CASE REPORT

Orbital Hematic Cyst: Case Report and Clarification of Terms

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ABSTRACT—We present a case of a hematic cyst and review the literature in order to clarify nomenclature discrepancies regarding terms used to describe cysts containing blood and blood breakdown products in and around the orbit. We believe orbital cysts containing blood and blood breakdown products should be separated into two major categories depending on the presence or absence of an epithelial or endothelial lining. A hemorrhage into a preexisting lesion such as a dermoid or lymphangioma, which contains blood breakdown products lined by an endothelial or epithelial lining, should be referred to as an acute or chronic hematoma in a dermoid or lymphangioma. In contrast, a hematic cyst contains blood breakdown products and has no epithelial or endothelial lining but rather a fibrous pseudocapsule.

Numerous terms, including hematoma, hematic cyst, blood cyst, chocolate cyst, orbital hemorrhage, hematic cyst, cholesteatoma, and cholesterol granuloma, have been used to describe orbital cystic collections of blood and its breakdown products. The multitude of terms and the inconsistency of their application, in addition to the relative infrequency of these lesions, cause difficulty in making useful statements about these entities. We present a case of a 39-year-old man with a hematic cyst of the orbit and review the literature in order to clarify nomenclature discrepancies.

CASE REPORT

A 39-year-old right-handed man presented with proptosis, inferomedial globe displacement, and diplopia. This patient was a healthy man with a 3-year history of “floaters” in the left eye which were stable until one month prior to admission when he developed diplopia and

proptosis. One week prior to admission the patient noticed left temporal headaches and nausea. A CT scan showed a left superolateral orbital mass arising within the bone at the junction of the lateral wall and roof of the orbit (Fig. 1). The mass was partially cystic, had some calcifications, and eroded into the lateral aspect of the frontal sinus. The patient denied any history of recent or remote trauma.

Physical examination was notable for 6 mm of proptosis of the left eye. There was mild ptosis of the left lid. There was marked limitation of upgaze as well as mild limitation of all other fields of gaze. Pupillary reactivity, visual fields, and color vision were normal. Dilated fundoscopic exam showed chorioretinal indentation of the left eye. The remainder of his cranial nerves were normal.

A left orbitofrontal craniectomy was performed. Arising within and expanding the bone of the superolateral orbital roof was a yellow and brown mass. The entire mass was removed including portions of the orbital roof and lateral orbital wall. The dura was preserved and the

Figure 1. (A). Coronal and axial CT scans showing an expansile lesion in the left superolateral orbital roof. (B). Coronal CT scan showing that the lesion extends into the frontal sinus.

periorbital repaired. Postoperatively the patient returned to normal. He has no evidence of recurrence after 18 months.

The resected tumor, measuring 3.2 cm × 2.8 cm × 2.0 cm in aggregate dimension, consisted grossly of fragments of soft, tan-brown friable tissue admixed with hard tan-gray bone. Histopathologic examination of the specimen showed discontinuous bands of fibrous tissue, mononuclear inflammatory cells, multinucleated foreign-body type giant cells, cholesterol clefts, hemosiderin and lipid-laden macrophages, and fragments of nonviable and viable lamellar bone with organizing hematoma (Fig. 2). There was no epithelial lining or osteoid formation. The pathological diagnosis was hematic cyst.

DISCUSSION

Traumatic and nontraumatic collections of blood and blood breakdown products in the bone and soft tissue structures of the orbit are inconsistently defined by a number of terms. Hematocele is a general term referring to any collection of blood in the orbit.^{1,2} In our opinion more specific terms than hematocele should be used to distinguish among the different blood-related lesions found in and around the orbit.^{3,4} The term hematoma should be reserved for an acute traumatic or spontaneous collection of blood which is reabsorbed in a matter of days to weeks.^{5,6} The etiology of orbital hematomas includes trauma, sickle cell disease, malaria, hemophilia, scurvy, uremia, venous hypertension, rickets, myelogenous leukemia, and blood dyscrasias.^{1,6-8}

In most instances, orbital hematomas break down and are reabsorbed without sequelae. However, occasionally the degradation of blood to its breakdown products results in a granulomatous reaction.⁸ Cholesterol and hemosiderin crystals are the blood breakdown products that are phagocytized by histiocytes, resulting in the formation of giant cells.³ Over time, calcium deposition occurs and hematoidin appears. Hematoidin is an iron-free pigment, closely related to bilirubin, which forms when hemoglobin is slowly broken down in a hemorrhage in tissue lacking a good oxygen supply.^{5,9} Hematoidin reacts to Gmelin stain for bile, while hemosiderin, a more immediate product of hemoglobin breakdown, reacts to Prussian blue stain for free iron.^{5,9}

Hemorrhage into a preexisting lesion such as a lymphangioma, dermoid, or hemangioma is usually referred to as a chocolate cyst or blood cyst.^{1,3,4,6,10,11} This lesion has an endothelial or epithelial lining on pathologic examination. Since some authors use the term blood cyst to refer to a nonepithelial lined fibrous cyst containing blood breakdown products,^{9,7,12} confusion is created. In our opinion, the best terminology would be to simply call these lesions an acute or chronic hematoma in a dermoid or an acute or chronic hematoma in a lymphangioma rather than use the term blood cyst or chocolate cyst.

Orbital cholesteatomas and orbital epidermoid cysts are synonyms which describe lesions with epithelial elements, keratin, cholesterol clefts, lipid-laden histiocytes, fibrous stroma, and in some cases hemorrhage.¹³ We believe if a hemorrhage is present this should be called an acute or chronic hematoma in an epidermoid cyst rather than orbital choesteatoma, blood cyst, or chocolate cyst.

The feature that we use to distinguish a hematic cyst from other lesions with blood breakdown products is the lack of an epithelial or endothelial lining. Numerous papers support this definition.^{1,3,4,6-9,11-15} Using the term hematic cyst to define lesions with an epithelial or endothelial lining, or those that result from hemorrhage into a preexisting lesion,^{5,16,17} in our opinion, groups different pathologic entities together under one name.

A cholesterol granuloma contains all of the elements of orbital cholesteatomas except for epithelial elements.^{3,13} Therefore, orbital cholesterol granulomas are essentially what we would call a hematic cyst. The etiology of petrous apex cholesterol granulomas is controversial but is believed to be distinct from that of a hematic cyst. One major theory is that a pneumatized petrous apex air cell becomes obstructed and the air cell no longer communicates with the outside world.¹⁸ This leads to relative hypoxia, mucosal edema, and then hemorrhage, resulting in a foreign-body granulomatous reaction.¹⁸ Although we do not believe our lesion developed from a trapped air cell, the lesion in our patient did erode into the frontal sinus. Histopathologically, petrous apex cholesterol granulomas and orbital hematic cysts are identical. However, it seems unlikely that the proposed etiology of petrous apex cholesterol granulomas applies to orbital hematic cysts.

Hematic cysts can be further classified into acute and chronic. Acute hematic cysts lack a thick capsule and cholesterol crystals and present typically with post-traumatic proptosis, conjunctival chemosis, choroidal folds and restricted ocular motility.^{11,14} Chronic hematic cysts may produce erosion of bone and consist of cholesterol clefts, giant cells, hematoidin, hemosiderin, and a fibrous pseudocapsule.^{3,11} These benign lesions may increase in size. Proposed mechanisms of growth include the presence of an osmotic gradient which draws fluid into the cyst and rebleeding into the cyst secondary to fibrinolytic activity.^{1,5,9}

Common clinical features seen in association with hematic cysts are recent or remote history of trauma, abrupt or acute onset, proptosis, ocular dysmotility, superior orbital location, and a high likelihood of cure following surgical intervention.^{1,6,14,19} Surgical excision is the typical treatment for a hematic cyst although some authors contend that incomplete removal^{6,7} or needle aspiration is adequate.²⁰

Our patient exemplifies all of the key clinicohistopathologic features of a hematic cyst. Clinically he presented with progressive unilateral proptosis. Imaging showed an expansive mass of the superolateral orbital

A

B

Figure 2. (A). Photomicrograph of a hematoxylin/eosin stained slide at 20X showing lipid-laden macrophages, multinucleated giant cells, mononuclear inflammatory cells, and small cholesterol clefts in a fibrous background. (B). Photomicrograph of a hematoxylin/eosin stained slide at 20X showing numerous needle-like cholesterol clefts.

bone. Histopathology showed a non-epithelial-lined cyst with a fibrous pseudocapsule containing blood breakdown products including hemosiderin, cholesterol clefts, lipid-laden macrophages, multinucleated giant cells, and mononuclear inflammatory cells. Surgical resection we believe was curative.

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