Primary Cranio-Orbital Bone Tumors

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15.1 Introduction

Primary bone tumors of the orbit are a group of bone lesions, both neoplastic and reactive, arising from the bone walls of the orbit. They are rare, accounting for 0.6–2% of all orbital tumors [1, 2].

This chapter discusses the clinical and diagnostic aspects and the management of the different tumor types.

15.2 Topographic and Pathological Classifications

Bone tumors may arise from all orbital walls. Those occurring in the superior wall arise from the frontal bone and frontal sinus wall; those occurring in the medial wall originate from the ethmoidal sinus and nasal cavities. Tumors of the lateral orbital wall are extension of sphenoid wing tumors; those of the inferior wall arise from the maxillary bone and sinus. Besides, bone tumors may exceptionally be located within the orbital cavity, with no relationship with the orbital walls (extraskeletal tumors) [3]. Thus, most bone lesions involving the orbit are really cranio-orbital tumors that arise from cranial structures.

According to their histological origin and pathology, the primary orbital bone tumors may be classified into dance groups (Table 15.1). The group of benign fibro-osseous and cartilaginous lesions includes the benign tumors and the fibrous dysplasia [4]. The group of reactive lesions

Table 15.1 Classifications of primary orbital bone pathologies

1. Benign Fibro-osseous and Cartilaginous Lesions
Osteoma
Osteoblastoma
Ossifying fibroma
Chondroma
Fibrous dysplasia
2. Reactive Bone Lesions
Aneurysmal bone cyst
Giant cell granuloma
3. Neoplasms
Osteosarcoma
Ewing sarcoma
Chondrosarcoma
Hemopoietic and histiocytic lesions
Giant cell tumor
4. Vascular Lesions
Osseous hemangioma

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includes the aneurysmal bone cyst and the giant cell granuloma. In the group of neoplasms are listed malignant tumors of bone, cartilaginous and hemopoietic origin, and benign lesions such as histiocytosis and giant cell tumor. Osseous hemangioma is the unique vascular lesion.

15.3 General Clinical and Diagnostic Aspects

The clinical presentation and evolution of primary orbital tumors mainly depend on their type and growth rate. Benign bone cartilaginous and reactive lesions may often be asymptomatic. Symptomatic cases present with slow progressive mass effect over years [4], causing gradual proptosis and eye displacement. Sudden increase of the mass effect due to hemorrhage causing worsening of the proptosis may be observed for several reactive bone lesions. Malignant tumors present with signs of mass effect and infiltration within weeks or months, including pain, decrease of the vision, and impairment of the eye movements.

Diagnostic imaging studies include computerized tomography (CT) and magnetic resonance (MR). CT with bone window and particularly 3D CT define the tumor components (compact bone versus cartilaginous, fibrous and soft components), the intraorbital tumor extension, the involvement of the optic canal and superior orbital fissure, the osteolysis of the surrounding involved bone, the tumor extension into the paranasal sinus cavities. MR better defines the compression and displacement of the intraorbital structures, mainly the optic nerve.

15.4 General Surgical Management

The surgical approach to orbital bone tumors mainly depends on their location and size. Endoscopic endonasal and orbital, external orbital and combined cranio-orbital approaches may be used. The endoscopic endonasal approach is mainly used for tumors arising in the frontal, ethmoid, and sphenoid sinuses and located in the medial orbital compartment [5, 6]. The traditional external orbital approach is represented by the orbitotomy that can be performed through a mininvasive eyelid anterior incision or through antero-lateral incision if an osteoplasty is needed. The orbital endoscopic approach, as unique procedure or combined with the endonasal endoscopic one, allows to resect even large bone tumors [7]; in these instances, the endoscopic endonasal and orbital approaches may be combined with a microsurgical external orbitotomy [8, 9]. On the other hand, the cranio-orbital approaches may be limited to cases with significant cranial extension.

15.5 Specified Pathological Entities

15.5.1 Benign Fibro-Osseous and Cartilaginous Lesions

15.5.1.1 Osteoma

Osteomas are benign bone tumors which frequently occur in the cranial sinuses. The frontal sinus is most frequently involved (58–68%) followed by the ethmoidal sinus [10]. The orbital involvement results from extension from the frontal bone and frontal and ethmoid sinuses. Obstruction of the sinus ostia by the tumor may result in a mucocele.

Most sinus osteomas are asymptomatic; those arising from the frontal bone may present with a palpable mass. In symptomatic cases, gradual proptosis is the main complaint, with globe displacement. Posterior osteomas arising from the sphenoid sinus may cause compression of the apical structures.

Osteomas are classically associated with Gardner syndrome, an autosomal-dominant disease that presents with intestinal polyposis, osteomas and other cutaneous soft tissue tumors [11]. Thus, patients with multiple skull osteomas must be investigated with gastrointestinal studies due to risk of colon cancer [12].

On CT scan osteomas appear as hyperdense bone masses with regular margins within the involved sinus and orbit (Figs. 15.1 and 15.2).

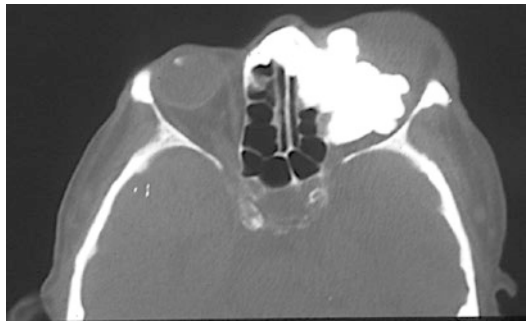

Fig. 15.1 CT of the skull: large lobulated osteoma of the ethmoid sinus largely extending in the left orbit and causing lateral globe displacement and proptosis

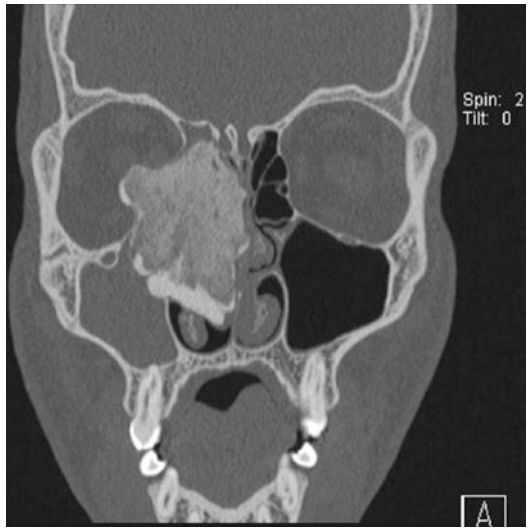

Fig. 15.2 CT of the skull, coronal section with bone window: **osteoma** of the right ethmoid and maxillary sinus extending into the inferomedial compartment of the right orbit

This imaging technique well defines the tumor size and degree of extension.

Some osteomas may be observed only because of their slow growth rate.

The surgical removal is warranted for enlarging and/or symptomatic tumors, for those with intracranial extension [5, 13, 14] or sinus outflow obstruction resulting in mucocele [4]. Many osteomas may be treated by endoscopic endonasal or/and orbital approach [6, 9]. Larger tumors require an external orbital or combined approach [14]. The cranio-orbital approach may be reserved to large posterior osteomas involving the orbital apex and optic canal [15]. The complete resection by drill cavitation up to the tumor attachment is the goal of surgery. The tumor attachment is more often with a large base, although a narrow pedicle may sometimes be found.

15.5.1.2 Osteoblastoma

Osteoblastoma is a rare benign bone tumor (1% of all bone neoplasms). Cranio-maxillo-facial locations account for about 15% and mainly involve the mandible and temporal bone. The endo-orbital location is exceptional, with a few reported cases [2, 16–20].

The orbital involvement arises from the orbital roof or ethmoid sinus and causes gradual globe displacement and proptosis.

CT scan shows a lesion with well-demarcated sclerotic borders, bone destruction, mottled calcifications, and variable contrast enhancement.

The complete surgical resection is suggested, because of the high recurrence rate after incomplete resection and the risk of malignant transformation (16–20%) [20].

15.5.1.3 Ossifying Fibroma

Ossifying fibroma is a rare distinct fibro-osseous neoplasm, characterized by fibrous tissue intermixed with a mineralized component. It is most frequently found in the mandible in young individuals. The orbital location is very rare [1, 2, 4, 21, 22] and occurs from tumors of the frontal, ethmoidal, and maxillary bones. It manifests with slow, painless proptosis, and globe displacement.

The radiological aspect is a well-circumscribed round mass with central osteoblastic and osteolytic areas. The tumor mass is often large.

Complete surgical excision is indicated.

15.5.1.4 Chondroma

Chondromas can arise in the intracranial and extracranial portions of the skull base (10% of all chordomas). They mostly occur in the parasellar region, middle fossa and ethmoid sinus [23–26]. Those involving the orbit are exceptional (0.07–0.15% of all mesenchymal orbital tumors) [1,

21]. These tumors occur in adolescents and young adults and present with a slowly growing mass near the orbital rim and trochlea [23, 27].

The CT aspect is a well-circumscribed hyperdense inhomogeneous sessile or pedunculated mass.

MR allows to differentiate the pure cartilaginous components from those variably mineralized; the high-water content in non-mineralized portions provides low T1 signal and high T2 signal [24].

Because of the risk of malignant degeneration, wide surgical resection is the treatment of choice.

15.5.1.5 Fibrous Dysplasia

Fibrous dysplasia is a benign congenital skeletal condition characterized by bone thin cortex and fibrous replacement with bone marrow [28]. It exists in two forms: monostotic and polyostotic.

The craniofacial involvement occurs in 10% of the monostotic forms and in 50–100% of the polyostotic forms. It presents with swelling and deformities of the affected bone areas. The orbital involvement occurs in fibrous dysplasia affecting the frontal, sphenoid, and ethmoid bones. The clinical presentation includes facial asymmetry, proptosis, and globe displacement, often lasting for years at the diagnosis and slowly worsening during the life [29–33]. Other symptoms, according to the location, are diplopia, cranial nerve palsies, intracranial hypertension. Decrease of the visual function due to optic nerve compression may also be present. Malignant sarcomatous degeneration is rare and associated to pain and rapid symptoms progression [34].

The radiological aspect of the fibrous dysplasia is often rather typical. The involved bone appears to be expanded with distorted anatomical form. CT scan shows an inhomogeneous density due to the ratio of fibrous and bone areas (Fig. 15.3); cystic and more sclerotic areas may be present [35]. This bone structure may also be evident on the skull radiograms. On MR, fibrous dysplasia shows low T1 intensity and heterogeneous T2 signal [30, 36].

Most patients with fibrous dysplasia may be treated conservatively, due to the very slow progression of the disease and long stable periods

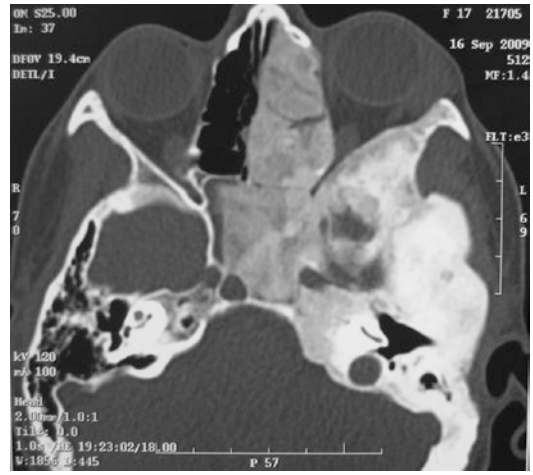

Fig. 15.3 CT scan: extensive **fibrodysplasia** of the skull base involving the sphenoid sinus, the left ethmoid sinus, and the left sphenoid wing; the left orbit is invaded and narrowing with conseguent proptosis

[37]. Bisphosphonates are the initial medical treatment and may result in pain relief, cosmetic improvement, and normalization of the bone turnover [38]. Indications to surgery include significant deformity, ophthalmological and neurological deficits, and the rare malignant degeneration.

Complete resection of the involved bone is often difficult and sometimes impossible; thus partial resection may be advisable. The surgery may require combined cranio-facial approaches with the cooperation of neurosurgeon, ophthalmic, and maxillofacial surgeons. The reconstruction is realized in one-step operation.

The optic nerve decompression is advised in patients with initial visual deficit; on the other hand, the prophylactic decompression of the optic canal is at risk of postoperative blindness and should be avoided [39].

15.5.2 Reactive Bone Lesions

15.5.2.1 Aneurysmal Bone Cyst

The aneurysmal bone cyst is a benign reactive lesion consisting in a red-brown friable mass containing blood-filled cysts separated by septa of trabecular bone and surrounded by fibrous tis-

sue and reactive bone. The mass of variable size often causes extensive osteolysis.

Aneurysmal bone cysts of the orbit are exceptional with only 33 reported cases [40–45]. Most are located in the orbital roof or lesser in the sphenoid-ethmoid bone. They may cause symptoms of chronic mass effect, such as proptosis and diplopia; sudden symptoms may result from intralesional hemorrhage. Some aneurysmal bone cysts are associated to the other bone pathologies, such as several tumours and fibrodysplasia [1, 4].

The radiological aspect on CT and MR is a round destructive bone lesion with irregular contrast enhancement and cyst with hemorrhagic component (Fig. 15.4) [44–46]. The biomolecular studies of detection of ubiquitin-specific peptidase 6/tre-2 gene may allow to confirm the diagnosis [45].

The surgical resection and curettage result in clinical remission. The preoperative embolization is useful for large cysts with high vascular flow. Recurrences may be observed, usually within 2 years. The medical treatment with receptor activator of nuclear factor kappa-b ligand (RANKL) inhibitors may improve the outcome [45].

15.5.2.2 Giant Cell Granuloma

Giant cell granuloma is a non-neoplastic reactive bone lesion which may result from trauma, inflammation, or infectious processes [47]. It consists in uniform cell stroma with fibroblasts, spindle-shaped and mononuclear infiltrative, and giant cells [48]. This reactive bone lesion mostly occurs in maxilla, mandible, and cranial bones; on the other hand, the orbital location is very rare [2, 4, 48–50]. The lesion occurs in the superolateral orbital compartment and causes variable bone erosion. It tends to be silent and relatively stable and may present with painless deformation of the involved bone and intraorbital mass lesion with proptosis [51].

The radiological aspect on CT is a high density mass with mildly enhancement and bone destruction. Surgical excision and additional curettage is indicated for large and symptomatic lesions.

The giant cell granuloma must be differentiated from other granular lesions, including giant cell tumor, cholesterol granuloma, Langherans cell histiocytosis, aneurysmal bone cyst, Brown's tumor of hyperparathyroidism [48].

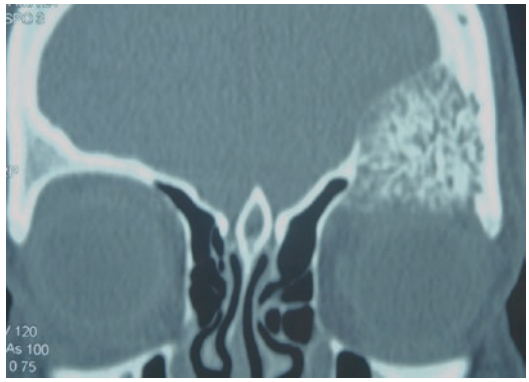

Fig. 15.4 CT in coronal scan: large hyperdense inhomogeneous mass lesion of the left frontal bone and left orbital roof causing diffuse osteolysis and invading the cranial cavity and the orbit (**aneurysmal bone cyst**)

15.5.3 Neoplasms

15.5.3.1 Osteosarcoma

Although osteosarcoma is the most common primary bone malignant tumor, the orbital involvement is rare and mainly occurs from a maxillary location. Orbital osteosarcomas may arise "de novo" or may be secondary to Paget disease, fibrous dysplasia, or radiotherapy [1, 2, 21, 52–55].

Orbital osteosarcomas present with a several month history of progressive mass lesion and infiltrative effects consisting in orbital pain, diplopia and decreased vision.

The radiological appearance is an irregular lytic and sclerotic bone mass with infiltrating soft tissue component [53, 54]. Exceptional cases of primary orbital osteosarcomas without connection to the bone have been reported [3, 56]; they are malignant mesenchymal neoplasms with osteoid matrix.

The treatment protocol includes preoperative chemotherapy, surgical resection, and postoperative chemotherapy. These tumors are resistant to

radiotherapy, which is reserved to residual and recurrent cases. The prognosis is poorer than osteosarcomas of other skeletal regions, because of the often incomplete resection.

15.5.3.2 Ewing's Sarcoma

Ewing's sarcoma is a highly malignant, small round cell neoplasms derived from primitive neuroectodermal cells with variable grades of differentiation. The two primary forms are skeletal and extraskeletal. Ewing's sarcoma accounts for 10% of all primary bone neoplasms and 4% of head and neck tumors. Skull neoplasms are mainly located in the maxillary bone and sinus, followed by ethmoid and frontal bones [57, 58]. Primary Ewing's sarcomas of the orbit are rare and mainly arise from the ethmoid sinus wall [58–62].

The clinical presentation occurs in the first two decades of life with non-axial proptosis of short duration and orbital pain. The infiltrative tumor mass causes bone destruction and is associated with a soft tissue component (Fig. 15.5).

Patients with orbital Ewing sarcoma must carefully be investigated for the possible presence of a primary tumor because the orbital locations are more often metastatic.

The treatment protocol first includes chemotherapy followed by surgery and adjuvant chemotherapy and eventually radiotherapy and proton beam therapy [62]; it may result in more favorable outcomes. Complete resolution followed by chemotherapy and radiotherapy alone has been reported.

15.5.3.3 Chondrosarcoma

Chondrosarcomas occurring in the orbit originate in the sinuses and nasal cavities [63–68]. The orbital extension causes medial or inferior mass effect with pain, proptosis, and globe displacement associated to symptoms of nasal sinus obstruction. CT and MR show a well-defined mottled lesion with calcified areas and moderate contrast enhancement associated to variable and often extensive osteolysis. The presence of areas of bone metaplasia may cause more hyperdense aspect on CT (Fig. 15.6).

Surgery through endoscopic endonasal approach [68, 69] or external route is the treatment of choice. The entity of surgical resection is the most important factor affecting overall survival. However, complete resection is often not possible, because of the extensive bone involvement. Adjuvant radiotherapy and chemotherapy are usually indicated. The 5-year survival is very variable (44–87%) and recurrence is estimated at 40–60% [68].

15.5.3.4 Hematopoietic and Histiocytic Lesions

Multiple myeloma and more rarely solitary plasmocytoma may involve the orbital bone [2, 52, 70]. The clinical presentation includes pain and

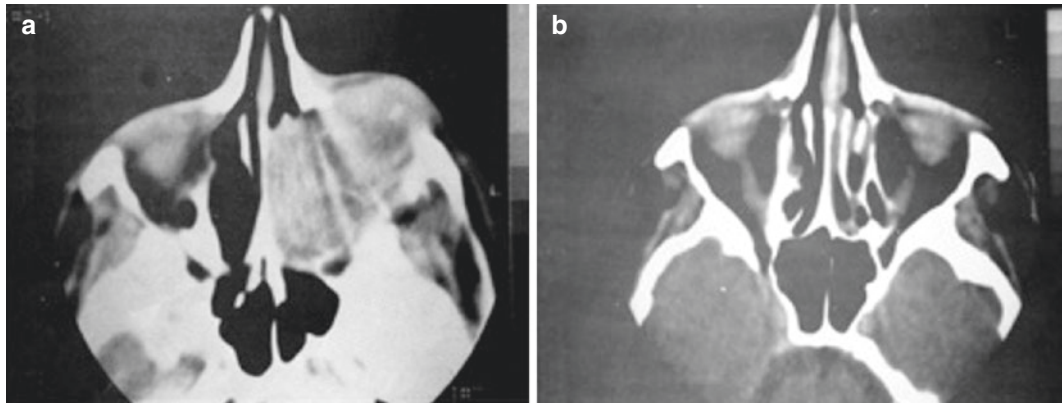

Fig. 15.5 (**a**) Post-contrast axial CT of the skull: mass lesion of the left ethmoid sinus invading the left orbit and causing moderate proptosis (**Ewing's sarcoma**); (**b**) Postoperative CT shows resection of the tumor and resolution of the proptosis

subacute proptosis. Symptoms and signs of systemic involvement may be present.

The Langerhan's cell histiocytosis, also defined eosinophilic granuloma, is a benign lesion which may rarely occur in the orbit [2, 71, 72], mainly in the supero-lateral orbital wall and in children. It causes osteolysis and intraorbital extension resulting in proptosis. The CT finding of central hypodensity with enhancing rim is rather typical (Fig. 15.7). The surgical curettage is curative.

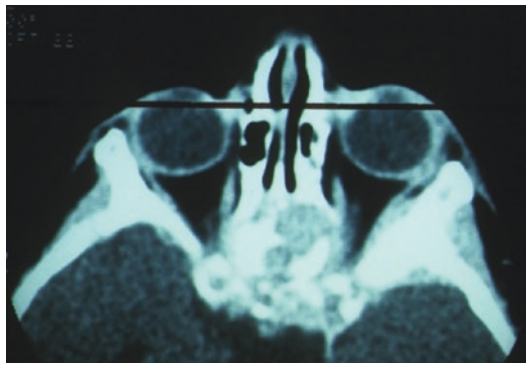

Fig. 15.6 CT of the skull base: large round hyperdense mass lesion of the sphenoid sinus, extending in both orbits, mainly on the left (**chondrosarcoma** with bone metaplasia)

15.5.3.5 Giant Cell Tumor

Giant cell tumor is a benign neoplasm of mesenchymal origin accounting for 15–20% of all benign bone tumors. It presents as a soft mass that erodes the bone and is surrounded by shell of reactive bone. The craniofacial location is exceptional (1%); the orbital involvement may occur from the temporal bone and sphenoid and ethmoid sinuses [2, 4, 73–77].

The orbital locations present as lytic or soft tissue masses causing headache, decrease of vision, or cranial nerve palsies. CT shows a lytic lesion with thin cortex; however, this radiological finding is aspecific [76].

The surgical treatment with wide resection is the recommended treatment, whereas radiotherapy should be reserved to inoperable cases. However, the recurrence rate ranges from 7% to 60% according to the extent of resection.

15.5.4 Vascular Lesions

15.5.4.1 Hemangioma

Intraosseous hemangiomas of the skull are rare (5% of all locations) [78, 79] and most frequently occur in the frontal bone; the orbital

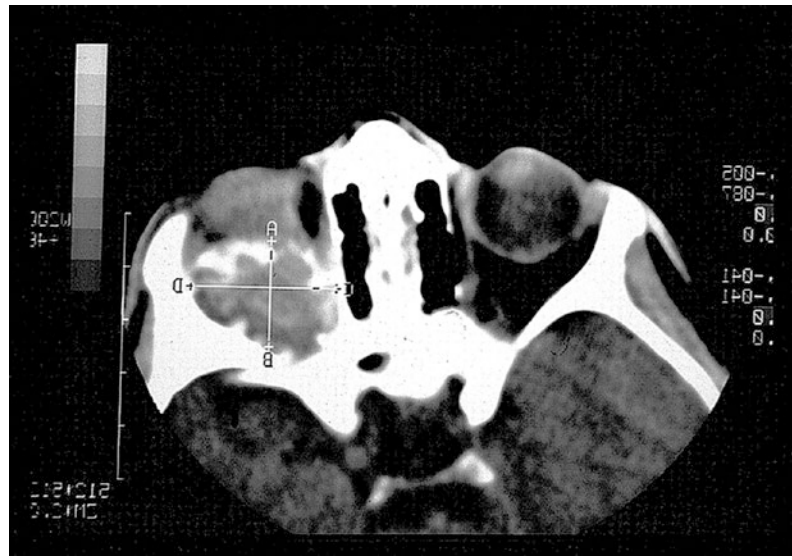

Fig. 15.7 CT of the orbits: large lesion of the right orbit associated to osteolysis of the lateral orbital wall; the lesion shows central hypodensity and enhancing rim (**eosinophilic granuloma**)

involvement is exceptional and mostly at the orbital rim [79–82].

Orbital cavernous haemangioma causes a painful or painless mass resulting in proptosis and visual impairment.

The radiographic aspect is a typical circumscribed area with pattern of trabeculation radiating from a common center [82]. CT clearly defines the typical trabecular pattern and stippled matrix [79]. On MR, the intensity signal varies according to the venous blood flow and the bone marrow.

The complete surgical resection is the treatment of choice in symptomatic cases; however, it may be difficult due to the profuse bleeding.

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