyp should include the other causes of nasal polyposis in children, such as chronic rhinosinusitis, cystic fibrosis, allergic fungal sinusitis, but these are almost always bilateral. Unilateral intranasal masses that should be considered include maxillary sinus mucocele, mucous retention cyst, juvenile nasal angiofibroma, meningoencephalocele, inverted papilloma, or hemangioma.

## Radiological Features

Antrochoanal polyps are well demonstrated on sinus CT as a soft tissue mass filling and sometimes expanding the maxillary sinus, passing through the natural or accessory maxillary ostium between the middle turbinate and the lateral nasal wall, and usually extending posteriorly through the nasal cavity to the choana. There is no bony destruction. The precise location of the mucosal origin within the maxillary sinus cannot usually be determined.

## Management

The management of antrochoanal polyp is endoscopic transnasal surgical resection, aided by powered instrumentation. Without identification and removal of maxillary sinus portion of the antrochoanal polyp, there will be a higher risk of recurrence through residual polyp regrowth. Angled-view endoscopes and curved instruments facilitate maxillary sinus clearance, especially of the lateral and anterior extent. In some cases, especially with recurrence, a combined transnasal and anterior canine fossa opening endoscopic approach may be required to ensure optimum access and visualization within the maxillary sinus. This may be particularly helpful where the mucosal origin of the antrochoanal polyp is from the anterior or anteroinferior wall of the maxillary sinus, which is challenging to reach transnasally.

## Clinical Example

A 9-year-old girl presented with long-standing history of nasal obstruction without any history of sinusitis or allergy. The parents had noted progressive heavy snoring and periods of obstructed breathing consistent with mild sleep apnea. She had a pale, non-sensate, easily visible polyp in the left anterior nasal cavity (Fig. 6.13). There was no family history of cystic fibrosis and the child was otherwise healthy.

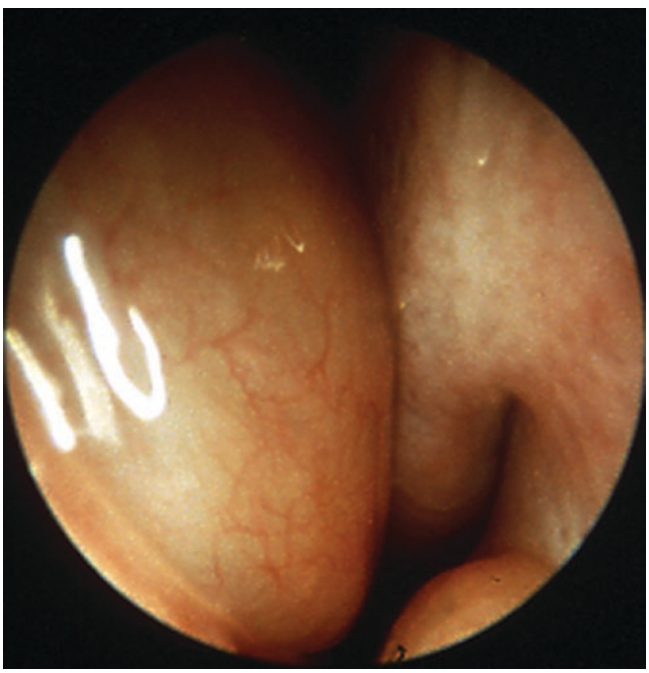

**Fig. 6.13** Polyp in left nasal cavity

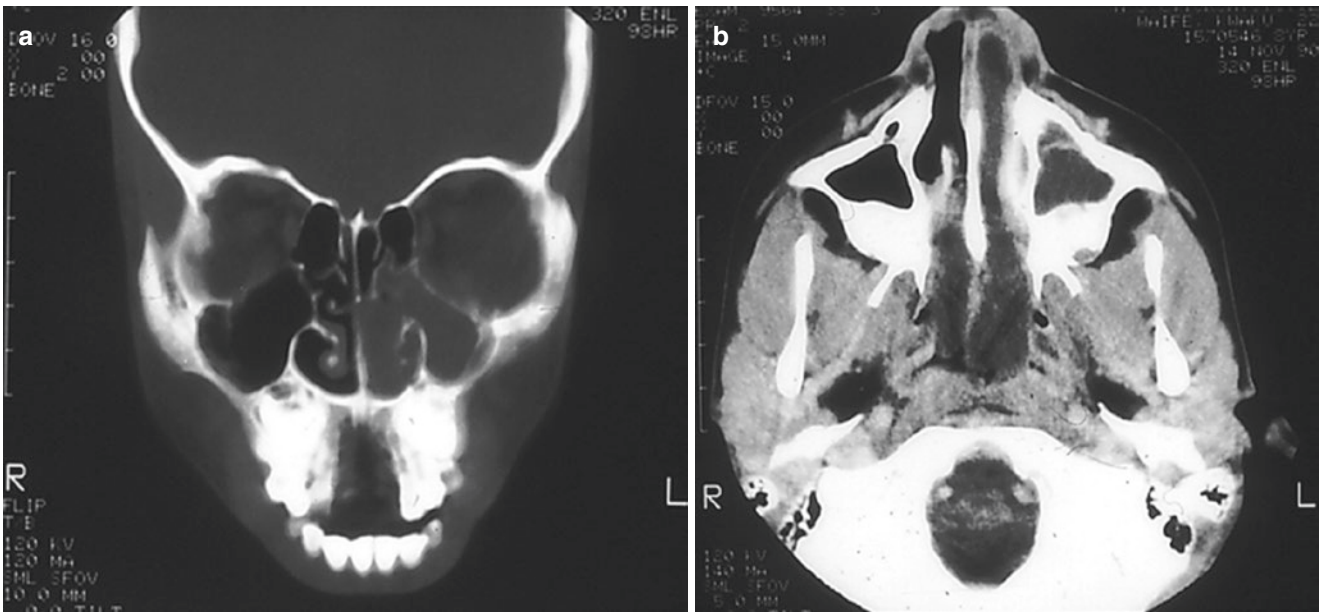


**Fig. 6.14** (**a**) Coronal CT showing mass in maxillary sinus and left nasal cavity. Other sinuses appear normal. (**b**) Axial CT showing complete filling of left maxillary sinus, left nasal cavity, and nasopharynx

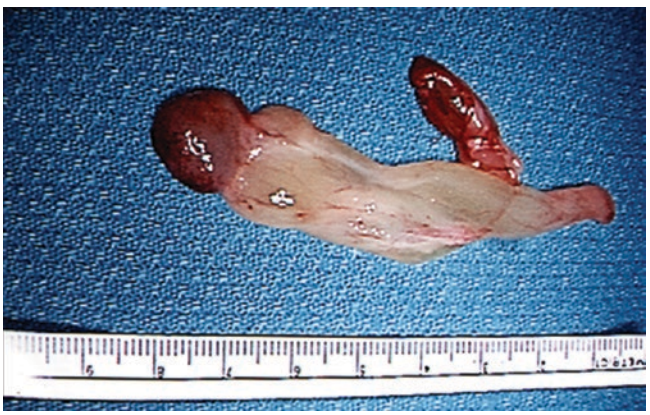

**Fig. 6.15** Gross appearance of large antrochoanal polyp after removal

A CT scan was done (Fig. 6.14a and b), and the polyp was successfully removed at surgery (Fig. 6.15).

Pathology showed an inflammatory polyp.

## Inverted Papilloma

### Definition

Inverted papilloma is a neoplastic growth of the nasal epithelium, which inverts into the underlying stroma, also known as Schneiderian papilloma or Ewing's papilloma. It is rare in children, most commonly arising in males over 40 years. The etiology is uncertain, but there may be an association with HPV virus or environmental exposure.

### Clinical Presentation

Inverted papilloma typically presents with nonspecific nasal symptoms, including unilateral nasal obstruction, rhinorrhea, facial pain, and epistaxis. Endoscopic examination usually demonstrates a unilateral polypoidal mass within the nasal cavity. The poly is typically reddish gray, irregular, and friable. The mass may extend into the nasopharynx and may cause bowing of the nasal septum to the contralateral side.

### Differential Diagnosis

Other intranasal lesions that can present in a similar way include benign nasal polyp, hemangiomas, pyogenic granulomas, and neoplasms such as rhabdomyosarcoma. On endoscopic examination, appearances are more characteristic, with a unilateral polypoid nasal mass originating from the lateral nasal wall.

### Radiological Features

CT scanning is necessary to identify any sinus involvement and bony erosion and should be performed before surgical excision is planned.

### Management

Inverted papilloma requires surgical excision. A transnasal endoscopic approach usually provides excellent visualization facilitate complete removal, avoiding an open approach.

### Clinical Example

A 15-year-old boy with a long history of right nasal obstruction had noted recent swelling and distortion of the face. There was no history of pain or bleeding, and the boy was otherwise healthy. Soft pale polyps were noted in the right nasal cavity. Inflammatory polyps were suspected after CT. Removal was performed via a nasal and sublabial transantral endoscopic approach. Pathology revealed the presence of inverted papilloma in one of the polyp fragments removed (see Chap. 7; Figs. 6.16, 6.17 and 6.18).

## Allergic Fungal Sinusitis

### Definition

Allergic fungal sinusitis (AFS) is a distinct eosinophilic subtype of chronic rhinosinusitis characterized by Type I hypersensitivity by history, skin testing, or serology; nasal polyposis; characteristic CT imaging findings; eosinophilic mucus; histologic evidence of eosinophilic mucus without evidence of fungal invasion into sinus tissue; and positive fungal stain of sinus contents [6]. The commonly encountered fungal species in children are *Aspergillus, Bipolaris, and Curvilaria* [7]. There may be a male preponderance, and the age at presentation is typically adolescents or young adults. The etiologic basis of AFS is an overly active immunologic response, manifest as an allergy to ubiquitous fungal species, and atopy and asthma are unsurprisingly associated. There exists a variation in prevalence with climatic factors, as there is a clear geographic pattern of distribution focused in temperate climates.

### Clinical Presentation

The presentation of AFS is fairly typical of chronic rhinosinusitis. In the pediatric population, this manifests as nasal airway obstruction, nasal discharge (sometimes purulent),

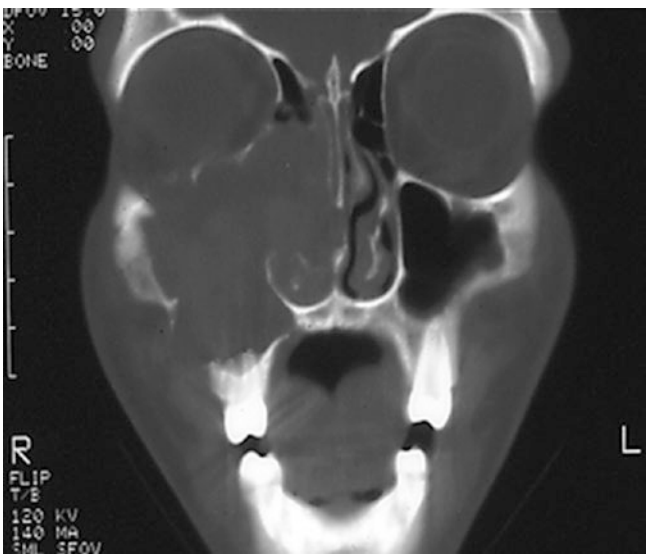


**Fig. 6.16** Coronal CT showing expansion of maxillary sinus by soft tissue density. There is thinning and destruction of bone of all the walls of the maxillary sinus

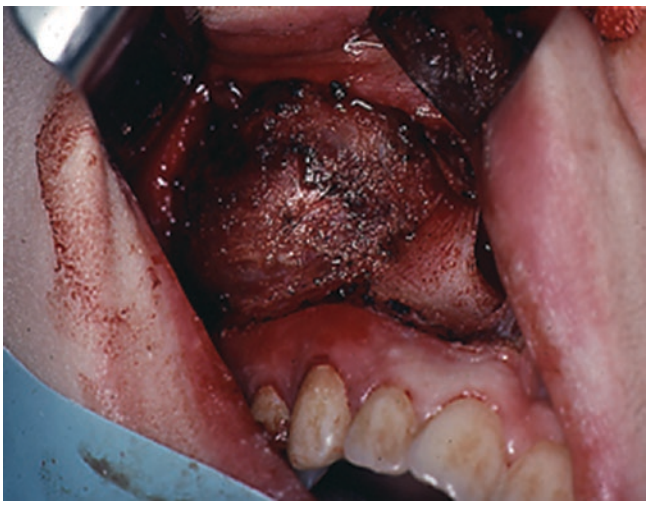

**Fig. 6.17** Sublabial approach at surgery showing expansion and thinning of anterior maxillary wall bone

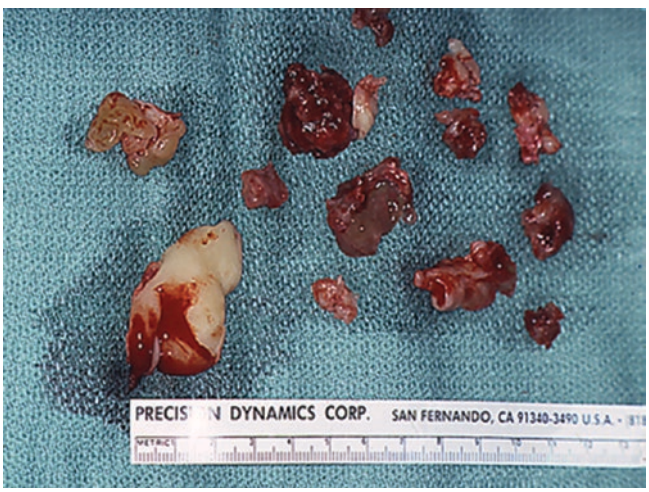


**Fig. 6.18** Soft tissue polyps removed with endoscopic approach via combined nasal and sublabial transantral approach

reduced sense of smell and/or taste, and less commonly headaches. The onset is typically a protracted indolent process, and pain is not usually present unless there is a concomitant bacterial infection. Concurrent atopy or asthma is usually elicited in the history. If endoscopy is tolerated, the examiner may note allergic mucin or nasal polyposis. Although history and physical examination may suggest AFS, definite diagnosis depends on histopathologic findings, so will not be made until after surgical intervention.

### Differential Diagnosis

The differential of AFS in children is broad, with signs and symptoms typically consistent with chronic rhinosinusitis. Other conditions that may mimic AFS include bacterial rhinosinusitis, adenoiditis, nasal polyposis associated with cystic fibrosis or primary ciliary dyskinesia, nasal tumor, nasal foreign body, antrochoanal polyp, or allergic rhinitis.

### Radiological Features

CT imaging plays a key role in diagnosis and surgical planning for this condition. Findings usually include opacification of multiple sinuses combined with more osseous expansion or erosion than is typical of other forms of chronic rhinosinusitis. The disease can often be asymmetric in children, with 70% of pediatric patients presenting with unilateral disease, compared with only 37% of adults [7]. A common feature is the presence of multiple densities without bony erosion (Fig. 6.19).

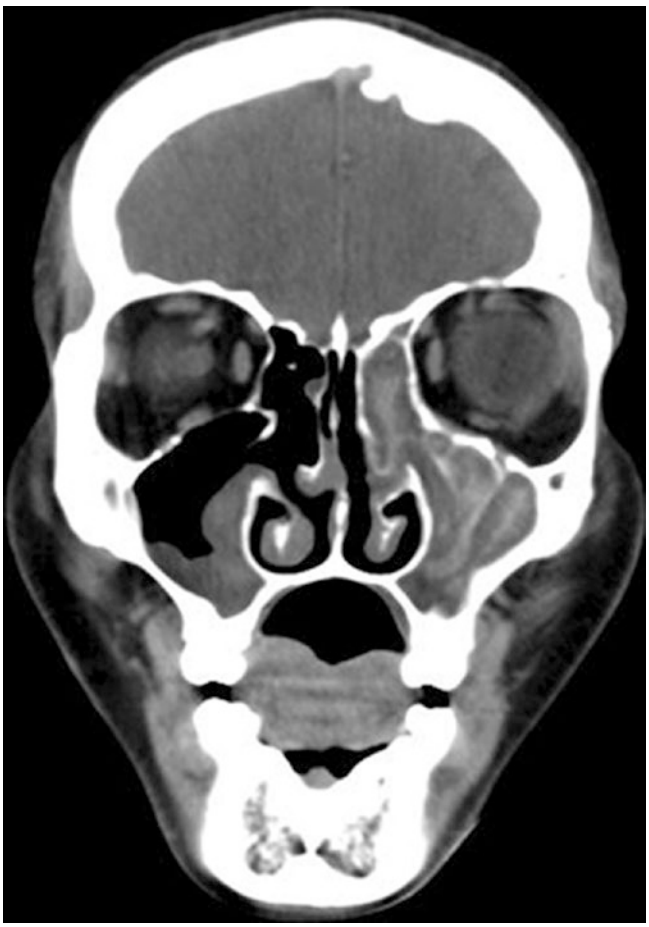

**Fig. 6.19** Coronal CT in patient with fungal sinusitis showing multiple densities in left maxillary and ethmoid sinuses without bony erosion or destruction

### Management

Unlike most chronic rhinosinusitis, the treatment of AFS is typically combined medical and surgical therapy, as opposed to initial medical therapy with failure warranting surgery. The surgical intervention of choice is functional endoscopic sinus surgery, with a focus on tissue preservation to maintain sinonasal mucociliary clearance pathways. The goals of this surgery are relief of obstruction, clearance of debris, and expansion of natural sinus drainage pathways. Preoperative planning using CT imaging is necessary to reduce risk of injury through osseous expansion distorting normal anatomy, and intraoperative image guidance may be helpful where available. Surgery will reveal the thick allergic mucin and sinus debris, which will contain fungal elements and eosinophils on histopathology confirming the diagnosis. If there is inadequate clearance of this mucin and fungal elements, the risk of early recurrence is increased [7]. Adjuvant medical therapy consists of preoperative systemic steroids to reduce the inflammation and polyposis, which improves surgical visualization, and postoperative nasal irrigation and further systemic steroids, eventually transitioning to local topical steroid therapy. Unfortunately, evidence has not supported significant benefit through the use of topical or systemic antifungal agents [7]. Immunotherapy, such as leukotriene receptor antagonists binding IgE, may have a role in the long-term control of disease. It should be made clear from the outset that surgical and medical therapies are not curative, and all patients have the potential for recurrent or persistent disease, which may require long-term therapy and follow-up.

## Invasive Fungal Sinusitis

### Definition

Invasive fungal sinusitis is often a life-threatening condition that uniformly requires surgical debridement and aggressive

antifungal therapy. There are two main subtypes that are distinguished from each other based on their onset and the affected population. Acute fulminant invasive fungal sinusitis (AFIFS) is a life-threatening condition that is rapidly progressive and occurs in immunocompromised patients. Whereas chronic invasive fungal sinusitis (CIFS) typically affects the immunocompetent population and has a gradual onset. AFIFS is the most aggressive form and can be a source of significant morbidity and mortality. The fungal infection rapidly invades the sinus bony structures, progressing over days to involve the orbit, cavernous sinus, and the brain. The filamentous organisms involved, typically *Aspergillus*, have the ability to invade blood vessels causing tissue necrosis and thrombosis. Although rare, immunocompromised children that may be affected are those with hematologic malignancy, specially stem-cell transplant recipients, or those with insulin-dependent diabetes. CIFS is characterized by significant fungal invasion into the mucosa of the paranasal sinuses, progressing slowly over many weeks or months. The fungal infection results in a granulomatous response. There is a gradual destruction of the bony paranasal sinuses, eventually extending posteriorly along the orbit resulting in blindness, or into the brain causing central manifestations.

## Clinical Presentation

The usual presentation of AFIFS is severe pain, in the face or forehead, associated with purulent or bloody nasal discharge. Patients may present with complications such as visual disturbance, signs of stroke, or conscious-level changes. Clinical signs may include darkened lesions on the nasal mucosa, altered facial cutaneous sensation, lack of bleeding from nasal mucosa with abrasion, facial asymmetry, periorbital swelling, proptosis, or cranial nerve palsies. The signs and symptoms of CIFS are similar, particular orbital involvement, but the onset is much slower.

## Differential Diagnosis

In the immunocompromised patient, a low threshold for a diagnosis of AFIFS should exist in the presence of the above features. Endoscopic assessment and CT imaging will assist the diagnosis and should be undertaken urgently. The differential would include complicated acute sinusitis or sinonasal malignancy. For CIFS, the differential is broader, with the need to rule-out sinonasal tumors, complicated chronic rhinosinusitis, or noninvasive allergic fungal sinusitis. Generally, CT imaging and histopathologic confirmation are required.

## Radiological Features

In CIFS, CT usually shows homogeneous dense opacification of the affected sinuses, relative lack of sinus expansion, irregular or mottled bone destruction, and largely extrasinus bone erosion. In contrast, AFIFS usually demonstrate hypoattenuating soft tissue opacification of the sinuses, with extensive bone destruction and fat-stranding on the outside of the sinuses. MRI is the best modality for assessing soft tissue extension in AFIFS, with intermediate to low signal on T1 and T2-weighted images, and absent sinus mucosal enhancement on contrast imaging, which suggests necrosis. There may be increased enhancement outside the sinuses where there is invasion, for example, in the intraorbital fat, masticator space, or pterygopalatine fossa.

## Management

CIFS should be aggressively treated with a combined medical and surgical approach, with debridement of affected tissues, although this may not be possible in extrasinus tissues or may sometimes require orbital exenteration may be required. This is combined with systemic long-term antifungal therapy, which may succeed in eradicating the disease even when complete surgical removal is not possible. In AFIFS, aggressive surgical debridement of necrotic tissue and antifungal therapy are urgently required, although even with prompt treatment mortality rates remain high. Immunosuppressive agents should be decreased if at all possible and any diabetic ketoacidosis controlled. In the more aggressive forms of AFIFS, antifungal therapy should be initiated without awaiting histopathologic confirmation of the diagnosis to avoid the delay causing a poorer prognosis.

## Clinical Example

A 10-year-old girl complaining of severe headaches and facial pain was diagnosed with acute fulminant invasive fungal sinusitis (AFIFS). She had been receiving aggressive chemotherapy for ALL that was poorly responsive to therapy. During treatment, she had developed severe neutropenia. Despite systemic antifungal therapy and G-CSF (granulocyte colony stimulating factor), she progressed to develop intracranial involvement that unfortunately lead to her death (Fig. 6.20).

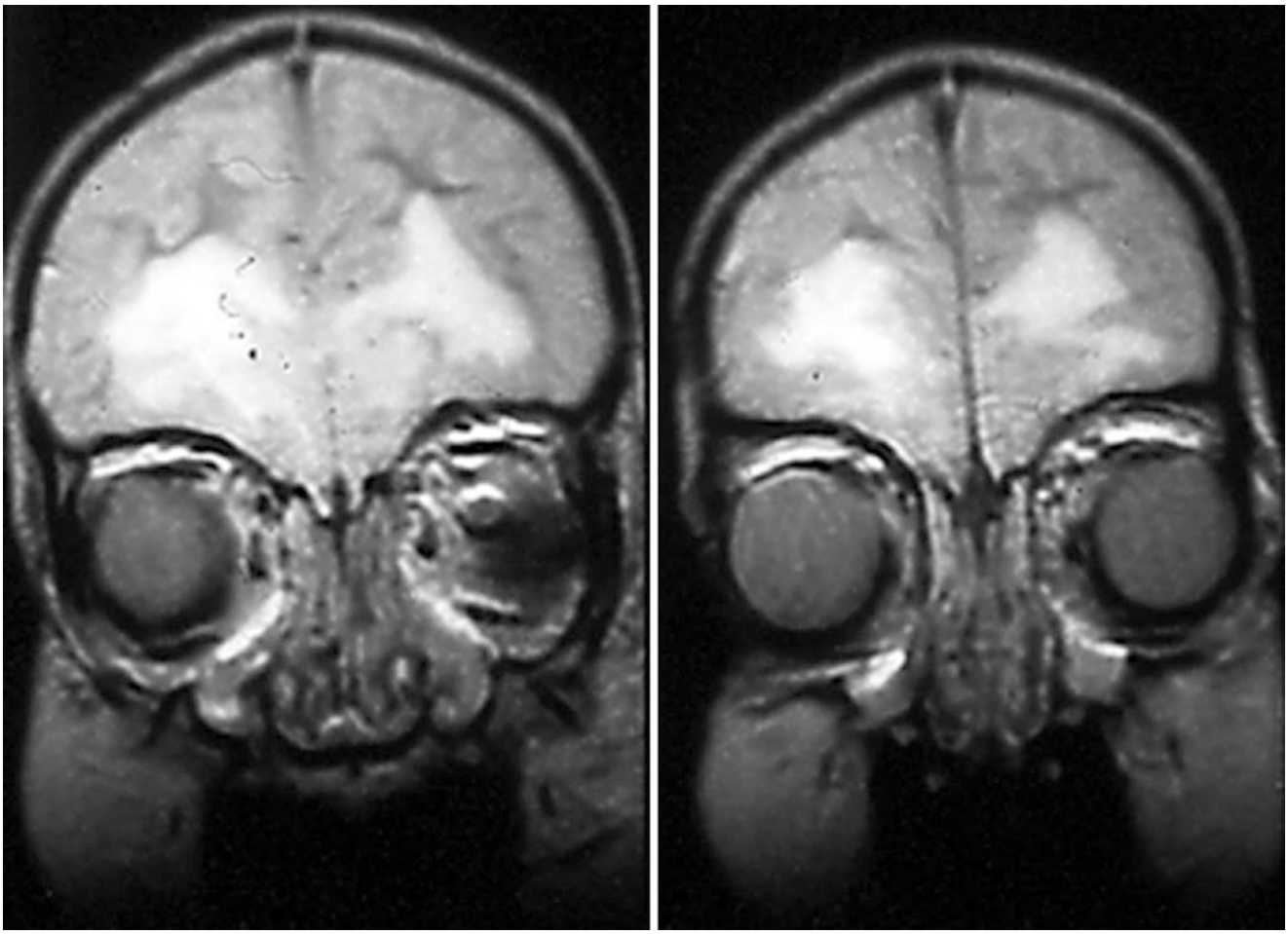

**Fig. 6.20** Coronal MRI showing invasive fungal involvement of both frontal lobes

## Pyogenic Granuloma of the Nasal Septum

### Definition

Pyogenic granuloma of the nasal cavity is also known as lobular capillary hemangioma, eruptive hemangioma, or granulation tissue-type hemangioma. The precise pathogenesis continues to be debated. It has been postulated to be associated with injury and hormonal factors and can occur in all age groups, including during pregnancy. The anterior septum is the most frequently affected site in the nasal cavity, followed by the inferior turbinate, but lesions have been described arising from the maxillary sinus, the roof of nasal cavity and the floor of nasal vestibule.

### Clinical Presentation

Nasal pyogenic granuloma usually presents as epistaxis and unilateral nasal obstruction. A soft fleshy vascular growth is either visible at the nasal vestibule by anterior rhinoscopy or can be identified endoscopically. The lesion tends to have a fairly rapid growth, and onset may be associated with a specific event of intranasal trauma.

### Differential Diagnosis

The differential diagnosis of nasal pyogenic granuloma in children includes nasal polyposis, juvenile nasal angiofibroma, granulomatous polyangitis (Wegener's disease), simple granulation tissue, recurrent respiratory papilloma, rhabdomyosarcoma, and lymphoma.

### Radiological Features

Radiological evaluation is only indicated to complement endoscopic assessment where the posterior extent of a large lesions cannot be determined and to exclude malignancy where there is concern that a rapidly enlarging mass may have skull base involvement or invade bone or soft tissues. In

most cases, the origin and attachment of the mass to the anterior nasal septum or lateral nasal wall can be clearly identified by endoscopy.

### Management

Total excision of the lesion by a transnasal approach. Endoscopic techniques provide better visualization of the mass and surrounding anatomy, thus facilitating the surgeon to remove the mass completely. Recurrence is uncommon, and malignant transformation of pyogenic granuloma has not been reported.

### Clinical Example

A 17-year-old girl was referred from hematology oncology. She had presented to them with right epistaxis and a vague familial history of a bleeding disorder. After hematological investigation, that was ruled out.

She was found to have a small polypoid mass in the right nostril based on the septum, which was suspected of being related to digital nasal trauma. Simple excision of the anterior nasal mass/polyp was carried out, and pathology was that of a pyogenic granuloma. See Chap. 7, Lobular Capillary Hemangioma.

## Fibrous Dysplasia

### Definition

Fibrous dysplasia describes a non-neoplastic condition of bone that is manifested by the accumulation of intermedullary fibrous tissue and immature woven bone, resulting in a mass of fibro-osseous tissue. This can occur in a single bone (monostotic) or multiple bones (polyostotic), and the latter form may be a manifestation of McCune-Albright syndrome, a separate condition that may have associated endocrinopathy and skin discoloration. In the polyostotic form of fibrous dysplasia, the craniofacial involvement is common, particularly affecting the ethmoid bones and less commonly the sphenoid, frontal bones, and bones of the cranial vault.

### Clinical Presentation

Fibrous dysplasia of the sinonasal region most commonly presents with craniofacial asymmetry and deformity or with a mass effect on adjacent cranial structures. Pain may occur due to the deformity or cranial nerve compression. Other local mass effects, depending on the location, may include nasal obstruction, proptosis, visual impairment, or other secondary lesions such as mucoceles and aneurysmal bone cysts. Extracranial involvement is rare.

### Differential Diagnosis

The lesion is usually diagnosed on clinical and radiological basis, but imaging differentials include a juvenile aggressive cemento-ossifying fibroma (more rapid growth, monostotic, and well-delineated radiographic margins), intraosseous meningioma (typically abuts intracranial compartment), Paget disease (usually skull vault sparing facial skeleton), sclerotic metastases (minimal expansion). If the imaging is consistent with the diagnosis, biopsy is generally not required [8].

### Radiological Features

Affected bones demonstrate a variety of radiographic features ranging from lucency to sclerosis. On plain radiography, features include blistering/bubbling cystic skull vault lesions, lesions crossing sutures, sclerotic skull base, a widened diploic space with displacement of outer table, sparing of the inner table (in contrast to Paget disease of bone, where the inner table is involved), and partial or total obliteration of paranasal sinuses.

CT typically demonstrates an expansible lesion, which is relatively homogeneous in appearance, described as "ground glass" (Fig. 6.21).

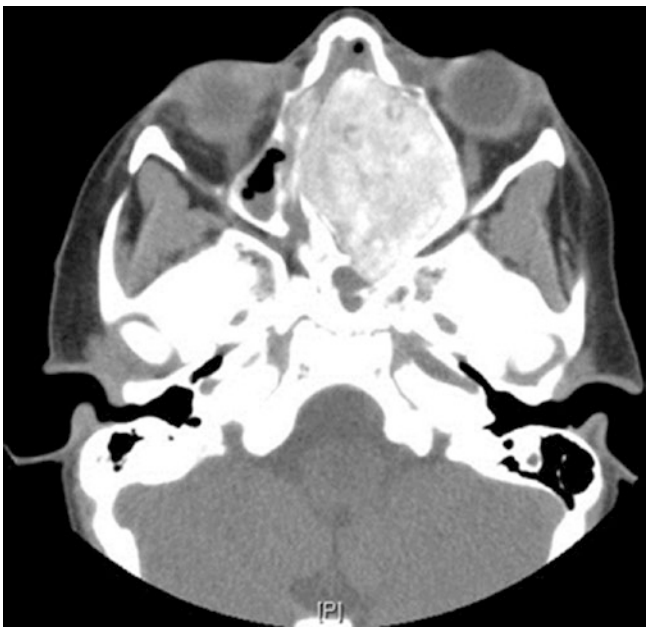

**Fig. 6.21** Axial CT typically demonstrates an expansible lesion that is relatively homogeneous in appearance, described as "ground glass"

The lesion may contain hypodense areas, cysts, or sclerosis, and the bony growth tends to expand with an intact cortex. The margin between abnormal and normal bone is often difficult to identify, the two regions blending with each other; however, a relatively sharp demarcation may be present and narrowing of neural foramina is rare. On MRI, appearance is variable depending on the degree of lucencies versus sclerosis, but there is typically a low to intermediate signal on T1-weighted images and a low signal on T2-weighted images, although where there are cartilaginous areas T2 signal will be bright. There may be heterogeneous gadolinium contrast enhancement.

### Management

Where asymptomatic, clinical and radiological follow-up is usually sufficient. The natural history is variable, and fibrous dysplasia may progress, particularly in the polyostotic form. Intervention is typically considered with the occurrence of significant disfigurement, compression causing cranial nerve impairment such as visual loss or pain. Medical therapy, such as bisphosphonates or corticosteroids, is of limited benefit, making surgery the mainstay of treatment. Malignant transformation of fibrosarcoma or osteosarcoma is described in approximately 0.4% [9]. Excision may be partial or total depending on the location and predicted morbidity, with sinonasal lesions potentially completely excised endoscopically, and skull base lesions often requiring a combined endoscopic/open approach. When the optic canal is involved in a sphenoid lesion, conservative management may be considered to avoid optic nerve injury if vision is stable, but where vision is deteriorating, an endoscopic partial decompression may be sufficient.

## Melanotic Neuroectodermal Tumor of Infancy

### Definition

Melanotic neuroectodermal tumor of infancy (MNTI) is a pigmented neoplasm typically arising in infants during the first year of life. In the majority of cases, MNTI originates from soft tissue overlying the maxilla, although involvement of the mandible, cranium, brain, and genitals have been described [10]. MNTI is a benign tumor of neural crest origin consisting of neuroblast-like cells and melanin-containing epithelial cells.

### Clinical Presentation

MNTI typically presents in a child under 6 months with a rapidly growing soft-tissue mass arising from the anterior alveolar maxillary ridge, which may be pigmented. The tumor causes feeding difficulty and can be locally aggressive with bony destruction, often extending upward into the maxilla.

### Differential Diagnosis

Other neoplasia of the newborn presenting in the maxillary location include congenital epulis and gingival cyst. The differential should include other small round cell tumors occurring of infancy, such as rhabdomyosarcoma, neuroblastoma, melanoma, desmoplastic small round cell tumor, primitive neuroectodermal tumor lymphoma.

### Radiological Features

Imaging is nondiagnostic but CT defines the extent of the lesion, clearly delineates osseous involvement and provides a good basis for surgical planning. It will typically demonstrate a hyperdense lesion with contrast enhancement, clear margins, and hyperostosis of adjacent bone. With MRI, the tumors are isointense or hypointense on T1- and T2-weighted MRI and will marked contrast enhancement in the non-ossified components.

### Management

The treatment of choice for MNTI is surgical excision. There remains controversy over the ideal margin required for this benign tumor, with suggested approaches ranging from curettage and enucleation to removal with a true margin [11]. Early conservative surgical excision with preservation of normal vital structures generally provides a good prognosis [11]. Both radiotherapy and chemotherapy are ineffective, including for recurrent or residual disease where further surgical resection would be indicated.

### Clinical Example

A 2-month-old baby girl was noted by mother to have a mass on the right alveolar arch prompting urgent referral. The mother felt the mass had been enlarging quickly over the past week or two. Examination showed the mass to be "spongy" in nature and nontender. There was a blackish discoloration seen through the intact mucosa, characteristic of MNTI. CT demonstrated an expansive lytic lesion. Direct excision was performed. All bony surfaces were "polished" intraoperatively with a burr under magnification. No recurrence was noted after 5-year follow-up (Figs. 6.22, 6.23a and b, 6.24 and 6.25).

## Nasal Xanthogranuloma

### Definition

Xanthogranuloma is an uncommon benign non-Langerhans cell histiocytosis lesion presenting in infancy, with a male preponderance.

### Clinical Presentation

The juvenile nasal form of xanthogranuloma usually presents as a solitary cutaneous asymptomatic soft lump on the surface of the nose. It may occur in other areas of the head and neck, such as the oral cavity or ear canal.

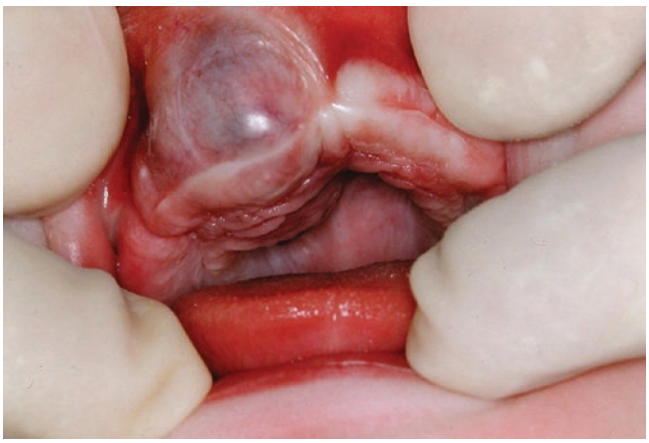

**Fig. 6.22** Expansile mass on the right alveolar arch with blackish hue seen through intact mucosa

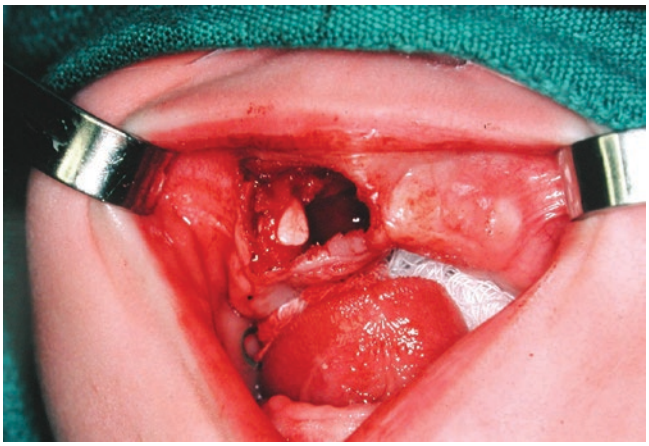

**Fig. 6.24** Defect after removal of tumor. All surfaces inspected and cleaned of potential tumor under endoscopic magnification. Tumor had characteristic black color making recognition of possible residual easy

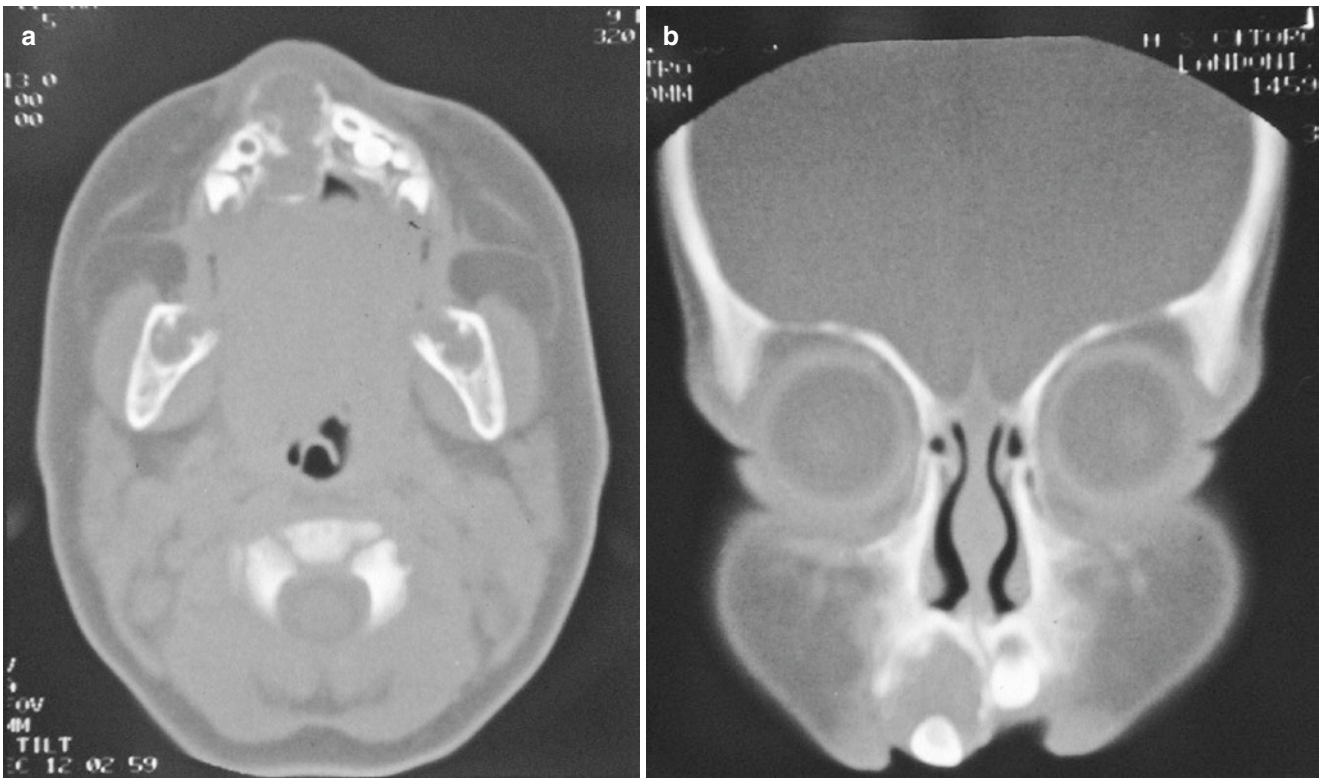


**Fig. 6.23** (**a**) Axial CT shows expansile lytic lesion of maxillary arch. (**b**) Coronal CT showing expansile mass displacing central incisor

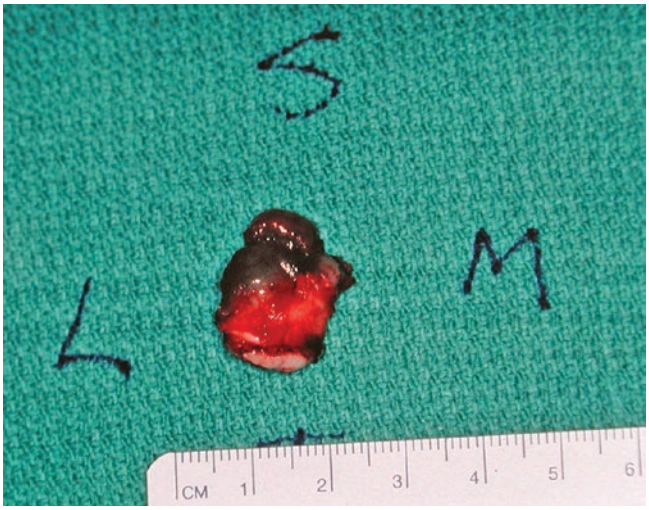


**Fig. 6.25** Gross tumor showing characteristic black color of MNTI

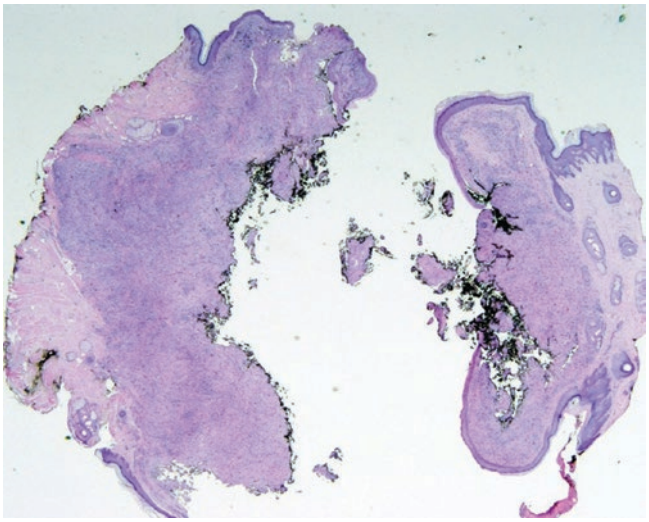

**Fig. 6.26** Simple excision of nasal lesion revealed xanthogranuloma

## Differential Diagnosis

The differential should include malignant soft tissue tumors such as rhabdomyosarcoma and malignant fibrous histiocytoma.

## Radiological Features

Radiological imaging is not generally required as the lesions are typically cutaneous.

## Management

Juvenile nasal xanthogranuloma may regress spontaneously, so conservative management may be indicated. Typically surgical excision by excisional biopsy is required for diagnosis and can provide good cosmesis.

## Clinical Examples

### Case 1

A 7-year-old boy with a past history of Crohn's disease was noted to have a right nasal vestibular mass for 2 months. No bleeding or any other nasal complaints were noted. Simple excision revealed xanthogranuloma. See Chap. 7 for complete description of pathology (Fig. 6.26).

### Case 2

A 1-month-old baby girl was noted to have a right nasal polyp since birth.

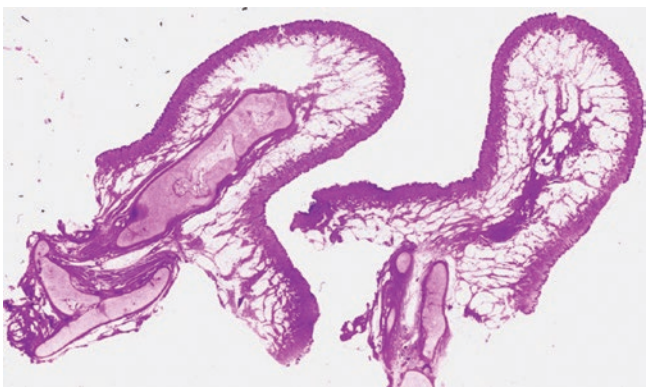

**Fig. 6.27** Simple excision of nasal vestibular lesion revealed lipoepidermal polyp

This was felt to be increasing in size slightly. The baby also had epibulbar dermoids and pre-auricular skin tags. A diagnosis of Goldenhar syndrome was also being considered. The nasal polyp was removed at surgery. The polyp was based on the right membranous and anterior cartilaginous septum and extended to the right upper lateral cartilage. Pathology revealed a lipoepidermal polyp. See Chap. 7 for full histological details (Fig. 6.27).

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