CASE REPORT

Compressive Optic Neuropathy Caused by a Cholesterol Granuloma of the Sphenoidal Sinus

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ABSTRACT

Cholesterol granuloma is a foreign body giant cell reaction to cholesterol deposits. The pathology is rarely seen in the paranasal sinuses. In this report we describe a case of compressive optic neuropathy caused by cholesterol granuloma arising from the sphenoidal sinus. A 46-year-old lady presented with painless progressive decrease in vision in her left eye over the past 7 years. Magnetic resonance imaging of the brain and orbit with gadolinium enhancement showed a homogenous T1 and T2 hyperintense lesion causing expansion of the sphenoidal sinus compressing the ethmoidal sinus and also the left optic nerve. An endoscopic marsupialisation of the lesion was performed. Histopathological examination showed multiple characteristic cholesterol clefts surrounded by numerous multinucleated giant cells, histiocytes, lymphocytes, haemosiderin-laden macrophages, and plasma cells. This case emphasises the importance of appropriate neuro-imaging in cases of progressive optic neuropathy: cholesterol granuloma of the paranasal sinuses is a rare cause of optic nerve compression.

KEYWORDS cholesterol granuloma, compressive optic neuropathy, sphenoidal sinus

INTRODUCTION

A case is described of compressive optic neuropathy due to a cholesterol granuloma of the sphenoidal sinus. The patient had experienced two episodes of transient visual loss with spontaneous recovery, mimicking optic neuritis.

CASE REPORT

A 46-year-old woman presented with painless progressive decrease in vision in her left eye over the previous 7 years. She was a known case of type II diabetes mellitus, hypertension, and migraine. For diabetes she was taking oral hypoglycaemics and calcium channel blockers for hypertension and was under good control. For migraine she was not on any medication. She gave a history of two episodes of painful reduction of vision 2 years previously in the left eye associated with headache that spontaneously recovered in 2 weeks. During these episodes neuro-imaging was not performed and visual loss was

ascribed to migraine. Details of clinical examination during these episodes were not available.

On examination, the best corrected visual acuity in the right eye was 20/20 and there was no perception of light in the left eye. Colour vision was normal in the right eye. Extraocular movements were normal in both eyes. Anterior segment examination of the left eye showed an amaurotic pupillary response. There was a trace of posterior subcapsular cataract in both eyes. Sensation along all three divisions of the Vth cranial nerve was normal bilaterally. The optic nerve head in the right eye was healthy and on the left showed primary optic atrophy. Automated visual field analysis (Humphrey 30-2) was normal in the right eye.

The following differential diagnoses were considered:

1. Recurrent retrobulbar neuritis: Due to history of two episodes of painful sudden loss of vision in a middle aged lady, which recovered spontaneously, recurrent retrobulbar neuritis was thought of as the initial differential diagnosis.
2. Compressive optic neuropathy: In view of the finding of no perception of light and non-recovery

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of vision, the possibility of left compressive optic neuropathy was also considered.

Magnetic resonance imaging (MRI) of the brain and orbits with gadolinium enhancement was carried out (Figure 1A, B; enhanced scans not shown as not available). A homogenous T1 and T2 hyperintense lesion causing expansion of sphenoidal sinus compressing the ethmoidal sinus and also left optic nerve was revealed. The sella was seen separate from the lesion. No heterogeneity was noted in the mass.

The differential diagnoses based on MRI findings were

1. Sphenoidal sinus mucocoele
2. Mucormycosis arising from sphenothmoidal sinuses
3. Sinus malignancy with intracranial extension causing compressive optic neuropathy
4. Non-ossifying fibroma

The patient was referred to an ENT surgeon. On examination the ENT surgeon made a provisional diagnosis of sphenoidal mucopyocoele. An endoscopic marsupialisation of the lesion was done and the specimen was sent for histopathological examination.

Gross examination of the specimen showed a fleshy white to grey tissue covered with mucosa. Sections of the tissue showed pseudostratified ciliated columnar epithelium. There were large areas of haemorrhage and fibrosis with presence of multiple characteristic cholesterol clefts surrounded by numerous multinucleated giant cells, histiocytes, lymphocytes, haemosiderin-laden macrophages, and plasma cells (Figure 2).

A final diagnosis of cholesterol granuloma arising from the sphenoidal sinus causing left compressive optic neuropathy was made. Postoperatively vision in the left eye improved to hand movements close to face and was maintained at this level until the most recent visit at time of writing, which was in the fourth post-operative month. Repeat MRI brain showed no residual lesion.

DISCUSSION

Compressive optic neuropathy (CON) usually causes painless, progressive, gradual loss of visual function, a relative afferent pupillary defect, and optic disc oedema or atrophy.¹ Various intracranial, intraorbital, and paranasal sinus lesions can cause CON. Cholesterol granuloma (CG) is a foreign body giant cell reaction to cholesterol deposits, with associated fibrosis and vascular proliferation, commonly seen in the temporal bone or middle ear cavity.² Its occurrence in paranasal sinuses is rare.³

In this report we present a case of CON caused by CG arising in the sphenoidal sinus. A MEDLINE search yielded only one similar case report by Hwang et al.⁴ In that case, the visual acuity in the affected eye was 20/20 but there were other signs of compressive optic neuropathy such as defective colour vision, relative afferent pupillary defect, temporal optic disc pallor, and visual field defects. The diagnosis of cholesterol granuloma was confirmed only on histopathology of the excised lesion. Visual field defects improved significantly after excision of the lesion.

In cases with unexplained optic atrophy, CON should be considered as a differential diagnosis and contrast-

FIGURE 1. (A) Unenhanced MRI brain T1-weighted image (axial view) showing hyperintense lesion causing expansion of sphenoidal sinus wall compressing the left optic nerve at the orbital apex. The right optic nerve is seen free from the lesion. (B) MRI brain T2-weighted image (sagittal view) showing the lesion to be separate from sella. Note that the lesion is also hyperintense on this T2-weighted sequence. Gadolinium-enhanced images were carried out but were not available for review.

FIGURE 2. Histopathology of the excised lesion (H&E stain, 10×) showing cholesterol clefts (*) surrounded by numerous multinucleated giant cells (small arrow), histiocytes, lymphocytes, haemosiderin-laden macrophages, and plasma cells.

enhanced MRI of brain and orbit performed. This may reveal unusual causes of CON such as CG, as in this case.

CG represents the end result of a variety of pathways for cholesterol deposition. The major factors thought to be responsible for the formation of a CG are firstly a poorly aerated, obstructed bony cavity and secondly intramucosal oedema and haemorrhage that fails to get resorbed. The degenerated red blood cell membrane precipitates in a crystalline form and incites a foreign body granulomatous reaction.⁵

The clinical presentation of the CG varies depending on its location and size. Computed tomographic (CT) scanning and MRI play a significant role in the diagnosis of CG. On CT scan, it appears as a well-marginated lesion, isodense with the brain, with no significant contrast enhancement; bone erosion may be present. The typical findings on MRI scan are lesions with areas of high signal intensity on both T1- and T2-weighted

images (characteristic of chronic haemorrhage) interspersed with either septations or foci of low signal intensity.²

Treatment of CG depends on the location and size of the lesion. Options include conservative therapy (serial neurological exams and MRI), steroids, or surgical excision. Surgical removal of the cyst, proper drainage, and permanent ventilation may prevent recurrence.⁶

We report a rare case of compressive optic neuropathy caused by cholesterol granuloma of the sphenoidal sinus. This case reinforces the need for appropriate neuroimaging in cases with unexplained optic atrophy.

Declaration of interest: The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

Note: Figures 2 of this article is available in colour online at www.informahealthcare.com/oph

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