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Frontal sinus cholesteatoma: a rare but important clinical entity to remember

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Abstract:

We aim to discuss the clinical presentation, investigations and successful management of frontal sinus cholesteatoma. A 26 year old Caucasian female, otherwise fit and healthy, presented with a 4-month history of swollen right eye and sudden visual impairment. The ophthalmological examination revealed right sided proptosis, diplopia and reduced visual acuity. The colour vision was normal and there was no afferent papillary defect. On nasendoscopy, the nasal cavity was unremarkable apart from some fullness at the area of the right uncinate process. Computerized tomography scan identified a space occupying lesion in the right frontal sinus eroding through the orbital roof and displacing the right globe. There was also extensive bony erosion through the posterior table. The patient underwent right sided endoscopic sinus surgery and fronto-ethmoidectomy via an external approach. Intra-operatively, the right frontal sinus was found to be full of keratin. The histological examination showed sheets of keratinous debris. She made a good post-operative recovery and remained disease free at 3 years follow-up. The frontal sinus cholesteatoma is a rare condition but should be included in the differential diagnosis of a slowly expanding lesion occurring in the frontal sinus.

Introduction:

The cholesteatoma or more accurately known as keratoma of the frontal sinus is a rare entity with variable frequencies and presentations described in the world literature. The clinical symptoms and complications are determined by the anatomical location and growth pattern of the disease, however, the true incidence of keratomas in the paranasal sinuses is thought to be lower than reported as some of the cases are believed to be cholesterol granulomas1, 2.

Objectives:

In this article, we describe a case of frontal sinus keratoma and discuss the clinical presentation, investigations and successful management of the disease.

Methods

A case report with literature review

Case Report:

A 26 year old Caucasian female, otherwise fit and healthy, presented with a 4-month history of swollen right eye and sudden visual impairment. There was no history of frontal headache, nasal discharge, facial pain or neurological complaints. There was no history of external frontal trauma nor previous sinonasal or ocular surgery.

The ophthamological examiniation revealed right sided proptosis and inferior displacement with restriction of upward gaze. Her visual acuity for the right and left eye was 6/36 and 6/4.8 respectively. The colour vision was normal and there was no afferent papillary defect. On nasendoscopy, the nasal cavity was unremarkable apart from some fullness at the area of the right uncinate process. The computerized tomography scan identified a space occupying lesion in the right frontal sinus eroding through the orbital roof and displacing the right globe (Fig. 1). There was also extensive bony erosion through the posterior table.

The patient underwent right sided endoscopic sinus surgery and fronto-ethmoidectomy via an external approach. Intra-operatively, the right frontal sinus was found to be full of keratin. The frontal sinus was thoroughly debrided, mucosal lining completely removed and the sinus tracts exenterated and smoothened with a diamond burr. An external drain was left in situ for 48 hours. She made a good post-operative recovery with resolution of her ocular symptoms. The histological examination showed sheets of keratinous debris (Fig 2). The microbiology analysis showed no pus and no organisms were grown culture. Her interval MRI imaging scan demonstrated minor opacification in the frontal sinus, but no disease recurrence. She remained disease free at 3 years follow-up.

Discussion:

Epidermoid cyst is a keratin filled cyst lined with squamous epithelium. It was first reported by Pinson in 1807 and a reproduction in Cruveilhier's Atlas in 1829, Cruveilhier described the 'tumeur perlee' referring to 'cholesteatomie' 3, 4. In 1838, Johan Muller noted these tumours to be made of layers of tissue derived from squamous epithelium and Virchow (1954) claimed that 'cholesteatoma' was a misnomer as cholesterol was not part of the lesion5, 6. Despite the pathologic separation of cholesterol from keratin, the term cholesteatoma has continued being used for lesions that do not contain cholesterol.

In the head and neck region, keratomas are classified into primary and secondary types. The primary keratomas are referred to as epidermoid cysts originating from congenital epithelial cell nests. The secondary keratomas occur due to squamous epithelial cell migration or implantation beyond its natural boundaries. This could be due to trauma or sequelae of any process resulting in sequestration of normal squamous epithelial cells in abnormal areas 2. Post-traumatic cholesteatomas differ from congenital keratomas as they are often associated with cholesterol granulomas resulting from degeneration of red blood cells. In the paranasal sinuses, keratomas are most often located in the frontal sinus and less commonly in the ethmoids and maxillary sinuses 7.

Characteristically, these tumors present as a unilateral, painless, slow-growing mass, often accompanied by frontal headache, proptosis and diplopia, and without a history of frontal sinus infection1, 2. As the keratoma enlarges, erosion of the bone occurs in all directions leading to forehead deformity. Our patient experienced diplopia and reduction in visual acuity secondary to pressure effect on the globe. In some cases unilateral headaches may be the only presenting symptom as a result of erosion of the posterior frontal sinus wall causing pressure effect on the dura, arachnoid and frontal lobe. Our patient did not experience any frontal headaches.

The CT scan of paranasal sinuses, therefore, plays an important role in the evaluation of the disease extension and helps surgical planning. Magnetic resonance imaging (MRI) can be used to differentiate sinusitis from a tumour or a soft tissue lesion 8.

The differential diagnosis includes mucocele, fungal infection, osteoma, fibroma, dermoid cysts, vascular abnormalities and malignant tumours in the frontal sinus. The mucocele is commonly associated with history of sinusitis 9. The CT scan would demonstrate a smooth, expansile lesion within the frontal sinus with inferior globe displacement. Fungal sinusitis presents with signs and symptoms of sinusitis or nasal polyposis. The presence of thick tenacious fungal mucin is often characteristic of allergic fungal sinusitis. The CT scan would demonstrate a heterogeneous opacification with mixed volume density and microcalcification. The osteomas have distinctive bony appearance radiographically. The Fibroma and dermoid cysts can present with the same symptoms and clinical findings as keratomas and are indistinguishable from epidermoids radiographically. Most malignant tumours in the region grow rapidly and give more intense pain and neurological

symptoms due to tumour mass pressure effect. The CT scan would demonstrate a destructive lesion often with bony erosion.

The curative treatment with total excision of the keratoma with adequate drainage 1, 2, 7, offers excellent prognosis although late recurrence has been reported 10. The osteoplastic frontal sinus obliteration procedure has been described to remove the disease successfully to provide a satisfactory cosmetic result 1, 2, 8. With limited anterior table erosion, reconstruction using titanium plate, mesh wire or methacrylate is recommended. In cases where there is erosion of the posterior table, frontal sinus obliteration with abdominal fat may be considered1, 2. In cases of isolated orbital roof defect, as seen in this patient, reconstruction of the orbital roof is deemed not necessary. In recent years, endoscopic approach with image-guidance had been described 11. Lai et al. described successful surgical resection of an extensive frontal sinus keratoma via a modified endoscopic Lothrop 12. Radical approaches, including craniotomy and dura resection of adherent cholesteatoma followed by primary repair with fascia lata graft, may be required in extensive disease 10, 13. It is recommended that all sinus skin tracts to be completely excised to avoid recurrence. In this case, the patient underwent a combination of endoscopic and external fronto-ethmoidectomy to remove the disease. This method provides good access to the area of concern, less morbidity and satisfactory cosmetic outcome. Untreated sinus cholesteatoma can lead to severe disfigurement, carcinomatous degeneration or death 7, 14, 15.

Patients should be followed up for a period of at least two years following surgery. Diffusion-weighted MRI may be considered when following up these patients to identify any recurrence.

Conclusion:

Frontal sinus keratoma is a rare condition but should be included in the differential diagnosis of a slowly expanding lesion occurring in the frontal sinus, especially in the presence of bony destruction. The management of both primary and secondary keratomas of the frontal sinus involves removing the disease completely and obliteration of the sinus.

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Legends

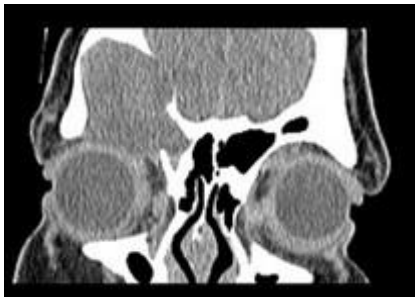

Figure 1: CT scan confirming right frontal sinus opacification with destruction of the orbital roof and adjacent skull base

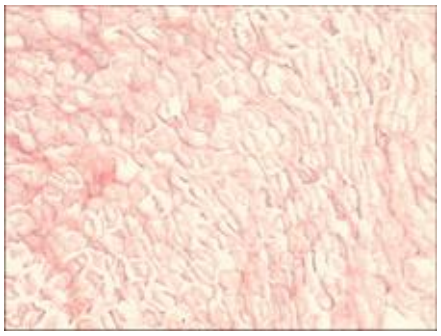

Figure 2: Sheets of keratinous debris without nucleated cells or evidence of intact squamous epithelium (cholesteatoma)