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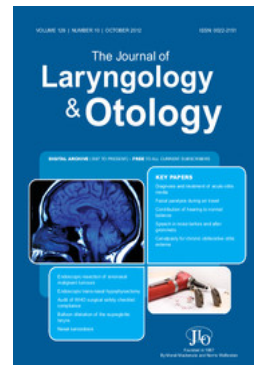
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Primary chordoma of the ethmoid sinus

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Abstract

Primary chordoma of the paranasal sinuses are extremely rare tumours, with only a small number of cases verified and published in the literature. It appears that only five of these cases involved the ethmoid sinuses either as a primary or by local invasion, and of these documented cases only one other has been found to solely involve the ethmoid sinus. We present a case of primary ethmoid sinus chordoma treated by wide local surgical excision and present a review of the literature with regard to prevalence and treatment rationale.

Key words: Chordoma; Ethmoid Sinus

Introduction

Chordomata are uncommon dysontogenic malignant neoplasms that are thought to arise either from residual remnants of the embryonic notochord or from ectopic chordal nodules.^{1,2} There are three main sites of occurrence: spheno-occipital, vertebral and sacro-coccygeal, of which approximately one third arise in the spheno-occipital,³ and it is amongst this group that the otolaryngologist may encounter this disease. Among the commoner presentations include the presence of a nasopharyngeal mass from direct spread from a lesion of the clivus. Presentation as a solitary lesion affecting one or more paranasal sinuses is extremely rare and is usually discovered by the pathologist. Table I shows the five cases of chordoma involving the ethmoid sinus that have been reported,^{4–7} with only of those confirmed as solely involving the ethmoid sinus.⁴ An additional case is presented and the radiographical findings and treatment rationale discussed.

Case report

A 42-year-old woman presented initially to the ophthalmic surgeons with a one month history of discomfort and swelling at the medial canthus of the left eye; she had no overt nasal symptoms. Examination confirmed a soft swelling at the medial canthus with no other ophthalmic signs. A computed tomography (CT) scan revealed opacification of the left frontal and ethmoid sinuses with a suggestion of some destruction of the medial orbital wall (Figure 1). Coronal MRI with gadolinium demonstrated enhancement of the tumour (Figure 2). She was referred for an ENT opinion and underwent an endoscopic examination and biopsy. This revealed a polypoidal lesion in the left fronto-nasal recess, which proved histologically to be a chordoma (Figure 3).

The patient underwent a lateral rhinotomy approach that showed the tumour filling the anterior ethmoid sinuses, extending into the middle turbinate and abutting the adjacent orbital periosteum. The ethmoid sinuses were cleared and the middle turbinate was dissected off the skull

base, the frontal sinus was opened and the mucosa stripped, and the affected area of orbital periosteum was excised.

The patient made an uneventful post-operative recovery and was discharged two days later. She has had a 12 month follow-up by endoscopic examination and magnetic resonance imaging (MRI) that has failed to show evidence of recurrence. However, the patient has been counselled that if recurrence occurred periorbitally, an orbital clearance might be required.

Discussion

A chordoma is a rare, malignant, slow-growing tumour that can present in the nasal cavity by anterior extension from the clivus involving the ethmoid sinuses. Isolated chordomas of the paranasal sinuses are extremely rare. Only five other cases in the English literature have been described, four of which occurred by direct extension from an adjacent sinus.

Virchow⁸ and Luschka⁹ independently described this lesion and at that time felt it most likely represented a tumour of cartilaginous origin. This view was held until Muller¹⁰ recognized it as notochordal tissue and coined the term 'ecchordosis physaliphora', because of the characteristic physaliphorous cells.

The presenting symptoms of chordoma depends on the site of involvement; clival lesions usually present late, after intracranial extension, and therefore, cranial nerve

TABLE I
CHORDOMAS INVOLVING THE ETHMOID SINUS IN THE LITERATURE

Number	Number of patients	Site
Berdal and Myhre ⁴	1	Ethmoid
	1	Antro-ethmoid
Wright ⁵	1	Spheno-ethmoid
Perzin and Pushparaj ⁶	1	Spheno-ethmoid
Harrison and Lund ⁷	1	Antro-ethmoid and orbit
Present study	1	Ethmoid

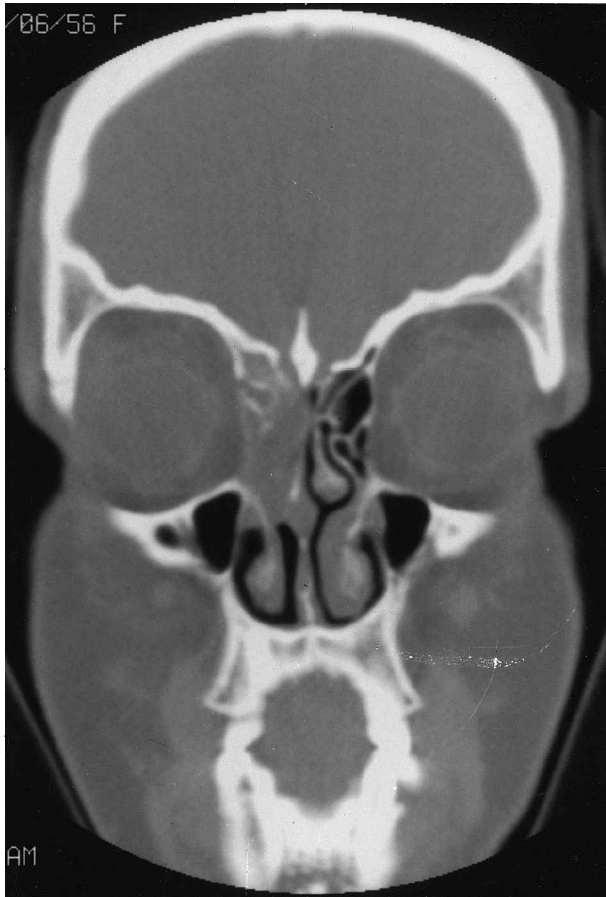


FIG. 1

Coronal CT scan image of the nose and paranasal sinuses showing opacification of the left frontal and ethmoid sinuses with a suggestion of some bone destruction of the medial orbital wall.

involvement and signs of increased intracranial pressure predominate. If the tumour spreads anteriorly, nasal obstruction, mucopurulent discharge and epistaxis may occur.¹¹ Computed tomography (CT) scanning is optimally employed to demonstrate bony destruction although the lesion is best shown by MRI, revealing a high T2 signal and enhancement on T1-post-gadolinium.

Macroscopically, the tumour is lobulated and gelatinous in nature, often with a pseudocapsule and areas of dystrophic calcification. Microscopically, the tumour is variable, and is usually composed of certain proportions of three cell types: stellate cells, intermediate cells, and physaliphorous cells.¹² There is abundant intercellular mucoid matrix, and stains for mucus and glycogen are positive. Immunohistochemistry for cytokeratins and S100 are positive. All the above make the diagnosis difficult and confusion can arise with the most common misdiagnoses being mixed salivary gland tumour, chondroma, chondrosarcoma, mucocele etc.

Treatment of the lesion depends on the site and extension of the disease. Clival or sphenoid lesions are often too advanced for complete surgical resection. However, debulking combined with post-operative radiotherapy is often employed, despite the tumour being relatively radioresistant. The more anterior, ectopic foci occurring in the frontal, ethmoid and maxillary sinuses are far more amenable to full surgical excision and the prognosis therefore appears more favourable. A relatively

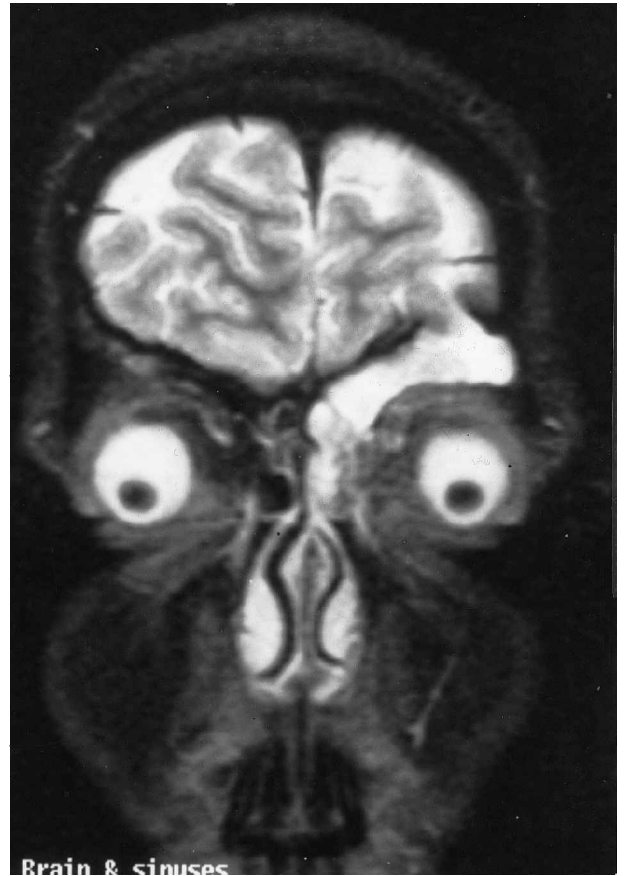


FIG. 2

Coronal MRI scan T1-gadolinium-enhanced image to show the high uptake of the tumour.

large series of cases (six) studied by Berdal and Myhre⁴ showed excellent survival rates with a number of probable cures, and one patient alive with recurrence 25 years following initial diagnosis.

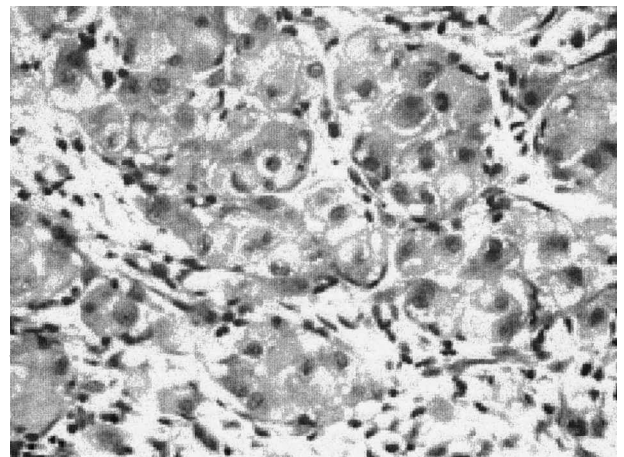


FIG. 3

Pathology. The specimens showed widespread infiltration of a neoplasm composed of small groups of epithelioid cells with nuclear hyperchromasia and moderate nuclear pleomorphism. Many of the nuclei contained intranuclear cytoplasmic inclusions, and the cytoplasm was abundant and eosinophilic. Immunostaining showed positivity for vimentin, EMA, S100 and, was weakly positive for LP34. This pattern of immunoprofile was in keeping with a chordoma. ($\times 400$)

We present an additional case of primary chordoma of the ethmoid sinus, and although the literature is somewhat sparse, we conclude that initial treatment for chordomata presenting in the paranasal sinuses is primary surgical excision; with the understanding that the slow growth rate of the tumour may mean late recurrences, that in themselves may require revision surgery. Radiotherapy has a place to play in such recurrences or in cases where excision is felt to be incomplete.

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