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Primary ethmoid sinus craniopharyngioma: a case report

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Abstract
A seven-year-old boy complained of intermittent epistaxis for several months. Computerized tomography (CT) scans showed increased soft tissue density over the left ethmoid sinus. Endoscopic sinus surgery was used to remove the mass completely. The pathological report was craniopharyngioma. No other focus of lesion was found, and he continues to do well.

Key words: Craniopharyngioma; Ethmoid sinus

Introduction

Craniopharyngiomas are benign cystic tumours (Trippi *et al.*, 1969). They are frequently located in a sellar or suprasellar region (Demaerel *et al.*, 1993). The infrasellar location is unusual. Akimura *et al.* (1989) found only 21 cases reported in the literature. The nasopharynx is the most common site of infrasellar craniopharyngiomas (Barnes and Peel, 1985). Graziani *et al.* (1994) reported 15 cases of nasopharyngeal craniopharyngioma in his paper. Other infrasellar locations have also been reported (Akimura *et al.*, 1989). However, a craniopharyngioma, which is primarily located in the ethmoid sinus, has not been found in the literature. Here, we report such a rare case.

Case report

A seven-year-old boy suffered from intermittent epistaxis for several months. He visited a local hospital, and a nasal biopsy was performed. The pathology was reported to be inverted papilloma. He was transferred to our department for further management. In our department, no nasal tumour was seen in the nasal cavity during the physical examination. A CT scan was carried out that showed increased soft tissue density over the left ethmoid sinus. A tumour mass with calcification seemed to be discernible (Figure 1). This lesion did not involve other neighbouring sites.

Endoscopic sinus surgery was undertaken to remove the lesion on August 29, 1995. Multiple pieces of bony and soft tissues were taken from the left ethmoid sinus (Figure 2). The ethmoid cavity was completely cleared. The left sphenoid, frontal, and maxillary sinuses were also checked, and were all free from the lesion. The pathological report found it to be a craniopharyngioma. Histopathologically, the tumour consisted of submucosal anastomosing epithelial islands with a palisaded basal layer and a centre of stellate cells (Figure 3), which are characteristic of craniopharyngioma. Cystic degeneration, focal calcification, foci of squamous metaplasia with solid nests of keratinization, and a mixed inflammatory reaction were also found in the specimens (Figure 4). A punch biopsy

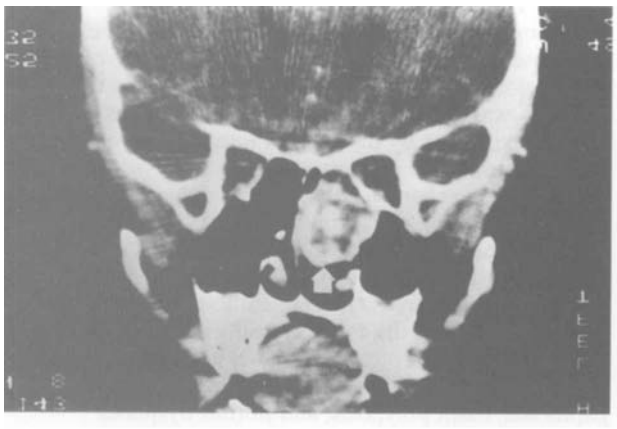

FIG. 1
A pre-operative CT scan showing a tumour mass (arrow) with calcification in the left ethmoid sinus.

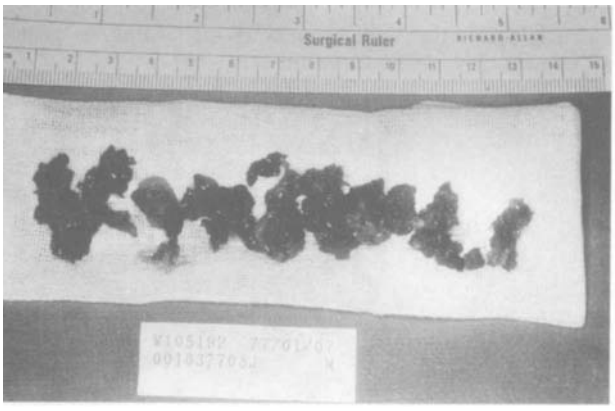

FIG. 2
Multiple pieces of bony and soft tissues were taken from the left ethmoid sinus.

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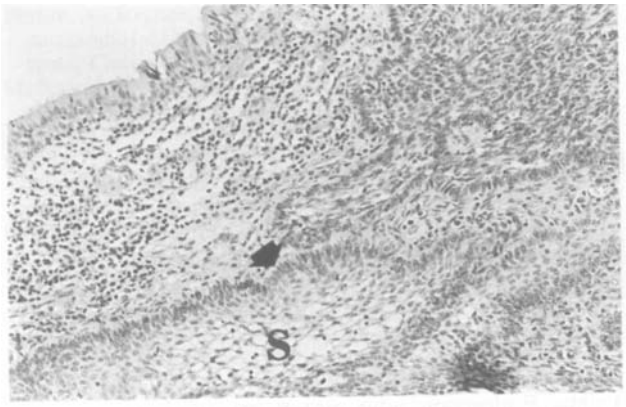

FIG. 3

Submucosal anastomosing epithelial islands with a palisaded basal layer (arrow) and a centre of stellate cells (S) shown (H & E; × 200).

was also done over the nasopharynx. The pathological report was chronic inflammation.

After surgery, this boy did well. A follow-up CT was done two months later. The left ethmoid sinus was free from any tumour mass (Figure 5). Only some high density was still seen in the left middle turbinate. One year after the operation, a flexible nasal endoscopy did not find any tumour mass in the nasal cavity.

Discussion

Craniopharyngiomas compose three per cent of intracranial neoplasms (Graziani *et al.*, 1994). They usually develop in the anterior lobe of the pituitary gland, which is derived from Rathke's pouch (Barnes and Peel, 1985). Therefore, it is generally believed that craniopharyngiomas arise from the squamous epithelial cells of remnants of the craniopharyngeal duct, which also comes from Rathke's pouch (Demaerel *et al.*, 1993). Craniopharyngiomas are usually localized in the suprasellar area (Graziani *et al.*, 1994). About one-third of them involve the sellar region (Maria *et al.*, 1995). It is very rare for craniopharyngiomas to arise from the infrasellar region (Akimura *et al.*, 1989). When craniopharyngiomas are localized in the infrasellar region, they are frequently nasopharyngeal or come from the sphenoid bone (Barnes and Peel, 1985; Graziani *et al.*, 1994). Both are also considered to originate from the remnants of Rathke's pouch (Akimura *et al.*, 1989; Graziani *et al.*, 1994).

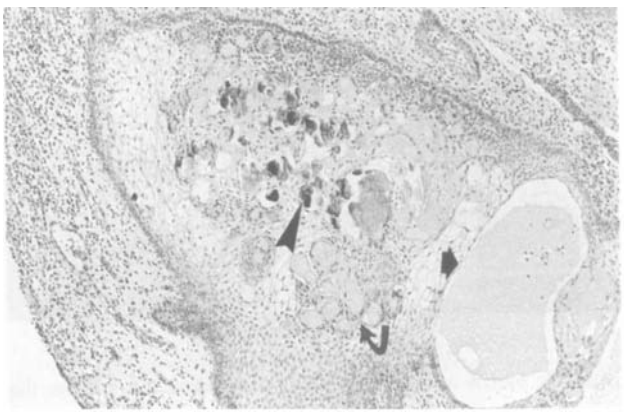

FIG. 4

The tumour specimen showing cystic degeneration (arrow), focal calcification (arrowhead), foci of squamous metaplasia with solid nests of keratinization (curved arrow), and a mixed inflammatory reaction (H & E; × 200).

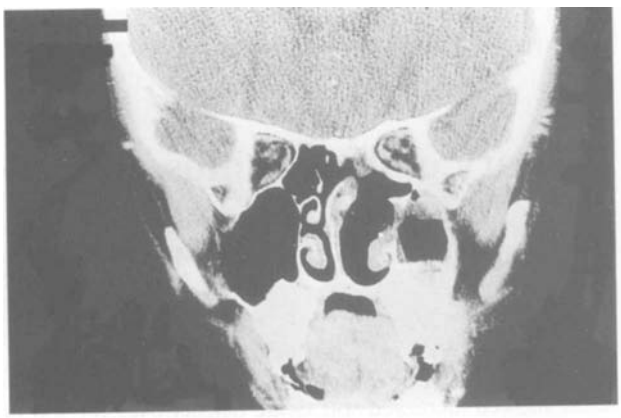

FIG. 5

A post-operative CT scan shows that the ethmoid sinus mass has been removed.

Paranasal sinus craniopharyngiomas, as mentioned above, usually involve the sphenoid sinus (Akimura *et al.*, 1989). On the other hand, the ethmoid sinus was only occasionally secondarily involved (Demaerel *et al.*, 1993). A primary ethmoid sinus craniopharyngioma has not been reported in the literature. The craniopharyngioma reported here was located only in the left ethmoid cavity. No other sinus or neighbouring structure was involved. Because of its location, it may be paradoxical to explain the origin of primary ethmoid sinus craniopharyngioma by Erdheim's theory in which craniopharyngiomas are thought to arise from remnants of Rathke's pouch (Graziani *et al.*, 1994). An ectopic explanation has been suggested to reconcile this paradox. Other ectopic sites except ethmoid sinus have been reported, such as third ventricle craniopharyngiomas (Graziani *et al.*, 1994). Apart from the probable origin from ectopic squamous remnants, other proposals may also explain the origin of this unusually located craniopharyngioma (Barnes and Peel, 1985). These include originating from misplaced tissue of embryonic enamel organ, an epidermoid metaplastic process or epitheliomas.

Nasal obstruction and impaired visual ability are the main symptoms of infrasellar craniopharyngiomas (Akimura *et al.*, 1989; Demaerel *et al.*, 1993; Graziani *et al.*, 1994). This can be expected from the location and extension of the tumour mass (Barnes and Peel, 1985). Conversely, epistaxis is not a usual symptom of craniopharyngiomas (Barnes and Peel, 1985). In this case, epistaxis was the only symptom. Although this symptom may be explained by the tumour location as well, it made the diagnosis more difficult.

The imaging characteristics of craniopharyngiomas have been widely reported (Akimura *et al.*, 1989). The presence of calcification is a diagnostic feature (Pusey *et al.*, 1987). On CT images, a mixed pattern which consists of high-density calcified areas, isodense non-cystic solid components, and low-density cysts is usually present (Akimura *et al.*, 1989). A similar picture was seen in this case. However, due to its rarity, the differential diagnosis includes inverted papilloma, nasal polyposis, and fibrous dysplasia.

Since craniopharyngiomas are generally radioresistant, surgical removal is the main treatment modality (Barnes and Peel, 1985). It has been said that craniopharyngiomas have a tendency to recur (Maria *et al.*, 1995). Therefore, complete removal of the tumour mass is the surgical aim (Maria *et al.*, 1995). A variety of operative procedures have been used, depending on the location and extension of the tumour mass, and the consideration of operative complica-

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tions and safety (Graziani *et al.*, 1994; Sethi and Pillay, 1994; Maria *et al.*, 1995).

Endoscopic sinus surgery has become popular recently (Sethi and Pillay, 1994). It has been applied for a variety of indications. It has been demonstrated to be able to treat inverted papilloma effectively (Waitz and Wigand, 1992). By the same token, it is assumed that endoscopic sinus surgery is a good way to treat primary ethmoid sinus craniopharyngioma. Under the endoscope, visualization is improved (Sethi and Pillay, 1994). Therefore, it is easier to remove the tumour mass completely and safely. Furthermore, since most craniopharyngiomas occur under the age of 25, as in our case, endoscopic sinus surgery is a minimally invasive surgical technique which is suitable for that condition (Barnes and Peel, 1985; Sethi and Pillay, 1994).

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