Orbital Tumors

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Tumors affecting the orbit include benign and malignant neoplasms arising from the various structures in the orbit including the blood vessels, nerves, bones, orbital fat, and other soft tissues. The first step in the management of orbital tumors is to differentiate the malignant tumors from the benign ones. A thorough clinical evaluation, aided by ancillary tests including imaging and biopsy, when necessary, can lead to an accurate diagnosis. Treatment of these tumors requires a basic knowledge of their biological behavior and their response to the currently available therapeutic modalities including surgery, chemotherapy, immunotherapy and radiotherapy. In the following chapter, some of the benign and malignant tumors of the orbit (Table 12.1) and their management are discussed.

12.1 Orbital Dermoid Cyst

Dermoid cyst is the most common orbital cystic tumor [1, 2]. They arise at the site of bony sutures secondary to the entrapment of the surface epithelium in the sutures during embryogenesis. The most frequent location is the superotemporal orbital rim (external angular dermoid), followed by the superonasal orbital rim (internal angular dermoid). In some instances, a dermoid cyst can have both extraorbital and intraorbital component connected through a defect in the bone, called as the dumbbell dermoid. Intraosseous dermoids can occur less frequently.

Anterior dermoids present as a well-circumscribed, firm, slow-growing subcutaneous mass (Fig. 12.1a–f). Dermoid cyst can also occur in the deeper orbital soft

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Table 12.1 Benign and malignant tumors of the orbit

Tumors of the orbit	Benign	Malignant
1. Cystic tumors	Orbital dermoid cyst Orbital teratomas Orbital mucocele	
2. Vascular tumors	Orbital capillary hemangioma Orbital cavernous hemangioma Orbital lymphangioma Orbital varix	Orbital hemangiopericytoma Orbital angiosarcoma Orbital glomus tumor
3. Peripheral nerve tumors	Orbital neurilemmoma Orbital neurofibroma Orbital paraganglioma	Orbital malignant peripheral nerve sheath tumor Orbital alveolar soft part sarcoma
4. Optic nerve and meningeal tumors	Optic nerve glioma Optic nerve sheath meningioma Orbital sphenoid wing meningioma	Optic nerve malignant astrocytoma Orbital neuroblastoma
5. Myogenic tumors	Orbital leiomyoma	Orbital malignant rhabdoid tumor Orbital rhabdomyosarcoma Orbital leiomysosarcoma
6. Fibrocytic tumors	Orbital fibroma Orbital solitary fibrous tumor Orbital myxofibroma	Orbital fibrosarcoma
7. Osseous and cartilaginous tumors	Orbital osteoma Orbital fibrous dysplasia Orbital ossifying fibroma Orbital giant cell reparative granuloma Orbital cartilaginous chondroma	Orbital osteosarcoma Orbital chondrosarcoma
8. Lipomatous tumors	Orbital dermolipoma Orbital lipoma	Orbital liposarcoma
9. Histiocytic tumors	Orbital juvenile xanthogranuloma Orbital Langerhans' cell histiocytosis Orbital Erdheim–Chester disease Orbital Rosai–Dorfman disease	
10. Lacrimal gland tumors	Lacrimal gland pleomorphic adenoma	Lacrimal gland pleomorphic adenocarcinoma Lacrimal gland adenoid cystic carcinoma
11. Lymphoid and leukemic tumors		Orbital non-Hodgkin's lymphoma Orbital plasmacytoma Orbital plasmablastic lymphoma Orbital Burkitt lymphoma
12. Metastatic tumors		
13. Secondary tumors		

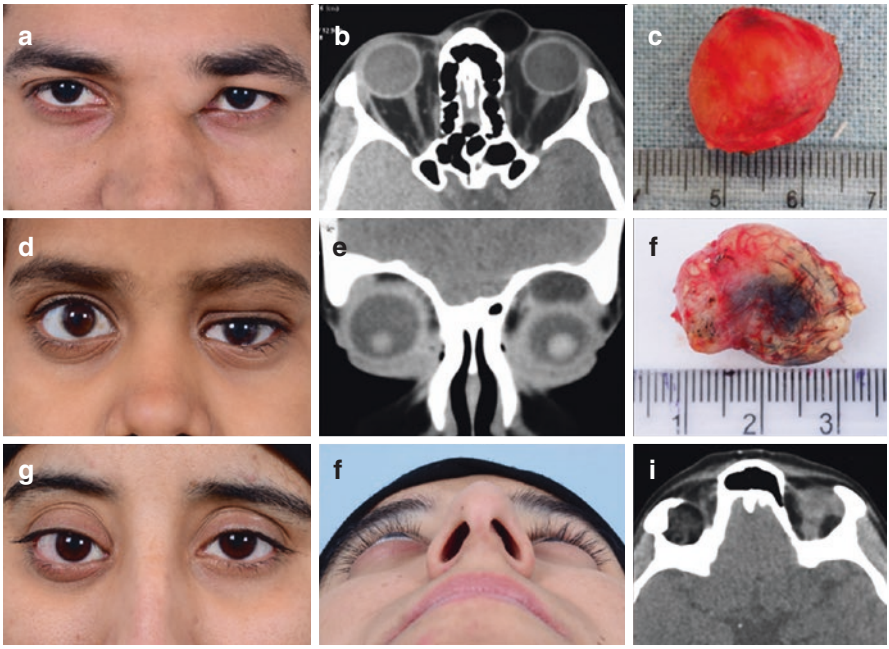

Fig. 12.1 Orbital dermoid cyst. (**a**) A 28-year-old male with an internal angular dermoid left orbit. (**b**) Axial CT orbit showing a cystic lesion in the left superomedial orbit. (**c**) Gross appearance of the intact dermoid of the same patient showing a well-circumscribed encapsulated lesion. (**d**) A 10-year-old female with a large dermoid in the superior left orbit. (**e**) Coronal CT orbit demonstrates downward displacement of the left globe by the cystic lesion (hypoglobus). (**f**) Gross appearance of the large dermoid with presence of hair on the tumor surface. (**g**) A 19-year-old female with right hypoglobus. (**h**) Worm's eye view shows right proptosis. (**i**) Axial CT orbit reveals the presence of a large deep orbital cystic lesion

tissues which causes proptosis (Fig. 12.1g–i). Sometimes, superficial dermoids rupture spontaneously or after trauma and produce an inflammatory reaction resembling cellulitis or dacryoadenitis. Rarely, squamous cell carcinoma may arise within these cysts.

The classic dermoid cyst can usually be diagnosed based on the clinical appearance of a subcutaneous firm mass located at the orbital rim. A computed tomography scan is preferred to assess any associated bony defects. It shows a cystic lesion with enhancement of the wall, but no significant enhancement of the lumen [2]. A bony defect is evident in dumbbell dermoid, and fluid levels and calcification are frequently seen. Histopathologically, an orbital dermoid cyst is lined by surface epithelium [2]. The cyst wall contains dermal appendages including sebaceous glands and sweat glands. The cyst lumen contains putty-like material composed of desquamated epithelial cells, sebaceous material, and hair.

The management of orbital dermoid cyst depends on the size, location, and symptoms. Asymptomatic small dermoids can be simply observed, while symptomatic large ones require complete surgical excision. Most anterior dermoid cysts can

be excised by anterior orbitotomy. Deeper and larger cysts require a lateral orbitotomy. Care should be taken to avoid surgical rupture of the cyst as its contents can incite an inflammatory reaction. If rupture occurs, copious irrigation and instillation of antibiotics or corticosteroids is advised.

12.2 Orbital Teratomatous Cyst

Teratoma is a congenital, multicystic mass arising from the primitive pluripotent germ cells that contains histologic structures representing all three embryonic germ layers: ectoderm, mesoderm, and endoderm [3]. Orbital teratoma is an uncommon condition that is almost always unilateral. Teratomas of the orbit are generally benign, and presents with severe unilateral proptosis at birth. The proptosis may increase over the first few days, and the resultant lid swelling and globe compression can lead to visual loss and corneal exposure. Larger lesions cause severe orbital disfigurement. The tumor can invade into the adjacent orbital tissue and extend posteriorly into the temporal fossa. Orbital teratoma should be ruled out in any neonate presenting with a large orbital mass.

On imaging of a teratoma, the scans demonstrate an enlarged orbit with a multiloculated mass. Histopathologically, a teratoma typically contains clear cysts lined by either epidermis or mucosal epithelium. A variety of tissues can be seen within the tumor including cerebral tissue, hyaline cartilage, choroid plexus, or rarely, well-differentiated tissues resembling a complete fetus or a portion of a fetus in the orbit [4].

Orbital exenteration is generally done in advanced cases with orbital disfigurement. Smaller orbital teratomas can be removed surgically, with preservation of the eye [5]. Aspiration of the fluid from a larger cyst to decrease the size of the mass allows complete surgical excision [6].

12.3 Orbital Mucocele

Orbital mucocele is an expansile lesion resulting from accumulation of mucoid secretion of a chronically inflamed paranasal sinus secondary to obstruction of the sinus ostium. It is often painless and demonstrates a fairly rapid growth. When a mucocele becomes secondarily infected, it is called a mucopyocele. It generally occurs in adults, but it is sometimes seen in children in association with cystic fibrosis. Most commonly, frontal sinus is involved (60–90%) which produces progressive proptosis that tends to displace the globe downward (Fig. 12.2a–d) [7]. Ethmoidal mucocele is less common, and maxillary and sphenoidal sinus involvement is very rare. The mass is generally visible and it is fluctuant to palpation beneath the orbital rim.

On imaging, a mucocele demonstrates opacification of the affected sinus and thickening of the mucosal lining, with erosion of the adjacent orbital bones [8]. Histopathologically, a mucocele is lined by the sinus mucosa, pseudostratified

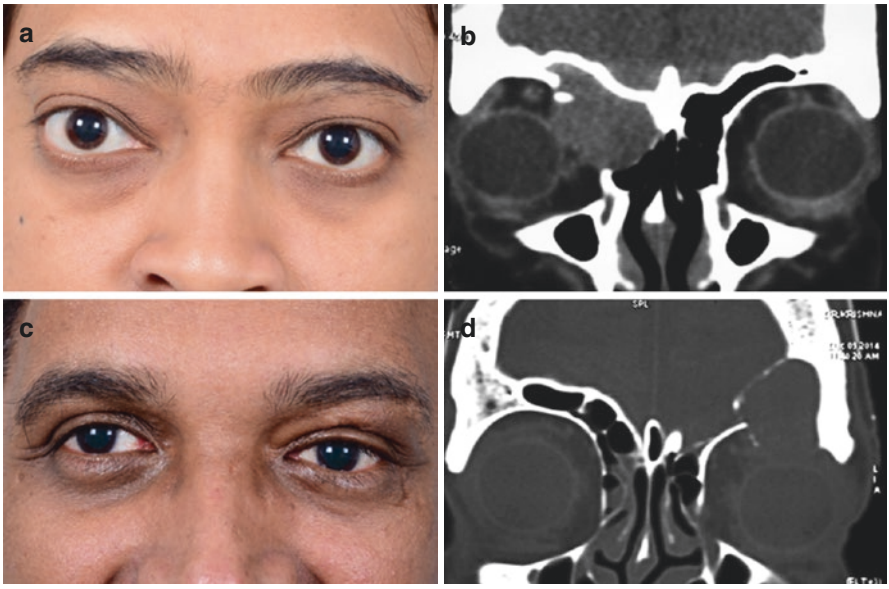

Fig. 12.2 Orbital mucocele. (**a**) A 32-year-old female with right proptosis. (**b**) Coronal CT orbit reveals an irregular homogenous lesion in the superomedial orbit with opacification of both ethmoid and frontal sinuses. (**c**) A 45-year-old man with left hypoglobus. (**d**) Coronal CT orbit shows left superior mass lesion with complete opacification of the left frontal sinus

columnar epithelium, with infiltration by the chronic inflammatory cells. The lumen contains mucous or pus.

The management of mucocele is complete surgical excision of the mucosal wall with marsupialization to avoid recurrence. Antibiotics to cover both aerobic and anaerobic bacteria should be administered before and after surgery.

12.4 Orbital Capillary Hemangioma

Periocular capillary hemangioma of infancy (strawberry hemangioma) can be superficial (located anterior to the orbital septum) or deep (located posterior to the septum). About 7% of periocular capillary hemangiomas arise posterior to the orbital septum [9]. Regarding its etiopathogenesis, due to the similar immunohistochemical characteristics with the placenta, it is believed that infantile hemangiomas could be of placental origin [10, 11]. Typically, capillary hemangiomas are not present at birth, but develop in the first few months of life and continue to enlarge over the first 6–12 months after the first year (proliferative phase), with 90% resolution occurring within 8 years of life (involutional phase). When located deeper within the orbit, it presents as progressive proptosis, which becomes more pronounced when the child strains or cries (Fig. 12.3a, b). In a child with proptosis, the diagnosis of orbital capillary hemangioma is more evident when associated with hemangioma

Fig. 12.3 Orbital capillary hemangioma. (**a**) A 19-month-old male with right upper lid fullness and mild right proptosis. (**b**) With crying, the fullness in the right upper lid increases with a prominent bulge in the right temporal fossa. (**c**) Coronal T2-weighted post-contrast MRI orbit reveals the presence of a diffuse mass in the deep right orbit which enhances well with contrast

of the eyelid. Lesions greater than 1 cm in diameter are more likely to cause complications, with an incidence of amblyopia upto 60% [9, 12]. Periocular capillary hemangioma of infancy may be seen in association with Kasabach–Merritt syndrome, which is characterized by large visceral hemangiomas, platelet entrapment, and thrombocytopenia.

On imaging, orbital capillary hemangiomas appear as a well-circumscribed homogenous soft tissue mass without adjacent bone destruction (Fig. 12.3c). With the use of contrast, the lesion shows enhancement. Histopathologically, an orbital capillary hemangioma is composed of lobules of proliferating small endothelially lined vascular spaces separated by thin fibrous septa.

Most tumors can be managed by observation, although those causing amblyopia should be treated with refraction and occlusive patching. Oral use or local injection of corticosteroids can hasten regression of the lesion. Oral prednisolone 2–4 mg/kg/day for 2–4 weeks is administered under the supervision of a pediatrician. The major risks include adrenal suppression and growth retardation. Intralesional corticosteroids are administered as a combination of triamcinolone 1 mL (40 mg/mL) and dexamethasone 1 mL (4 mg/mL). Interferon α-2a upto three million units/m^2 of body surface area can be given as daily subcutaneous injections for life-threatening or vision-threatening hemangiomas to cause complete regression of the lesions [13–15]. Propranolol is a nonselective beta blocker that can be used systemically for orbital capillary hemangiomas with high efficacy [16]. The recommended dosage of oral propranolol is 2–3 mg/kg/day until regression and additionally for a month to prevent recurrence. Surgical treatment is rarely necessary, but can be considered in those with visual symptoms not responding to pharmacologic modalities.

12.5 Orbital Schwannoma

Schwannoma (neurilemoma) is a benign, encapsulated tumor that arises from the cells forming the peripheral nerve sheath (Schwann cells). It develops as an eccentric growth from the nerves and causes proptosis (Fig. 12.4a–d). They are usually solitary lesions, and occur between the ages of 20 and 50 years [17]. Neurilemmomas

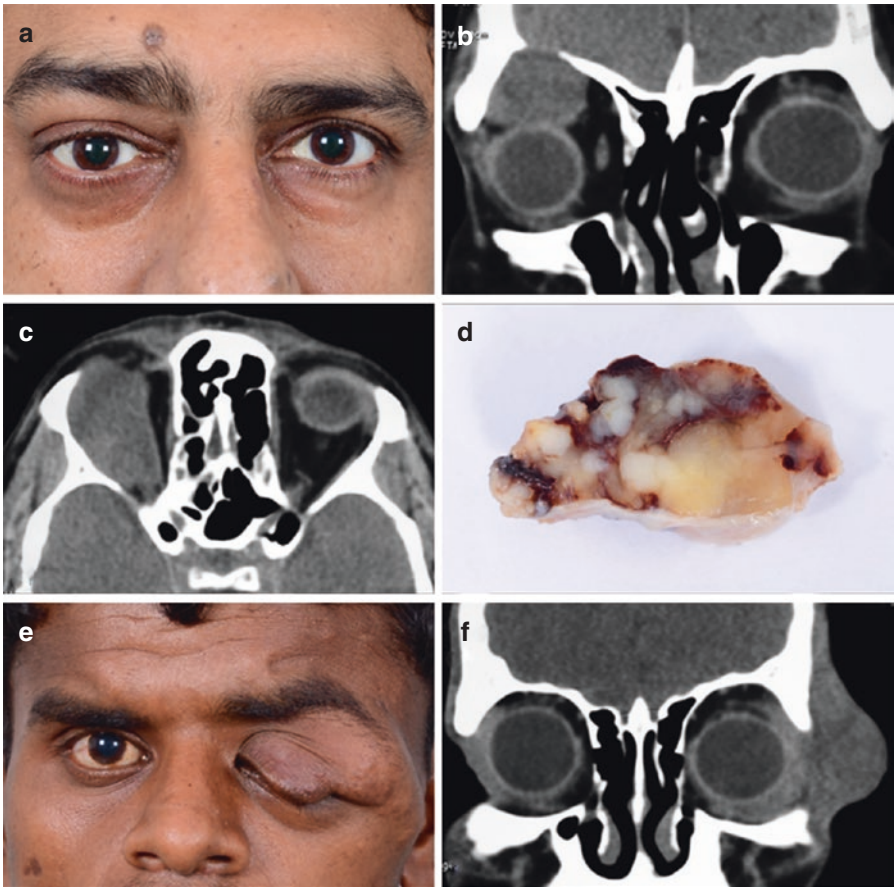

Fig. 12.4 Orbital peripheral nerve tumors. (**a**) A 48-year-old male with right proptosis. (**b**) Coronal CT orbit shows right superior orbital homogenous lesion. (**c**) Axial CT orbit shows an irregular homogenous mass extending upto the posterior orbit. (**d**) Gross appearance of an orbital schwannoma, multi-lobulated with a capsule. (**e**) A 20-year-old male with left orbital plexiform neurofibromatosis. (**f**) Coronal CT orbit illustrates left temporal fossa homogenous mass lesion with minimal orbital involvement

usually form along the course of the supraorbital or supratrochlear nerve, and less frequently along the infraorbital nerve.

On imaging, a neurilemmoma appears as an ovoid to elongated extraconal mass, although intraconal neurilemmomas also occur. Large cystoid spaces are sometimes present within the lesion. On histopathology, a schwannoma consists of benign proliferating Schwann cells, with Antoni A pattern (ribbons or fascicles of spindle cells) interspersed with Antoni B pattern (ovoid clear cells).

Surgical excision offers complete cure. Recurrences are common when the tumor is incompletely excised [21]. However, if the tumor has an extension upto the orbital

apex, the apical component is best left alone to avoid injury to the structures passing through the orbital apex and superior orbital fissure.

12.6 Orbital Neurofibroma

Neurofibromas in the orbit can occur in three different forms-as a solitary tumor, as a diffuse infiltration, and as a plexiform lesion. Isolated, solitary neurofibromas are generally not associated with neurofibromatosis. Diffuse neurofibromas occur as a part of the systemic syndrome in only 10% of patients [18]. On the other hand, plexiform lesions are typically suggestive of neurofibromatosis type 1. Patients with neurofibromatosis can have congenital absence of the sphenoid bone that can produce a pulsating proptosis.

Localized neurofibroma are solitary well-circumscribed tumors within the orbit that can produce proptosis and optic nerve compression. Diffuse and plexiform neurofibromas are seen in the first decade of life and cause the typical S-shaped curve to the upper eyelid due to the subcutaneous involvement, but the plexiform variant can cause severe disfigurement and morbidity secondary to involvement of the orbit, eyelids, intraocular structures, and maxillofacial region (Fig. 12.4e, f).

On imaging, a neurofibroma appears as a homogenous mass that can be either well-circumscribed or diffuse. Plexiform neurofibromas additionally show periorbital involvement. Histopathologically, it consists of Schwann cells and endoneural fibroblasts with distinct perineural sheath separating the axons within the involved nerve.

Symptomatic solitary tumors are managed by complete excision. Diffuse and plexiform lesions cannot be completely excised and hence, debulking is done to offer relief from compressive symptoms or for cosmesis.

12.7 Optic Nerve Glioma

Optic nerve gliomas are low-grade pilocytic (juvenile) astrocytomas that resemble those which occur elsewhere in the central nervous system. Although most of these are isolated lesions, an association with neurofibromatosis type1 (NF1) must be looked for, as the clinical behavior is different in this context. Approximately, 10% of patients with NF1 have optic pathway gliomas, and 30% of patients presenting with optic nerve gliomas have NF1 [19]. An optic nerve glioma in the background of NF1 offers a favorable prognosis [20]. Most lesions present in the first decade of life with a female preponderance. At the time of diagnosis, gliomas frequently involve the chiasma. Clinically, the usual presentations are proptosis, visual loss, motility restriction, optic atrophy, and unilateral disc edema. These tumors show an indolent growth or are non-progressive, with little or no change in the clinical status over long periods [21].

On imaging, optic nerve glioma has a characteristic ovoid appearance along the course of the optic nerve (Fig. 12.5a, b). A characteristic kink in the midportion of

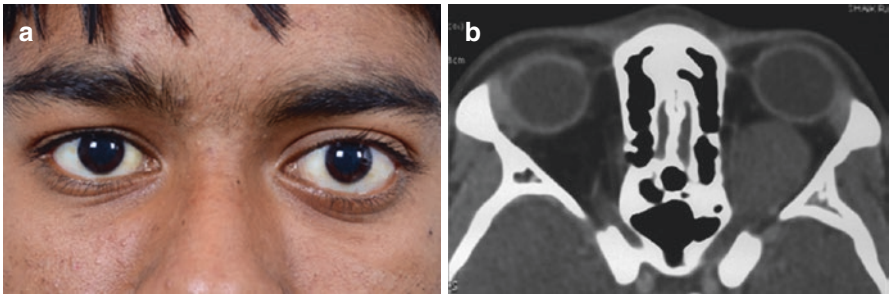

Fig. 12.5 Optic nerve glioma. (**a**) An 18-year-old male with left axial proptosis. (**b**) Axial CT orbit shows a homogenous ovoid mass along the course of the optic nerve extending upto the posterior orbit

the tumor is often present. Gadolinium-enhanced MRI is the best modality for diagnosis, monitoring disease progression, and response to treatment. Histopathologically, the tumor is composed of fibrillary astrocytes, which may develop many brightly eosinophilic nodes (called Rosenthal fibers) in their processes. Rarely, malignant degenerative features or mitoses are observed.

The general principle of management is essentially conservative. The patient is followed up periodically for vision, pupil, fundus, and visual field examination, while the tumor is monitored by imaging. Treatment is recommended in tumors which are either large or rapidly progressive. Patients in the younger age group are known to have a higher chance for progression [22]. In children less than 10 years of age, chemotherapy is the treatment of choice. In older children and adults, excision is performed when the tumor is still confined in the prechiasmatic optic nerve. With significant chiasmal or parachiasmal involvement and progressive growth, radiotherapy is treatment of choice [23].

12.8 Orbital Meningioma

Meningiomas affecting the ocular and orbital structures can arise from the optic nerve sheath or intracranially. Majority of the meningiomas involving the orbit are extensions from intracranial sites, most commonly the sphenoid wing. True primary optic nerve sheath meningiomas are rare. About 90% of the primary optic nerve sheath meningiomas originate within the intraorbital nerve sheath, while less than 10% are intracanalicular in origin [24].

Optic nerve sheath meningiomas typically affect middle-aged women (Fig. 12.6a, b). Bilateral and multifocal optic nerve sheath meningiomas are more likely to present in childhood in association with neurofibromatosis type 2 [25]. Visual loss is often seen, especially from tumors of the optic canal which can cause early visual disturbances. The classical triad of progressive visual loss, optic nerve atrophy, and presence of opticocilliary shunt vessels is pathognomonic of an optic nerve sheath meningiomas. Frequently, there is evidence of an underlying optic neuropathy, as

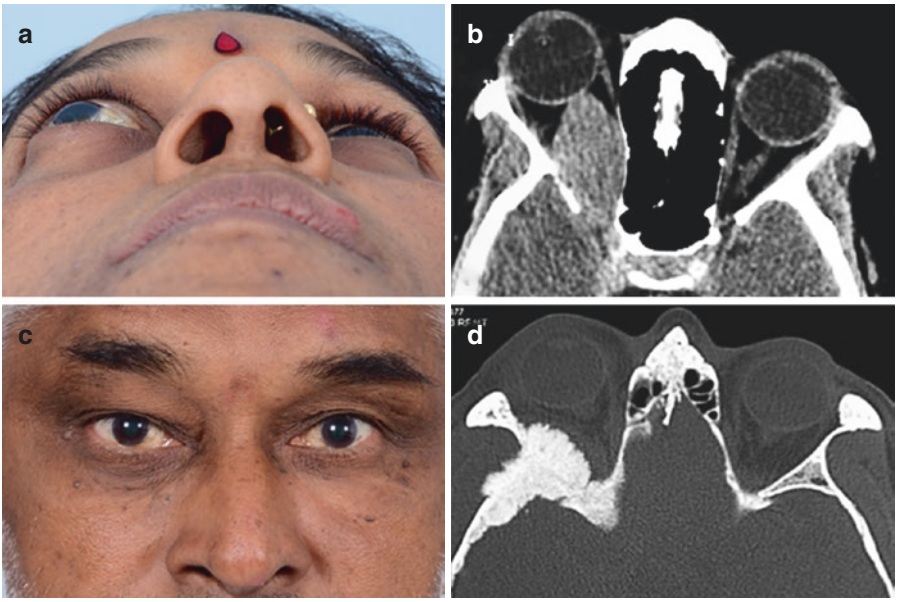

Fig. 12.6 Orbital meningioma. (**a**) A 46-year-old female with Worm's eye view demonstrates left proptosis. (**b**) Axial CT orbit shows a homogenous fusiform mass along the course of the optic nerve extending with optic foramen widening. (**c**) A 52-year-old male with right proptosis and bulging of right temporal fossa. (**d**) Axial CT orbit shows left sphenoidal wing hyperostosis

also peripapillary hemorrhages and disc swelling. Proptosis is present in most patients. Sphenoid wing meningioma arises from the arachnoid lining the sphenoid bones and secondarily invades the orbit. Like optic nerve sheath meningioma, the sphenoidal wing meningioma is also seen in middle-aged women. It presents as a slowly progressive abaxial proptosis, enlargement of the temporal fossa, and cranial nerve palsies (Fig. 12.6c, d). Visual disturbances occur late in the disease when the tumor compresses the optic nerve at the optic canal.

On imaging, optic nerve sheath meningiomas show fusiform enlargement in the arachnoid with a relatively normal optic nerve in its center. Areas of calcification are often seen in the tumors. On the other hand, a sphenoidal wing meningioma causes the characteristic bony hyperostosis, with a soft tissue tumor mass extending into the orbit, temporal fossa, and cranial cavity [25–27]. Histopathologically, meningiomas arise from meningothelial cap cells of the arachnoid with occasionally dispersed calcification (psammoma bodies).

For optic nerve sheath meningiomas, observation alone suffices where there is no significant visual deterioration, especially when the tumor is located at the orbital apex. Surgical resection is offered to patients with aggressive tumors with intracranial extension, in order to prevent spread to the contralateral optic nerve. The

surgical approach is either by a lateral orbitotomy, or via transcranial route by a neurosurgeon. Various new techniques of conformal radiotherapy are now used in the treatment of optic nerve sheath meningiomas including intensity-modulated radiotherapy (IMRT) and stereotactic fractionated radiotherapy (SFRT) [27]. Cyberknife surgery and proton beam therapy are now increasingly being used with good tumor control. Meningioma of the sphenoid wing is managed by surgical debulking in conjunction with a neurosurgical team, and subsequent postoperative radiation for the residual disease.

12.9 Orbital Rhabdomyosarcoma

Rhabdomyosarcoma is the most common primary orbital malignant tumor in the pediatric age group. Orbital rhabdomyosarcoma is primarily a disease of young children with a mean age at diagnosis of 10 years [28]. The presenting features include proptosis, globe displacement, ptosis, conjunctival and eyelid swelling, and a palpable orbital mass (Fig. 12.7a, b). Rhabdomyosarcoma is an aggressive neoplasm that can invade orbital bone and even extend into the cranial cavity. Metastasis most commonly occurs to lung and bone via hematogenous dissemination.

On imaging, rhabdomyosarcoma appears as a moderately well-circumscribed orbital mass that is limited to soft tissue without the involvement of extraocular muscles. Less frequently, it can erode into the adjacent orbital bones or sinuses. The tumor shows enhancement with contrast material. Histopathologically, the tumor is composed of cells that resemble histologically to striated muscle in various stages of embryogenesis. Four different histopathological types can be seen, and embryonal rhabdomyosarcoma is the most common variant in the orbit.

The management of orbital rhabdomyosarcoma has evolved over the last two decades. A multimodal management including a combination of surgery, irradiation, and chemotherapy is shown to give excellent results. Intergroup Rhabdomyosarcoma Study Group IV currently recommends a specific regimen

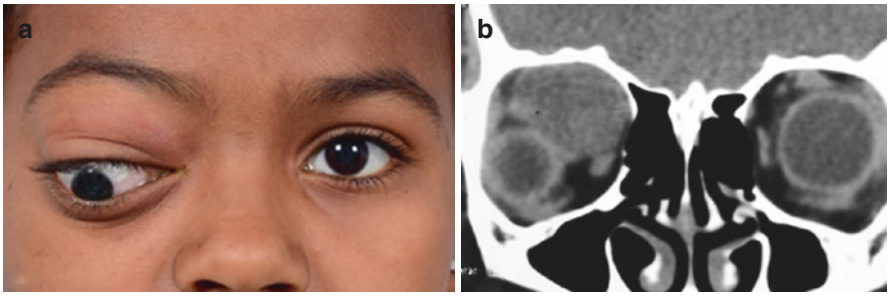

Fig. 12.7 Orbital rhabdomyosarcoma. (**a**) A 10-year-old female with right hypoglobus. (**b**) Coronal CT shows a large right superomedial orbital lesion displacing the globe downward and outward

including a combination of radiation and chemotherapy based on the tumor staging. An incisional biopsy is first performed to establish a histopathologic diagnosis, followed by chemotherapy consisting of vincristine, etoposide, cyclophosphamide/and ifosfamide [29–31]. Radiation is generally sandwiched between chemotherapy cycles, as deemed appropriate by the treating radiation oncologist.

12.10 Orbital Dermolipoma

Dermolipoma is a congenital lesion that is often detected in adulthood as the lesion tends to be very small in children. It is a choristoma and is seen as a pink-yellow mass in the superotemporal conjunctival fornix. It is a sessile, soft to firm mass, most commonly unilateral. It accounts for 3% of all orbital lesions [1]. Fine hair is sometimes found on the tumor surface (Fig. 12.8a, b). Dermolipoma is sometimes seen in association with Goldenhar syndrome.

On imaging, a dermolipoma appears as a well-circumscribed lesion, with its posterior border not very distinct from the superotemporal orbital fat and the lacrimal gland. Histopathologically, a dermolipoma is lined by stratified squamous epithelium, made up of mature fat, pilosebaceous units, and glandular acini.

Dermolipoma is managed by observation when small and asymptomatic. Larger tumors are excised by a conjunctival approach by unroofing the lesion, with care taken to avoid injury to the lacrimal ducts and levator aponeurosis. After excision of the lesion, the conjunctiva overlying it can be easily reconstructed.

12.11 Lacrimal Gland Pleomorphic Adenoma

Pleomorphic adenoma (benign mixed tumor) accounts for accounts for 50% of all the epithelial tumors of the lacrimal gland. It is a disease of the young age, and it usually arises from the orbital lobe of the lacrimal gland. Pleomorphic adenoma

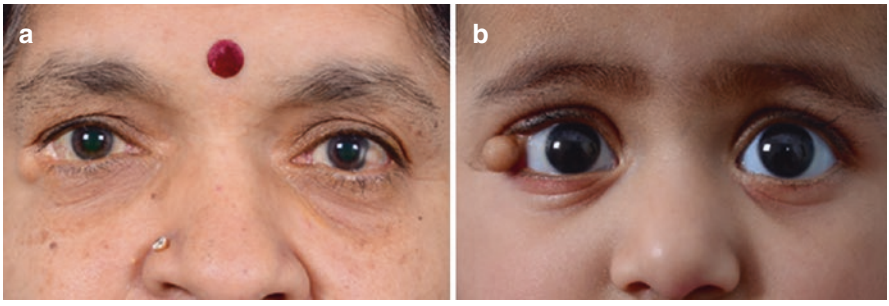

Fig. 12.8 Orbital dermolipoma. (**a**) A 63-year-old female with right dermolipoma, right lateral canthal cutaneous choristoma, left conjunctival dermoid (patient also had preauricular skin tags, all features suggestive of Goldenhar syndrome). (**b**) A 2-year-old female with a large right dermolipoma with fine hair on the tumor surface

presents as a unilateral progressive mass in the superotemporal orbit. It is generally painless and produces proptosis as it enlarges and grows into the orbit posteriorly. It causes downward displacement of eyeball and lid swelling (Fig. 12.9a, b). Rarely, the tumor demonstrates malignant transformation (pleomorphic adenocarcinoma).

On imaging, the tumor is seen as a well-circumscribed mass in the lacrimal gland fossa with an irregular surface. There is generally an associated pressure indentation of the lacrimal gland fossa, but without erosion of the orbital bones. The histopathology of pleomorphic adenoma reveals a combination of benign epithelial elements and mesenchymal elements, consisting of ducts, squamous cells, myxoid material, and cartilagenous tissue.

The diagnosis of pleomorphic adenoma of the lacrimal gland can be generally made based on the clinical and radiological findings. When the diagnosis is evident, an incisional biopsy is best avoided to avoid breaching the capsule. The tumor is excised with an intact capsule by a superolateral orbitotomy through an eyelid crease or a sub-brow incision with a trans-septal approach.

12.12 Adenoid Cystic Carcinoma of the Lacrimal Gland

Adenoid cystic carcinoma of the lacrimal gland is the most common malignant epithelial neoplasm of the lacrimal gland. The age at diagnosis has a bimodal pattern, with patients generally belonging to the first to second decade or the sixth to seventh decade. Periocular pain is frequently reported due to perineural invasion by the tumor. The tumor causes abaxial proptosis, with progressive downward and medial displacement of the globe (Fig. 12.9c, d).

Generally, CT demonstrates a round or elongated soft tissue mass with bone erosion, and moderate contrast enhancement. Histopathologically, the tumor is characterized by solid areas or cords of bland-appearing malignant epithelial cells arranged in various patterns. The typical pattern with cystic spaces lined by malignant cells is called the "Swiss cheese" pattern. Adenoid cystic carcinoma of the lacrimal gland has been subdivided into several histopathologic subtypes, namely cribriform, sclerosing, basaloid, comedocarcinoma, and tubular types [32].

Lacrimal gland adenocarcinoma is known to carry a dismal prognosis with fatality up to 60–80%. The tumor has a tendency for perineural infiltration and bone invasion, making it difficult to achieve surgically clear margins. Furthermore, distant metastasis occurs in about half of the patients. The rarity of this cancer makes it difficult to study the efficacy of various therapeutic modalities. Orbital exenteration is the widely accepted form of treatment. Neoadjuvant chemotherapy by the intra-arterial route or as systemic therapy has also been tried as a part of multimodal management [33]. Adjuvant radiotherapy for lacrimal gland carcinomas is indicated in all cases. Stereotactic radiotherapy, intensity-modulated radiation therapy (IMRT) technique, 3D conformal radiotherapy or proton radiotherapy deliver radiation more efficiently to the tumor site and reduce side effects to the normal orbital structures [34]. For those with a limited disease extent, tumor excision through lateral orbitotomy followed by adjuvant radiotherapy has achieved eye and vision salvage.

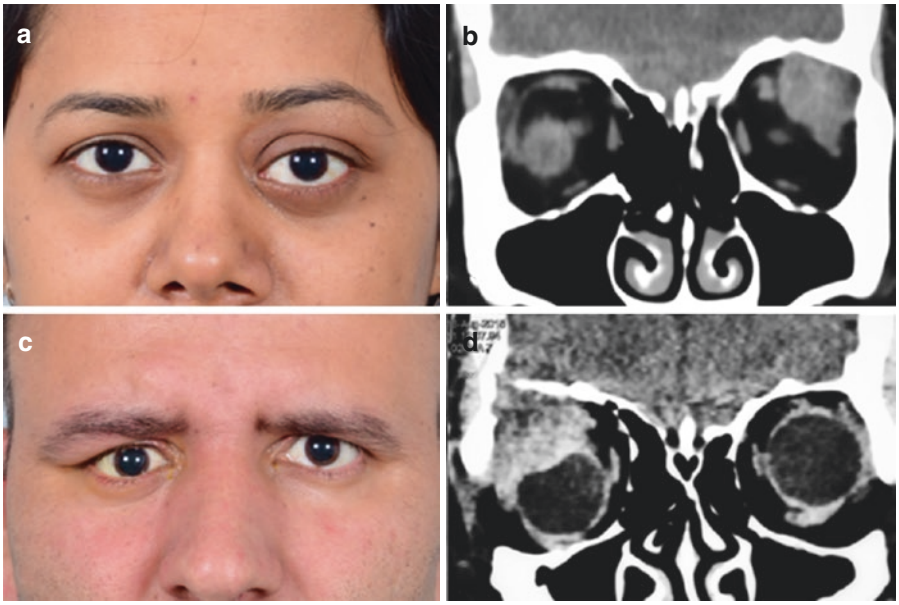

Fig. 12.9 Lacrimal gland tumors. (**a**) An 18-year-old female with left proptosis. (**b**) Coronal CT orbit reveals a left supertemporal homogenous orbital mass in the lacrimal fossa, with bony indentation and no bone destruction. (**c**) A 38-year-old male with right proptosis complaining of severe diplopia in right lateral gaze. (**d**) Coronal CT orbit shows a nonhomogenous ill-defined mass lesion in the right lacrimal fossa with evidence of bone destruction

12.13 Orbital Lymphoma

Lymphomas of the ocular adnexa constitute approximately 8–10% of all extranodal lymphomas. Orbital lymphoma is the most common orbital lymphoproliferative lesion, which form a spectrum ranging from reactive lymphoid hyperplasia to lymphomas. These lesions can involve the conjunctiva, eyelids, lacrimal glands, extraocular muscles, orbital soft tissue, or lacrimal sac, either individually or in combination.

Orbital lymphoma is a disease that occurs in the middle-age and the elderly. It is most commonly unilateral, although bilateral involvement is also known. Conjunctival lymphoma presents as a well-circumscribed "salmon-patch," while other orbital lymphomas most often cause proptosis or an eyelid swelling. Orbital involvement may present as an enlarged lacrimal gland, an enlarged extraocular muscle, or as a diffuse extraconal lesion (Fig. 12.10a–d). Most orbital lymphomas are primary tumors and are usually non-Hodgkin's lymphoma (NHL) of B-cell type. The most common primary lymphoma subtype is the low-grade malignant extranodal marginal zone B-cell lymphoma of MALT type (mucosa-associated lymphoid tissue). In those presenting with orbital lymphoma alone, systemic lymphoma eventually develops in one-thirds by 10 years, whereas only about 5% of patients with NHL develop ocular adnexal involvement during the course of their disease [35, 36].

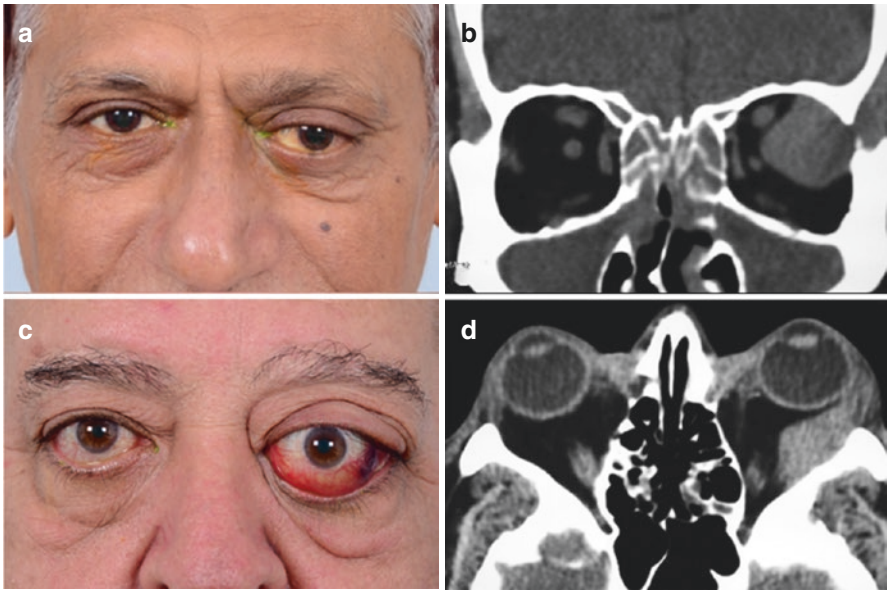

Fig. 12.10 Orbital lymphoma. (**a**) A 59-year-old male with left proptosis. (**b**) Coronal CT orbit reveals a left supertemporal orbital mass in the lacrimal fossa, with bony indentation and no bone destruction. (**c**) A 60-year-old male with left proptosis and conjunctival injection. (**d**) Axial CT orbit shows a mass lesion along the lateral rectus muscle extending upto the mid-orbit

A thorough systemic evaluation is warranted in all patients at the time of presentation. An incisional biopsy for a histopathological confirmation of the disease is mandatory. If the patient has associated systemic lymphoma, chemotherapy is recommended. Various regimens with or without immunotherapy are now available. In the absence of systemic lymphoma, small and well-circumscribed lesions are amenable to complete resection. Intralesional rituximab offers some benefit in the resolution of small orbital lesions. For larger lesions, a low dose radiotherapy offers an excellent local control with prolonged regression [37, 38]. Intravenous rituximab infusion followed by 90Y ibritumomab tiuxetan has also proven to be effective in early stage extranodal indolent B-cell lymphoma of the ocular adnexa [39].

12.14 Orbital Metastatic Tumors

Metastasis to the orbit occurs mainly by the hematogenous route. The primary tumor causing metastasis to the orbit is different in adults and children. In adults, the most common primary tumors metastasizing to the orbit are carcinomas of the breast, prostate gland, lung, and gastrointestinal tract [40]. Rarely, cutaneous and uveal melanoma can metastasize to the orbit. In children, orbital metastases occur from adrenal neuroblastoma, Wilms tumor, and Ewing tumor [40].

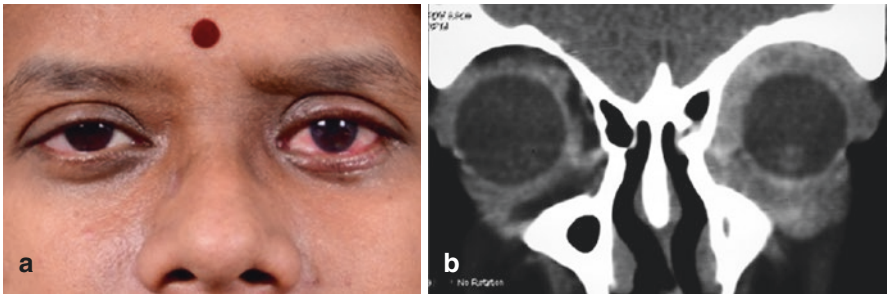

Fig. 12.11 Orbital metastatic tumors. (**a**) A 56-year-old female with a history of breast carcinoma post treatment presenting with left inferior orbital mass. (**b**) Coronal CT orbit shows a diffuse mass in the left orbit and the biopsy proved to be metastatic carcinoma from breast

The presenting features of orbital metastasis depend on the exact location of the tumor in the orbit and the type of primary neoplasm. Some of these lesions are confined to the extraocular muscles while others occur as solitary intraconal or extraconal mass producing rapidly progressive proptosis, pain, diplopia, ptosis, and eyelid swelling (Fig. 12.11a, b). Enophthalmos is seen in tumors which cause fibrosis of the orbital tissues, namely the scirrhous carcinoma of the breast and stomach [41, 42].

A thorough history, systemic examination and whole-body imaging is undertaken to diagnose the primary tumor. Imaging of the orbit for the size and location of the tumor is mandated. The diagnosis should be confirmed by an orbital incisional biopsy. The histopathology of orbital metastasis is same as that of the primary neoplasm. Once the orbital tumor is proven to be a metastasis, further management should involve a team of medical and surgical oncologists. The orbital tumor may subsequently require radiation therapy.

12.15 Orbital Secondary Tumors

An orbital secondary tumor is a malignant tumor that has extended into the orbit from the adjacent tissues around the orbit including eyelid, conjunctiva, intraocular structures, sinuses, nasopharynx, and brain. The main eyelid tumors that can involve the orbit secondarily include sebaceous gland carcinoma, squamous cell carcinoma, and basal cell carcinoma. Conjunctival tumors extending into the orbit include squamous cell carcinoma and melanoma (Fig. 12.12a, b). Intraocular tumors include uveal melanoma and retinoblastoma. Sinus tumors like carcinoma of the ethmoid or maxillary sinus and intracranial tumors like sphenoid wing meningioma also invade the orbit.

The clinical features of a secondary orbital tumor depend on the type and location of the primary neoplasm, with proptosis being the most common symptoms. Sometimes, the tumor is detected accidentally during follow-up for a previous eye and ocular adnexal tumor.

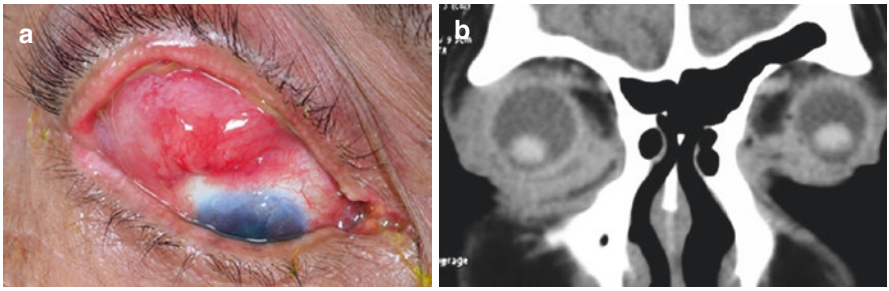

Fig. 12.12 Orbital secondary tumors. (**a**) A 59-year-old male with recurrent conjunctival squamous cell carcinoma right eye. (**b**) Coronal CT orbit shows diffuse right orbital involvement by extension of the conjunctival tumor

A detailed history, clinical exam, imaging, and an orbital biopsy are necessary to establish the diagnosis. Management depends on the extent of the lesion in the orbit. Smaller tumors may be managed by excision biopsy, while orbital exenteration is necessary for larger tumors. Subsequent chemotherapy and radiation therapy are essential in many cases.

In conclusion, orbit is an important site for a variety of primary and secondary tumors. Common tumors of the orbit maybe distinguished in most cases due to their characteristic clinical and radiological features. However, common tumors with atypical presentation or rare tumors of the orbit can cause a diagnostic dilemma. A well-planned biopsy of the tumor can yield correct diagnosis in such cases. Treatment of benign tumors is mainly observation or surgery, while malignant tumors are managed by using one or more of the various modalities of treatment available, as deemed appropriate by the treating ocular oncologist in conjunction with a medical oncologist.

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