

Orbitofrontal Cholesterol Granuloma: Report of Two Cases and Review of Literature

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Abstract

Purpose: To report two cases of orbitofrontal cholesterol granuloma, which is a rare progressive destructive disease

Case reports: We report two patients with orbitofrontal cholesterol granuloma; a 50-year-old man and a 20-year-old woman; both with progressive proptosis due to a cystic lesion in the superior part of the orbit with frontal bone erosion and intracranial extension. The second case had a previous incomplete surgery 5 years before and presented with recurrent orbital tumor. We performed orbitotomy and cyst evacuation for both cases, which resulted in complete resolution without any complications. At last follow-up (2 and 6 years after operations respectively), no recurrence was observed.

Conclusion: We presented the clinical and para-clinical manifestations and surgical results. Complete resolution of symptoms without any recurrence was observed after surgery.

Keywords: Cholesterol Granuloma, Hematocele, Surgery, Proptosis

Iranian Journal of Ophthalmology 2011;23(3):60-64 © 2011 by the Iranian Society of Ophthalmology

Introduction

Orbitofrontal cholesterol granuloma is a rare disease of orbit with progressive destruction of frontal bone and intracranial invasion, sometimes misdiagnosed as malignancies. It is also called organized hematocele.¹ This lesion usually arises in the superior temporal part of the orbit and results in diplopia and proptosis. The granuloma contains cholesterol clefts with surrounding giant cells in addition to granulation tissue and capillaries.¹

Few cases have been reported in the literature and recurrent cases are even rarer.^{1,2} We report two patients with

orbitofrontal cholesterol granuloma, one of them was a recurrent case, and discuss the clinical, imaging, surgical and pathologic features.

Case reports

Case 1

A 50-year-old man was referred to oculoplastic clinic in Rassoul Akram hospital. The main presenting symptoms were progressive proptosis of the left eye and diplopia in up gaze since 2 years ago. The lesion was not painful.

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Received: March 6, 2011

Accepted: August 4, 2011

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No history of systemic disease was reported. The patient was asked specifically for any previous trauma including multiple trauma or skull or eye trauma and no positive history was found.

Best corrected visual acuity (BCVA) was $20/20$ in both eyes without afferent pupillary defect. On external examination 3 mm proptosis and 3 mm inferior displacement of the left globe were noted. The left eye supraduction was restricted. Intraocular examinations were normal.

The computed tomography scanning (CT) revealed a cystic lesion in superior lateral part of the orbit extending from the rim up to the mid orbit (measuring 30x30 mm) with pushing effect on the globe (Figure 1). There was destruction of the frontal bone and intracranial extension of the lesion. The lacrimal gland and frontal sinus seemed intact. In magnetic resonance imaging (MRI), the lesion was hyperintense and attached to dura on both T₁ and T₂ views (Figure 1).

Orbitotomy was performed completely and the cyst walls were removed. There was a brown appearing cystic lesion with brown liquid contents as well as some solid components. No perioperative complications occurred.

In pathologic evaluation, cholesterol clefts with surrounding giant cells, compatible with cholesterol granuloma was reported. No epithelial components were found. Two years after surgery proptosis and supraduction limitation were resolved.

Case 2

A 20-year-old woman was referred with the complaint of right eye proptosis and diplopia in

up gaze since 6 months ago. She was specifically asked for history of skull or orbital trauma and the result was negative. BCVA was $20/20$ in both eyes and no afferent pupillary defect was detected. On external examination, 4 mm proptosis, 5 mm inferior displacement and 4+ supraduction restriction of the left globe were noted. Intraocular examinations were negative except for mild disc edema in the left eye.

On CT scans a cystic lesion in the superior part of the orbit (30x35 mm) with inferior displacement of the globe, associated with destruction of the frontal bone and intracranial extension of the lesion was detected. Lacrimal gland and frontal sinus seemed intact. The size of the tumor on the last CT scan of 6 months before was 20x30 mm. On MRI, the lesion was hyperintense and attached to the dura (Figure 2).

She reported a similar history which she had 5 years before at age of 15 years. The CT scan of that date showed a superior orbital cyst measuring 30x27x18 mm with frontal bone destruction. The diagnosis of lacrimal duct cyst had been imposed at that center and an incisional biopsy and aspiration had been performed. The pathological report included cystic wall structures with cholesterol clefts, foreign body granuloma and ossification. All signs and symptoms had been resolved after the first surgery, but recurred 5 years later. Considering the diagnosis of orbitofrontal cholesterol granuloma, orbitotomy and tumor removal was performed. Cyst contents were brown color liquid with some solid yellow components. Pathology report of cholesterol clefts surrounded by histiocytes confirmed the diagnosis.

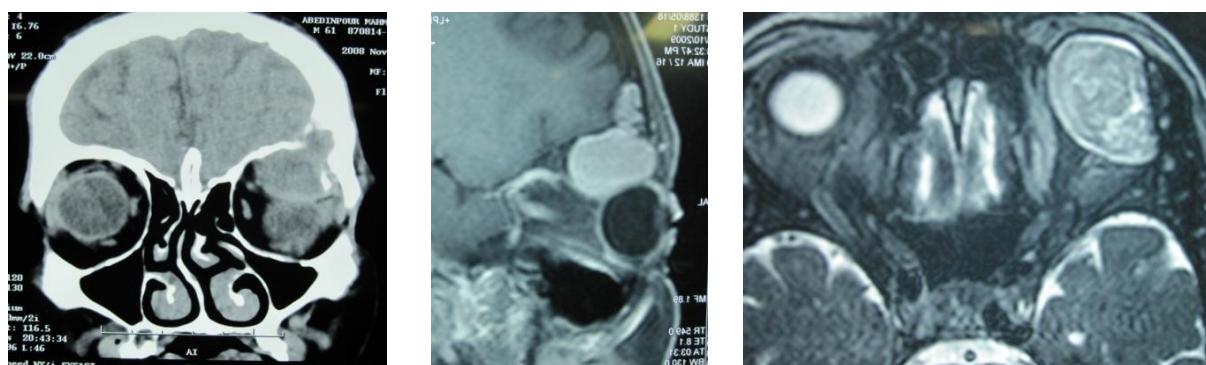


Figure 1. Coronal CT (left), sagittal and axial MRI (middle & right) of a 50-year-old man showing frontal bone destruction and intracranial invasion in orbitofrontal cholesterol granuloma



Figure 2. Coronal MRI (left) and CT scan (middle and right) of a 20-year-old woman with recurrent cholesterol granuloma at the time of second surgery

At the last visit (6 years after surgery) proptosis and diplopia remained resolved. Ocular motility was normal and no complication was noted. No recurrence was observed on orbital CT scan.

Discussion

Cholesterol granuloma has been reported in different organs such as peritoneum, breast, etc. but the most common site is adjacent to skull bones specially temporal bone and petrous apex.¹ Denig³ was the first to report the orbitofrontal cholesterol granuloma in 1902 and few cases were reported thereafter. Most of the reported cases are middle age men (30-50 years) and the lesion almost always has been found in the superior temporal part of the orbit with involvement of frontal bone. There are some reports of primary paranasal sinus lesions with secondary orbital involvement.⁴

Several theories have been offered for orbitofrontal cholesterol granuloma formation.¹ The first theory was hemocele formation and according to this theory, orbital cholesterol granuloma was also named as organized hemocele.^{1,5} It is usually caused by trauma in the subperiosteal part of the frontal bone. McNab and Wright⁵ reported 27 patients with orbitofrontal cholesterol granuloma and seven cases (23%) had past history of trauma, but in Eijpe⁶ report of 11 cases, no one had history of trauma. Other theories are; hemocele formation secondary to vascular anomalies such as hemangioma, primary abnormal bone lesions and primary pagetoid lesions.⁷⁻⁹ It is not clear whether osteolysis is secondary to hemocele or vice versa. For example in

Arat¹⁰ report, a small frontal intraosseous lesion was seen in the first stage of disease. After hemocele formation if the blood is not absorbed, it may be degraded, and organized. Cholesterol is released from plasma and blood cells break down, crystallized and granuloma reaction is formed around crystals. In addition, blood and bile pigments are released. Granulomatous reaction releases platelet driven prostaglandins, which destruct surrounding soft tissues, and causes bones resorption.¹ Macroscopic finding depends on the stage of blood degradation, the color may be red, brown, yellow, muddy or purple and the contents may be liquid, viscid or semisolid containing yellow crystals.¹¹

Hallmark of microscopic findings are cholesterol clefts and surrounding giant cells (multi-nucleated histiocytes) in addition to granulation tissue and capillaries. Sometimes blood degradation products such as hematoidin (red brown), hemosiderine (brown), occasionally calcium deposits, and some bone particles are found in the progressed stages. The surrounding tissues become hard and a fibrotic wall forms around it. The lesion is cystic but there is no epithelial cells and keratin, so it is a hemocele, not a true cyst as is seen in cholesteatoma.^{12,13}

In cases that have past history of orbital trauma have the latency of 1 month to 3 years from trauma to clinical manifestations.¹⁴ Clinical signs and symptoms depend on the disease stage. For example, there is a report of very small lytic lesion in the frontal bone found by chance in CT scans, on the other hand a very extensive lesion with destruction of frontal bone, sphenoid bones, orbital apex,

lateral and superior orbital wall and cranial and optic canal invasion was reported.¹⁴ The most common symptom and sign is progressive proptosis, although diplopia, pain, decreased visual acuity (VA) due to induced astigmatism and palpable orbital tumors have been reported. In case of intracranial invasion, there may be central nervous symptoms such as headache and convulsion.¹⁵ There is the chance of rapid progression due to rebleeding in the lesion.¹⁶

In imaging examinations, MRI may better show soft tissues such as dura and brain, hematoma, abscess, and dermoid cyst, but CT scan shows bone lesions better, so usually both modalities are needed. On CT scan, there are well-delineated cystic lesion with medium density (hypodense or isodense with brain) without enhancement, in addition, osteolysis of superior orbital wall with smooth bone erosion but absence of sclerosis (pathognomonic of dermoid cysts) is evident.^{6,14,17} Sometimes several small bony fragments and calcifications are found. On MRI, usually there are characteristic high signals in both T1 and T2 without enhancement.¹⁸ Sometimes MRI findings are non-characteristic.¹⁹

There are reports of orbital B scan, bone scan and angiography for evaluation of cholesterol granuloma but their findings are not specific in comparison to CT scans and MRI.²⁰

Two most important differential diagnoses are dermoid cyst and malignant tumors. Orbital dermoid cysts are usually dumbbell shape located between orbit and temporal fossa, whose bony defect has characteristic regular sclerotic margins.²¹ It may be homogenous or heterogeneous and does not enhance (except its capsule), the most characteristic MRI sign is hypointensity in T1 and T2 in comparison to brain.²²

Malignancies usually are not cystic but may be circumscribed. They cause bone erosion, usually moth eaten and irregular. Clinically there is pain and usually lacrimal gland enlargement or involvement. On MRI they are hyperintense in comparison to the brain and are usually enhanced with contrast.¹⁴

Other lesions such as aneurismal bone cyst, ossifying fibroma and eosinophilic granuloma are seen in children with their characteristic findings easily differentiating from orbitofrontal cholesterol granuloma.²³

Considering progression of the disease, all cases require surgical removal. The usual technique is superior orbitotomy via upper lid crease approach with cyst aspiration and curettage and complete drainage.⁵ Some authors believe that complete curettage is not necessary,^{5,7,9} Some authors advice complete removal of cyst^{2,22,24} and some advice that craniotomy is prudent if there is sphenoid or intracranial involvement.²⁴ Sometimes by orbitotomy approach and dilation of bone defects, curettage of intracranial cyst can be performed. Usually bone defects have thin blue-yellow margins.²⁵ Also, endoscopic surgery through skin approach has been reported.²⁶

Prognosis depends on the extension and severity of associated lesions. Usually recurrence is rare by complete removal of the cyst^{5,22} but there is chance of recurrence in incomplete removal.^{1,2,14}

Conclusion

Orbital cholesterol granuloma is a rare, progressive disease, usually with frontal bone destruction and intracranial invasion. It must be differentiated from dermoid cyst and malignancies. Imaging findings (CT scan and MRI) are very important to have accurate diagnosis before surgery. It must be managed surgically, usually with orbitotomy. Success rate is high, and recurrence is rare.

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