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Intraorbital Cystic Lesions: An Imaging Spectrum

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Presence of a cyst or a cystic component in an intraorbital mass often narrows the list of differential diagnoses to specific entities. Such a lesion in the orbit may arise from structures within the orbit, globe, and lacrimal system or from neighboring paranasal sinuses or meninges. Common congenital and developmental lesions encountered within the orbit include dermoids and epidermoids, and infrequently coloboma. Parasitic cysts (cysticercus), orbital abscess, mucocele, and vascular lesions are the most common acquired pathologies giving rise to fluid-containing lesions within the orbit. The role of a radiologist is crucial in expediting the diagnosis of orbital lesions with the help of characteristic imaging features on ultrasound, computed tomography, or magnetic resonance imaging. It also helps in identifying complications in others where formulation of an early and effective management strategy is vital for preserving vision.

Introduction

A cyst is a sac like structure lined by epithelium and containing fluid or semisolid material. Cysts may be extraocular or rarely intraocular in location. The spectrum of such lesions is wide. It consists of rather easily managed benign lesions such as uncomplicated dermoids and parasitic cysts at one end to more sinister pathologies such as orbital abscesses and cystic malignant tumors on the other. These lesions are encountered across all age groups with varied clinical presentations. An orbital cyst may be an unsightly congenital cystic eye or colobomatous cyst in a neonate, which portends a poor visual outcome, a venolymphatic malformation in a young child with an acute and often dramatic presentation, or an insidious, slowly enlarging mucocele presenting with proptosis in an adult.[1] A superficial lesion often presents as a subcutaneous mass or nodule, whereas lesions located deep in the orbit may present with visual disturbances, proptosis, squint, or signs and symptoms of inflammation.[2,3]

Shields and Shields[3] have classified orbital cystic lesions according to the cell of origin (Table 1). Kaufman et al[2] proposed a simpler classification based on the etiology (Table 2). They classified cystic lesions of the orbit as congenital, acquired, and those arising from adjacent structures[1,2] such as paranasal sinuses, brain, and meninges. The organ of origin, cyst wall, internal contents, and relationship to adjacent structures are well depicted on both computed tomography (CT) and magnetic resonance imaging (MRI). Bony abnormalities are well depicted on CT, whereas MRI because of its excellent soft tissue contrast helps characterize the internal contents and wall characteristics of the lesions. However, because of widespread availability and short imaging time, CT often remains the workhorse of orbital imaging.

A stepwise approach is useful in arriving at the correct diagnosis of orbital cystic lesions.

Step 1: Look at the cyst contents for the presence of fat and calcification; dermoids are fat-containing lesions.

Step 2: Look at the exact location of the lesion within the orbit. Dermoids are usually located along lines of closure of sutures; orbital varices and colobomatous cysts are intraconal masses. In case of large masses, the epicenter of the lesion is usually the site of origin.

Step3: Look at the relationship of the lesion to the globe, paranasal sinuses, and floor of anterior cranial fossa. Colobomatous cysts lie adjacent to a microphthalmic eye; a mucocele will usually communicate with the frontal or ethmoid sinus, and meningocele will be associated with defect in the anterior skull base. In the following sections, we discuss the most commonly encountered lesions, their clinical presentation, and radiologic features.

Congenital and Developmental Orbital Cysts

Choristoma (Dermoid, Epidermoid, and Dermolipoma)

A choristoma is defined as benign, disorganized proliferation of histologically normal tissues in an abnormal location.[4] Dermoid, epidermoid, and dermolipoma are choristomas seen in orbit and are among the most common benign orbital tumors of childhood.[1,5] They arise from embryonic cell rests that get sequestered or implanted along lines of closure of sutures, meninges, or in the diploe, or due to failure of separation of these cell rests from mesoderm or endoderm.[1] Both dermoid and epidermoid are lined

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Table 1
Classification of cystic lesions of the orbit (Shields and Shields[3])

I. Cysts of the surface epithelium
A. Simple epithelial cysts
1. Cutaneous epithelial cyst (epidermal cyst)
2. Conjunctival epithelial cyst
3. Respiratory epithelial cyst
4. Apocrine gland cyst
B. Dermoid cysts
1. Epidermal dermoid cyst
2. Conjunctival dermoid cyst
II. Teratomatous cyst (Teratoma)
III. Neural cysts
A. Neural cysts associated with ocular maldevelopment
1. Congenital cystic eye
2. Colobomatous cyst (neuroectodermal cyst with microphthalmia)
B. Neural cysts associated with brain and meningeal tissue
1. Cephalocele and ectopic brain tissue
2. Orbital optic nerve meningocele
IV. Secondary cysts from adjacent structures
A. Mucocele
B. Dentigerous cyst
V. Inflammatory cysts (parasitic cyst)
A. Echinococcal cyst
B. Cysticercosis
C. Others
VI. Noncystic orbital lesions with cystic component
A. Adenoid cystic carcinoma
B. Rhabdomyosarcoma
C. Lymphangioma
D. Others

by squamous epithelium, but dermoids also contain cutaneous appendages as hair follicles, sweat, and sebaceous glands.

Dermoid cysts can also be classified as cutaneous and conjunctival dermoid cysts, which represent 59% of the total. Based on location, they can be classified as *superficial or deep*. Superficial dermoids are most commonly located along frontozygomatic (60%) and frontolacrimal (25%) sutures[4] and present as painless, firm, subcutaneous periorbital masses in infancy or childhood. On imaging, they are seen as well-defined, unilocular masses, with fluid or fat density contents, causing scalloping of the adjacent bone (Fig 1). Approximately 15% may contain foci of calcification.[4] The presence of fat on imaging is pathognomonic.

Deep dermoids and epidermoids present later in life as slow-growing, discrete, well-circumscribed, freely movable, intraorbital or rarely intraosseous masses causing proptosis and diplopia or with signs and symptoms of inflammation if they rupture or get infected. They are situated along suture lines, or even at the orbital apex, and may extend into the adjacent bones. Imaging is essential to establish the exact extent of these masses, because these may have dumbbell-shaped extensions into the temporal fossa, intracranial cavity, or paranasal sinuses. If a choristoma is complicated by rupture or infection, CT/MRI will reveal an irregular wall, presence of inflammation in adjacent structures (Fig 1C), and irregularity of bony orbital walls. There may be a documented change in the longstanding image appearances (Fig 2).

Dermolipomas are composed of mature fat cells with very little epithelial cells. They are located on the outer canthus of the eye beneath the conjunctiva or in the superotemporal epibulbar region and occasionally causes intermittent ocular irritation. These lesions are often noncystic and infiltrate into the peribulbar region, lid, and surrounding orbital tissues. When present, these lesions should not be surgically excised. Bilateral dermolipomas are seen in Goldenhar syndrome.[1]

The differential diagnosis for choristomas depends on the location—it may be of lacrimal gland origin for lesions located in the superotemporal quadrant, mucocele and encephalocele for superomedially located masses, and solid orbital tumors for inflamed masses in other locations.

Superficial masses are managed by direct excision, whereas deep masses require combined orbital and neurosurgical approaches for removal. Occasionally, fistulization is seen after incomplete removal of orbital dermoid tumors. Hence defining the exact extent of the cystic mass and its relationship to adjacent structures on preoperative imaging is crucial for management.

Conjunctival Cysts

Conjunctival cysts are sequestrations of conjunctival and caruncular epithelia occurring in the superonasal aspect of the orbit without a bony defect. They are lined by nonkeratinizing conjunctival epithelium and may be congenital or traumatic in origin. These are distinguished based on a lack of historical or clinical evidence of trauma. They are most commonly located medially at or behind the caruncle and may have fat or fluid contents (Fig 3). When large, they can cause motility disturbances, pain, or refractive errors.[6] In a case series of 11 patients, Goldstein et al[6] found that larger conjunctival cysts caused mass effect leading to distortion of the globe in 6 patients and bone remodeling in another 6. Hence, imaging is essential when excision is being planned for a large conjunctival cyst; smaller lesions are managed by complete extirpation.

Orbital Germ Cell Tumors

Orbital germ cell tumors are very rare tumors, with an unknown incidence. Teratomas are benign germ cell tumors that are seen as unilateral, rapidly growing orbital masses in infant girls.[2] Primary orbital teratomas are confined to the orbit and are not associated with a bony defect, whereas combined teratomas extend into the periorbital tissues and cranial cavity. They are composed of cells derived from more than one germ layer and present as large masses that cause diffuse expansion of the orbit and massive unilateral proptosis.[7,8] Vision is threatened by exposure keratopathy of the displaced globe or owing to stretching of the optic nerve. Defects of orbital bones are present in combined teratomas. Orbital yolk cell carcinomas and endodermal sinus tumors are malignant orbital tumors seen in children.

On CT or MRI, germ cell tumors are seen as complex solid cystic masses that cause expansion of the orbit (teratomas) that displace

Table 2
Classification of cystic lesions of the orbit (Kaufman et al[2])

Classification	Examples
Congenital	Dermoid, epidermoid, teratoma, colobomatous cyst, congenital cystic eye, optic nerve sheath meningocele, and dacryocele
Acquired	Mucocele, vascular lesions, dacryocele, epithelial and appendage cysts, lacrimal gland cysts, hematic cysts, orbital abscess, parasitic cysts, tumors, and tumorlike conditions
Cysts arising from adjacent structures	Cephalocele, dentigerous cyst, and enterogenous cyst