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Case Report

An atypical case of compressive optic neuropathy and cranial nerve 6th palsy caused by a cholesterol granuloma

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Abstract

In this case report we present an unusual case of orbital cholesterol granuloma associated with compressive optic neuropathy and cranial nerve 6th palsy. Cholesterol granuloma results from a foreign body response to the presence of crystallized cholesterol. Cholesterol granuloma affecting the orbit are a rare presentation as they typically occur in the petrous apex of the temporal bone. Computerized tomography scan and magnetic resonance imaging are extremely valuable in the diagnosis of cholesterol granuloma as they have characteristic radiological features. The treatment of cholesterol granulomas is based on the presentation and the symptoms and may include active surveillance versus surgical drainage.

Cholesterol granuloma (CG) is a slow-growing benign lesion which results from a foreign body giant cell reaction to the presence of cholesterol deposits as a result of breakdown of blood products [1]. It is more common in men and presents with gradual mass effect [2]. CG typically occurs in the middle ear, mastoid antrum and petrous apex. Less frequently it has been reported in the frontal bone, zygoma, and paranasal sinuses [2].

The exact pathophysiology of CG is not well understood. However, several mechanisms including trauma as well as ventilatory obstruction in the bony cavity resulting in a negative pressure and subsequent haemorrhage and cholesterol deposition has been proposed [2,3]. The alternative hypothesis is the exposed marrow theory where the hyperplastic mucosa invades the underlying bone resulting in bleeding and chronic inflammatory response as the blood is metabolized [4].

CG has characteristic appearance on imaging. CG appears as a well-marginated lesion, isodense within the brain, usually with evidence of bony erosion on the CT. CG have a high signal intensity on T1-weighted sequence of the MRI due to their cholesterol component and methemoglobin content [3,5,6]. On T2-weighted sequence of the MRI, CG demonstrate central high signal which does not attenuate on FLAIR sequence. Furthermore, they do not demonstrate fat suppression and have no enhancement post-gadolinium [3,5,6].

In this case report we describe an atypical presentation of CG associated with compressive optic neuropathy and cranial nerve 6th palsy.

Case Report

A 33-year-old female presented to the emergency department with a 4-day history of gradually worsening headache, nausea, vomiting and left sided retro-orbital pain. Her past medical history included iron deficiency anaemia. There was no history of head trauma, previous head and neck surgery, proptosis, double vison or decrease in visual acuity.

On examination her visual acuity was 6/6 in each eye. Initially extraocular movements were normal and fields to confrontation were unremarkable. Pupils were reactive to light and accommodation and there was no evidence of a relative afferent pupillary defect (RAPD). Intraocular pressures using non-contact tonometer were 14/16 mmHg right/left eye. Anterior segment examination was unremarkable and fundoscopy revealed normal optic nerves and retina. In the following 4 hours; while being managed with analgesia, and antiemetic and awaiting the result of the CT brain, she developed double vision with

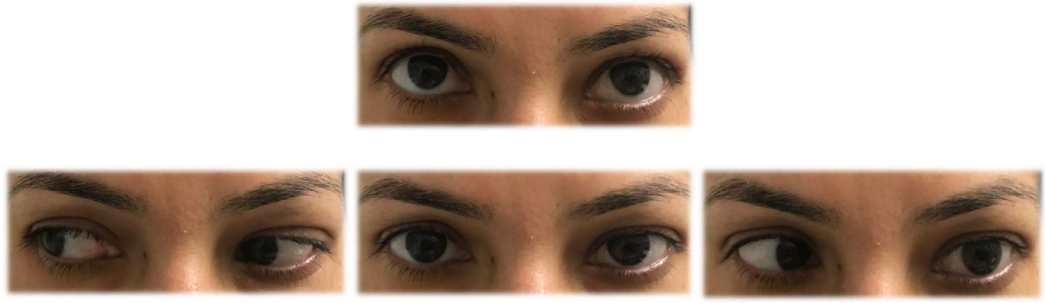
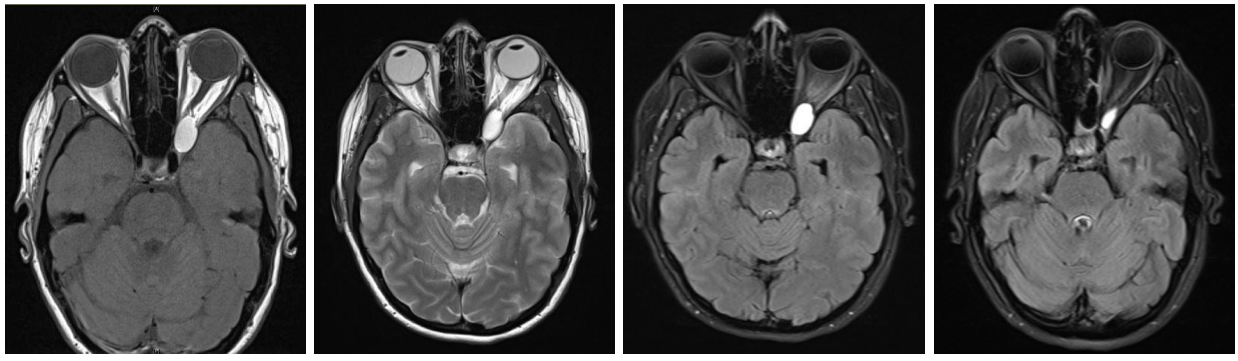


Figure 1: Examination of ocular motility during the presentation demonstrating a left abduction deficit.

a complete left 6th cranial nerve palsy. There was no evidence of "RAPD". Figure 1. Demonstrates her ocular movement and left 6th cranial nerve palsy.

CT brain demonstrated a non-enhancing homogenous left orbital apex soft tissue mass, measuring 2.0 x 1.0 x 1.3 cm, and associated bony remodelling of the lateral wall of left sphenoid sinus and superior displacement of the optic nerve. Patient was transferred to a tertiary hospital and admitted under ophthalmology team. Within 24 hours the patient developed left RAPD. Colour vision on Ishihara was normal in each eye. Red saturation was decreased to 90% in the left eye. Cover test demonstrated left esotropia. MRI of the brain and the orbit with contrast on day 1 admission demonstrated an ovoid lesion centred at the left superior orbital fissure and orbital apex. The lesion extended into orbit via superior orbital fissure, which was secondarily widened. It had mass effect on the optic nerve, deviating it superiorly and medially. The lesion was hyperintense on T1 and T2, with no suppression on FLAIR or fat suppressed sequences. There was no restricted diffusion, nor appreciable enhancement post contrast and no dural tail. Internal signal characteristics were in keeping with complex fluid. The radiologic characteristic of the lesion was thought to be most consistent with CG with differential diagnosis of a trapped complex left sphenoid sinus fluid. Other differentials for CG are dermoid or epidermoid cysts [7]. On MRI, dermoid cysts have the same brightness as subcutaneous fat on T1 and T2 weighted images due to their sebaceous secretions [7]. However, they become dark on fat-supressed images [7]. On the other hand, dermoid cysts are generally hypointense on T1 weighted images and hyperintense on T2 weighted images [7]. Figure 2. (A-C) Demonstrates the findings of the MRI on admission.

The patient was referred to the Ear Nose & Throat surgical team and underwent functional endoscopic sinus surgery (FESS) and decompression of the left orbital apex lesion. Intraoperatively the lesion's appearance was consistent with CG. Unfortunately, a sufficient sample could not be obtained for pathology. Postoperatively the patient was managed with dexamethasone 8 mg twice a day for two days, followed by dexamethasone 4 mg twice a day for 3 days. She was also prescribed cephalexin 500 mg twice a day for 1 week. The postoperative MRI confirmed a reduction in the size of the apical lesion. The patient's diplopia and abduction deficit resolved over the next three months. Figure 2D demonstrate the MRI images post-surgery.



A.T1. B. T2 C.T2 flare D. T2 Flare – post FESS

Figure 2: A) Unenhanced MRI brain axial view T1-weighted image demonstrate an hyperintense lesion causing compression on the left optic nerve at the orbital apex. **B)** MRI brain axial view T2-wighted image also demonstrate an hyperintense lesion. **C)** MRI brain axial view post Gadolinium contrast with nil further enhancement of the lesion. **D)** MRI brain axial view post Gadolinium contrast following functional endoscopic surgery (FESS) demonstrating a reduction in the size of the lesion with less mass affect on the optic nerve.

In Summary

This case is a rare presentation of CG of the orbit resulting in a compressive optic neuropathy and cranial nerve palsy arising from the sphenoid sinus. The diagnosis of GC can be challenging as it is extremely rare. However, it should be considered in the cases of compressive optic neuropathy and cranial nerve palsy. Imaging, in particular MRI, can be extremely valuable in the diagnosis of CG. Management of CG depends on the presentation and progression of symptoms and includes conservative therapy (with serial imaging and neurological exam) in the case of incidental finding or minimally symptomatic patients, steroids, and surgical excision [8].

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