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Epidemiology and Classification of Cystic Benign Orbital Tumors: A Review Article

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ABSTRACT: Benign orbital tumor is known to be with a broad variation in the clinical features and findings. Tumors in this small cavity could quickly result in crucial symptoms in vision, even though complete loss of vision. These tumors may be infectious, inflammatory or neoplastic. Management and follow up may differ based on the tumor type, site, and spread into other systems. Because of what is known that the orbital cavity constitutes a small space, so that tiny projections like that observed with tumors and infection could result in huge physiologic disorders in near structures leading to anatomical and weak or total loss of vision. Deep awareness about orbit structure and physiology is needed to be aware about the complications of different orbital disorders and surgical therapy. Orbital cystic tumors comprise a collection of masses with various sings, histopathology and pathogenesis. This article cast a light on the epidemiology and classification of "cystic benign orbital tumors".

INTRODUCTION

The bony cavities are orbits that contain fat plus the lacrimal apparatus and vessels, nerves and muscles of the eye along with the eyeball itself. The globe is protected by each orbit; the supportive tissues allow it to move torsionally, vertically and horizontally. Orbit anatomy is an interesting topic, quite difficult because of its complexity but very important to understand for normal connection between the central nervous system and the eye; also for pathways that infection or malignancy could spread through. (Gospe et al., 2018).In the orbit, connective tissue structures play a role in maintaining support and protecting what is inside. The ocular muscles and the globe are supported by orbital adipose tissue which acts as a cushion to protect these structures; it also helps in eye locomotion. The orbital septum is another connective tissue structure that forms an anterior border between the skin of the face plus fat and what lies inside the orbit preventing spread of infection into the orbit from surrounding facial skin structures (Luibil et al., 2023).

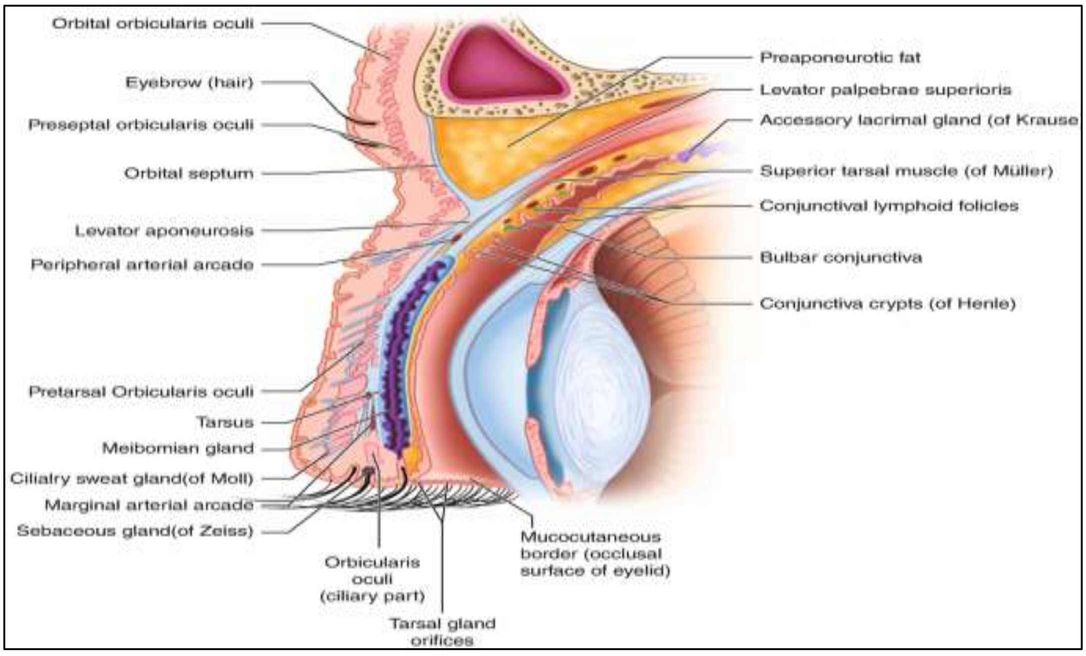

Figure 1. Anatomy of the orbit (Satterfield and Chambers, 2022)

The orbital cavity is a restricted space with well-defined anatomical points of reference. The orbits are usually symmetrical in the horizontal plane, bounded by straight lines along their medial walls and narrowing posteriorly towards the optic foramen. Bony medial wall components include the maxilla, lacrimal bone and ethmoid bone. The superior orbital rim is made up of in part by the frontal bone and contains the supraorbital foramen; while trochlear fossa and lacrimal fossa are also present in superior orbit. Lateral bony orbitarum includes zygomatic bone plus sphenoid greater wing which is thick anteriorly till equator allowing good field vision on temporal side, as in Figure 1. (Turvey & Golden, 2012).

The orbital floor is formed from three bones: zygomatic, palatine, and maxillary. The optic canal, inferior fissure of the orbit and superior fissure of the orbit harbor important neurovascular structures in the orbit. Ophthalmic artery, which is the major arterial blood supply of the orbital srtuctures, arises from the internal carotid artery. It anastomoses with branches from the external carotid to create a rich vascular network for supplying the periorbital region. Drainage of blood from the orbit is primarily through the inferior and superior ophthalmic veins, as in figure 2. (Satterfield and Chambers, 2022).

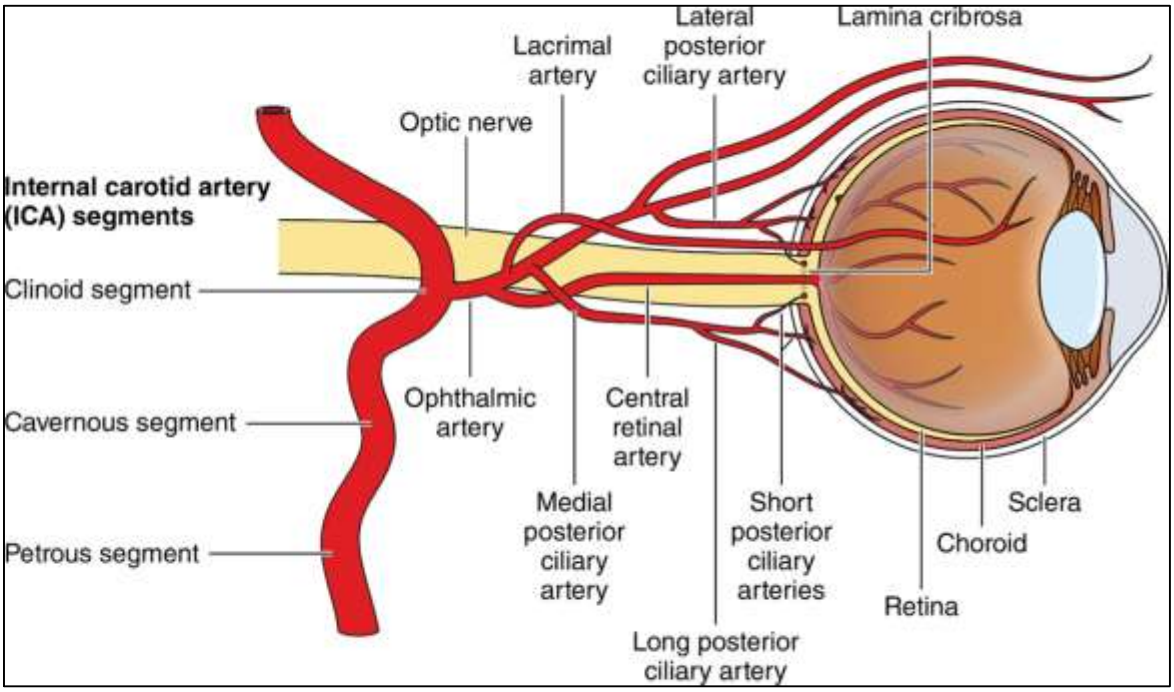

Figure 1. Blood supply in the orbit (René, 2006)

Tumors within such a small space could easily lead to major vision symptoms— even complete loss of vision. These tumors can either be infectious, inflammatory or neoplastic. Tumors that are situated in the front 66% of the orbit can usually be accessed ventrally through the conjunctivae and palpebrae; laterally based on the site through lateral orbital wall; or superiorly through a transcranial approach. Retrobulbar lesions medially located to the optic nerve can typically be dealt with by endoscopic endonasal approaches. Lesions at the orbital apex, more so if they are on the medial side of the optic nerve, usually need endonasal endoscopic intervention while those located laterally to the optic nerve are often approached through a lateral orbitotomy or transcranial intervention (Salgado-López et al., 2020; Murdock et al., 2023).

EPIDEMIOLOGY

The primary orbital tumors are not frequently encountered: the incidence is decreased and about 1 in 105 people. The majority of primary orbital masses are benign but there is a greater likelihood of malignancy in geriatric patients. While in adults, most primary benign orbital tumors have an equal distribution between males and females, although some subtypes including meningiomas and cavernous hemangiomas of the sphenoid wing and optic nerve sheath are more frequent in women. The risk factors based on geography or race for primary orbital tumors have not been established yet among adults. In a large retrospective series that included more than 2 thousand cases of orbital lesions sent to a specialized center for tissue diagnosis, benign tumors were identified in about 68% cases; cavernous hemangiomas constituted only 9% while dermoid cysts were more common with 14% (Bonavolontà et al., 2013).

The majority of masses were found in the upper outer orbital quadrant. The researchers observed that masses located in the lower inner quadrant have a greater chance of being malignant. Other patterns were noted as well. Different tumors tended to present more commonly based on their anatomic location. In the upper outer quadrant, dermoid cysts were identified as the most frequent masses. Mucoceles constituted most masses present in the upper inner quadrant. Cavernous hemangiomas were predominantly found in the lower outer quadrant while basal cell epitheliomas had a higher frequency among masses found in the lower inner quadrant. Among central orbital masses, meningiomas were most common (Murdock et al., 2023).

CLASSIFICATION

Essential orbital masses could be categorized depending on their original tissue that is affected by tumor. There are seven types of these tumors: lymphoid tumors, lipomatous tumors, osseous and fibro-osseous tumors, fibrocystic tumors, neural tumors, vasculogenic tumors, cystic tumors. As in figure 3. Orbital cystic tumors comprise a collection of masses with various sings, histopathology and pathogenesis (Günalp & Gündüz, 1996).

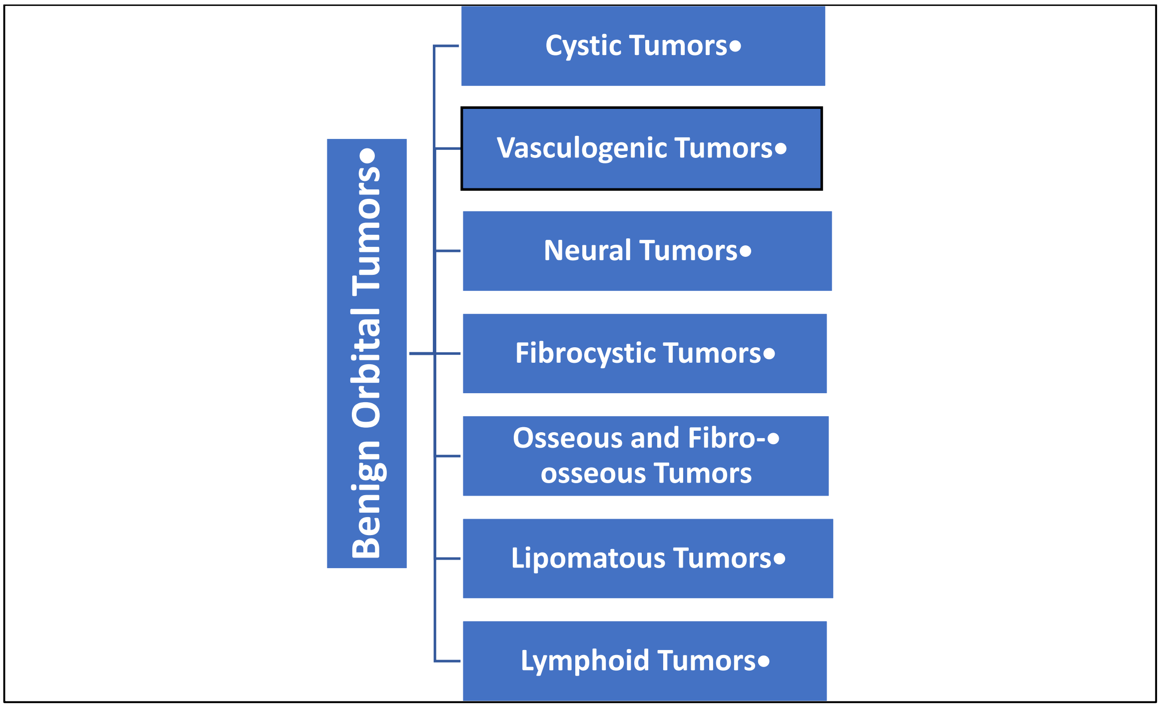

Figure 3. Classification of orbital benign tumors

Types of Cystic Benign Orbital Tumors

The broad classifications involve non-cystic lesions with cystic component, inflammatory cysts, secondary cysts, neural cysts, teratomatous cysts and cysts of surface epithelium. In the surface epithelium, the cysts are further classified into dermoid cyst (conjunctival and epidermal) and simple epithelial cyst (apocrine, respiratory conjunctival and epidermal gland). Amongst pediatric patients the most frequent orbital lesion is epidermal dermoid cyst (dermoid) which is considered to be an orbital lesion of childhood: it makes up about 41% of all orbital lesions in children, which also make up about 90% of all childhood cases that come to biopsy or surgical incision. Neural cysts are those related to ocular maldevelopment (colobomatous cyst and congenital cystic eye) as well as those related to meninges and brain tissue (Shields & Shields, 2004).

Epidermoid Cysts

Epidermoid cysts have a lining of a stratified squamous epithelial tissue with lamellae having desquamated keratin. Infrequently located in the orbital space but to be comprised the differential for deepest orbit cystic tumors, epidermoid cysts may occur as essential congenital masses or formed after traumatic dislocation of surface epithelial tissue (Hoang et al., 2019).

Dermoid Cysts

The differentiation between epidermoid and dermoid cysts is that the latter contains mesodermal elements. While dermoid cysts make up almost fifty percent of orbital childhood tumors they are hardly ever detected in adulthood (Shields & Shields, 2004).

Mucoceles

Mucoceles are sterile mucus-filled cystic cavities. They usually develop as a result of an obstruction in the outflow from the ethmoidal or frontal sinuses. Rarely formed and weak-growing, these masses can still enter the orbit if they grow large enough; this then results in symptoms similar to other orbital masses. Mucoceles have had more frequent reports among patients who have had trauma or sinus surgery. The cysts have lining with a simple epithelial tissue and might contain chronic or acute inflammatory contents: thus, acting more as secondary lesions themselves (Devars du Mayne et al., 2012).

Apocrine hidrocystomas

Apocrine hidrocystomas are often found on the eyelid but not commonly documented within the eye socket. They are cystic lesions described by having a capsule made up of two layers of columnar epithelial tissue with small outer projections (called "apocrine snouts") from the lumen. These tend to come back if they are not totally taken off, mainly because of this capsule's recurrent nature (Ssi-Yan-Kai & Pearson, 2012).

Hematic pseudocysts

An unusual lesion of the orbit rarely reported is an orbital hematic pseudocyst. Acute proptosis at onset would be the most frequent feature of a hematic cyst following trauma to the orbit, even though remote. Chronic orbit hematic cysts are recorded and are approximately identical to chronic subdural hematomas in histology as they are both cystic lesions with blood breakdown products and/or only blood; based on the chronicity of the tumor, granulomatous reactions or products of inflammation can also be found (Ali et al., 2012).

Meningoencephaloceles

Intracranial tissues can herniate into the orbit through defects in the orbital roof leading to the formation of meningoencephaloceles. Although uncommon, these intraorbital masses are an important consideration for patients with manifestations of a superonasal or superior orbital mass. Pulsatile proptosis places meningoencephaloceles on the differential along with vascular tumors and carotid-cavernous fistulas as inappropriate biopsy or resection, which can be devastating outcomes. It is crucial to obtain radiographic studies for diagnosing these masses (Umerani et al., 2018).

CONCLUSIONS

Benign orbital tumor are known to be with a broad variation in the Clinical features and findings. Tumors in this small cavity could quickly result in crucial symptoms in vision, even though complete loss of vision. These tumors may be infectious, inflammatory or neoplastic. Management and follow up may differ based on the tumor type, site, and spread into other systems. Because of what is known that the orbital cavity constitutes a small space, so that tiny projections like that observed with tumors and infection could result in huge disorders in near structures leading to anatomical and weak or total loss of vision. Deep awareness about orbit structure and functions is needed to be aware about the complications of different orbital disorders and surgical therapy. Orbital cystic tumors comprise a collection of masses with various sings, histopathology and pathogenesis.

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