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Infrasellar craniopharyngioma presenting as epistaxis, excised via Denker's medial maxillectomy approach

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Abstract
Infrasellar craniopharyngioma is an uncommon neoplasm. Rarely, it presents with epistaxis. To date, 39 cases of infrasellar craniopharyngioma have been reported in the world literature. Involvement of the maxillary sinus has only been reported once. We report a second case exclusively involving the nasal cavity, ethmoid sinus and maxillary sinus. Excision was performed using a Denker's medial maxillectomy approach.

Key words: Infrasellar Craniopharyngioma; Denker's Medial Maxillectomy

Introduction

Craniopharyngioma is a rare, benign epithelial neoplasm.[1] These tumours arise in the area of the sella turcica. They can occupy both intra- and suprasellar regions, with the suprasellar location being the most common.[2] Occurrence of craniopharyngioma along the tract of the obliterated craniopharyngeal duct (which includes the sphenoid bone, vomer and the nasopharynx) is extremely rare.[3] Infrasellar craniopharyngioma was first reported in 1924.[4] To our knowledge, only one case with extension into the maxillary sinus has been reported in the literature. This was managed surgically via a lateral rhinotomy approach.[5] Our report discusses this rare condition and we describe a different, aesthetically superior surgical approach to managing such tumours.

Case report

A 59-year-old man with known muscular dystrophy presented to the accident and emergency department with a 3-day history of intermittent, left-sided epistaxis. He was initially managed with left-sided nasal packing and then referred to the otolaryngology department for further management. The pack was removed after 24 hr and, as per routine, rigid nasendoscopy was performed. This revealed a lesion in the left nasal cavity that was vascular and polypoidal in appearance. An examination under anaesthesia of the nose and biopsy of the lesion was performed. The lesion appeared to be arising from the left middle meatus and extending into the postnasal space. There was profuse bleeding on biopsy of the lesion. A computed tomography (CT) scan was performed to delineate the extent of the lesion. The CT images revealed the presence of a large soft tissue mass measuring approximately 4.4 × 4.1 cm centred in the left maxillary sinus, extending through the medial wall of the maxillary sinus, filling the nasal cavity and abutting the nasal septum. Posteriorly, it occupied the nasopharynx; anteriorly, it was bounded by the anterior wall of the maxillary sinus, with erosion. Superiorly, it extended into the sphenoid and ethmoid sinuses. There was no defect in the skull base or the orbit. (Figures 1, 2 and 3).

Surgical excision of the craniopharyngioma was undertaken, with the lesion being macroscopically completely excised via a left Denker's medial maxillectomy and spheno-ethmoidectomy approach.[6] At the time of surgery, the tumour was noted to have eroded through the anterior maxillary wall. Bulky tumour was seen in the antrum and was excised piecemeal. The nasomaxillary buttress was reduced to open into the nasal cavity. Mucosa of the inferior meatus was incised along the length of the inferior turbinate, and the inferior turbinate excised. Tumour in the middle meatus, arising from the middle turbinate, was excised. At this point in the procedure, bleeding is always encountered from the spheno-palatine artery, which is easily controlled with suction diathermy. The ethmoid sinuses were opened and cleared up to the anterior skull base. The sphenoid sinus was opened and contained only mucopus. In keeping with the CT findings, there was no breach of cribiform plate, lamina paperacia or orbital floor. The tumour therefore appeared to have arisen entirely intranasally.

A Bismuth Iodoform Paraffin Paste pack was inserted and this was removed 1 week post-operatively under general anaesthetic. Post-operative management included regular saline douches to keep the cavity clean and crust-free. Clinical follow up involving endoscopic examination of the naso-ethmoid-maxillary cavity revealed no sign of recurrent disease, and at the time of writing the patient remained well and under review, 12 months post-surgery.

Histology

Examination showed almost identical histological appearances in the initial biopsy and in the piecemeal resection specimen. Normal mucosa or respiratory epithelium was not identified in the initial biopsy, but

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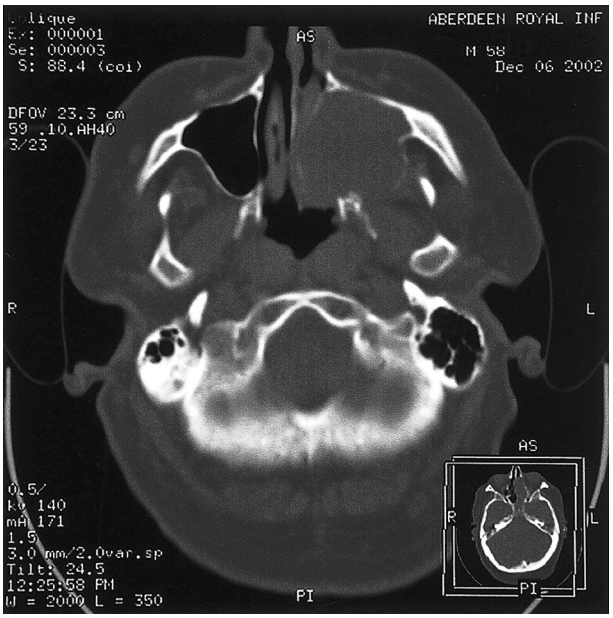

FIG. 1

Axial CT image showing mass in the maxillary sinus, and bulging of medial and lateral walls of the maxillary sinus.

focally, within the resection specimen, tumour was evident growing out of (or invading under) the nasal mucosa (Figure 4).

The lesion consisted of vascular, myxoid or oedematous fibroconnective tissue stroma containing innumerable closely packed islands, anastomosing cords and strands of epithelial cells. Many of the islands of epithelial cells had a characteristic appearance, with a well-defined basal layer and a more loosely arranged central network of stellate cells (Figure 5). Foci of cystic change were also visible. The anastomosing strands or cords had a more papillary appearance in places and comprised mostly basaloid cells. Keratin pearls and keratinization were not significant

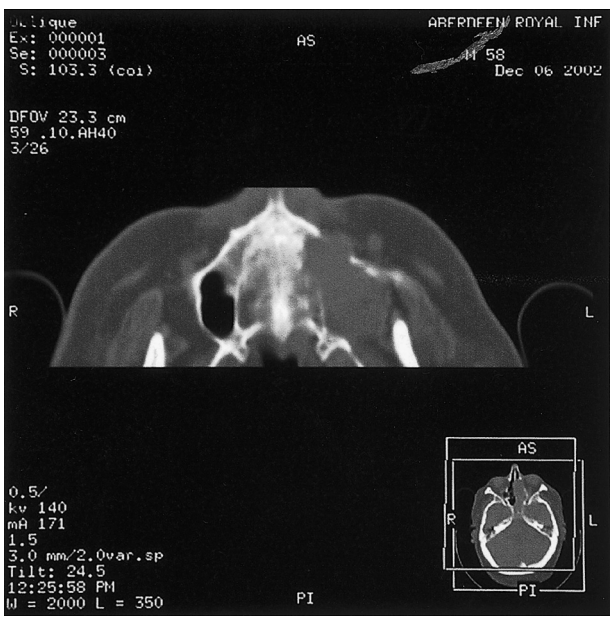

FIG. 2

Axial CT image showing soft tissue mass, with bony destruction of anterior maxillary sinus wall.

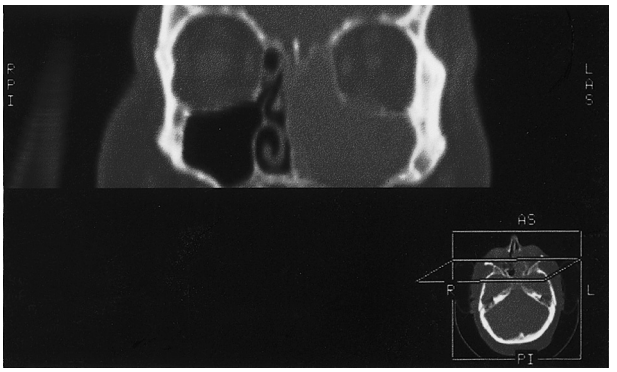

FIG. 3

Coronal CT image showing opacification of maxillary and ethmoid sinuses, with intact skull base and orbit.

features. Mitotic activity was visible in the epithelial component but this was not excessive, and there was no evidence of cytological atypia or malignant transformation. There was no histological evidence of bony invasion or involvement. The histological features were consistent with those of a craniopharyngioma.

Discussion

Craniopharyngiomas are rare, constituting approximately 3 per cent of all intra-cranial tumours[1] with a similar frequency in children and adults and a slight preponderance in the first two decades of life. In 90 per cent of cases, craniopharyngiomas are located extra-axially in the sellar or suprasellar area.[7] They can extend to the anterior (2–5 per cent of cases), middle (2 per cent) or posterior (1–4 per cent) cranial fossae, and infrasellar extension is found in about 5 per cent of cases.[8]

Although benign, they are locally aggressive epithelial neoplasms. Theories as to the origin of this tumour suggest that they arise from the tract of the obliterated craniopharyngeal duct (Rathke's pouch) or perhaps from the functioning pharyngeal hypophysis.[3,9,10] An infrasellar location of the tumour along the posterior

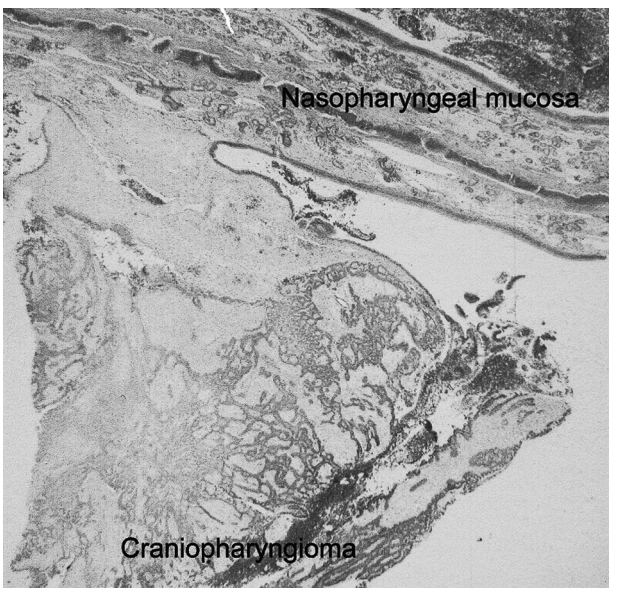

FIG. 4

Craniopharyngioma is seen projecting out from under the nasopharyngeal mucosa (H&E × 20).

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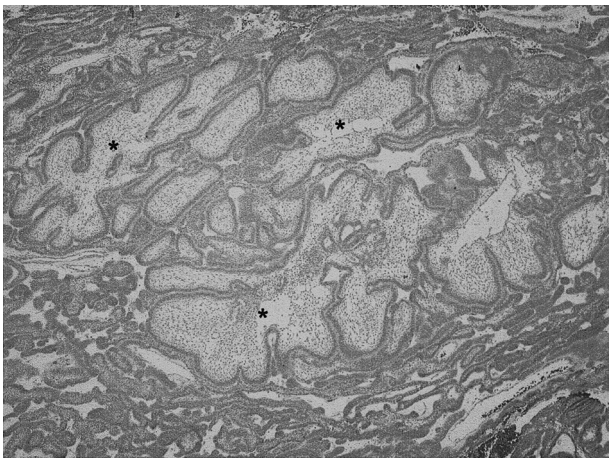

FIG. 5

Typical appearance of craniopharyngioma, comprising vascular stroma with epithelial lobules in the centre, and anastamosing cords and strands of more basiloid epithelial cells around the periphery. The epithelial lobules show peripheral pallisading, with a central loose network of cells (*) (H&E × 40).

aspect of the vomer above the junction of the soft palate and nasal septum, the sphenoid bone and the undersurface of the floor of sella could be accounted for by these theories.

Symptoms and clinical findings are consistent with the tumour's location and its mass effect, with compression of surrounding structures. In suprasellar tumours, this is often characterized by visual field defects, pituitary insufficiency and symptoms of raised intracranial pressure.[7] Tumour located in the sphenoid sinus presents with headache and cavernous sinus syndrome.[11–13] Tumours in the nasopharyngeal region usually present with frontal headache, nasal obstruction, epistaxis, and nasopharyngeal and nasal fossa masses.[4,5,8,11,14] Our case presented as an emergency with epistaxis.

Computed tomography is the investigation of choice as it reveals the heterogeneous nature of the tumour, with its solid and cystic components. It also defines the extent of the soft tissue mass and any bony destruction.[15] Other diagnostic investigations include analysis of cerebrospinal fluid for protein content, which is normal if tumours are confined to the sella turcica but raised if extrasellar extension is present.[16] The anterior pituitary function is evaluated to assess the extent of gland involvement.

The treatment of choice for craniopharyngiomas is surgical excision with the aim of completely removing all tumour. One approach described for surgical excision of both infra- or intrasellar craniopharyngiomas is transpalatal (Loeb, 1927),[17] while the lateral rhinotomy approach (described in 1845 by Ferguson) has been used for excision of tumours occupying the nasopharynx, the nose and paranasal sinuses.[18] The frontopterional approach is also commonly used.[19]

In our case we adopted the sublabial Denker's approach described in 1906 for medial maxillectomy and spheno-ethmoidectomy.[6] Using this technique, via an extended anterior antrostomy, complete visualization and excision of the tumour was possible. This approach enables direct vision whilst working near vital structures, and thereby potentially reduces the risk of operative complications. Bleeding from the sphenopalatine artery is generally encountered but easily controlled by direct suction diathermy. We would recommend this approach not only for excellent visualization of infrasellar craniopharyngioma at the time of excision but also because the incision is sublabial and hence aesthetically superior.

When complete removal of tumour is not possible surgically, post-operative radiotherapy is proposed as this has been shown to increase survival rates, with 10-year survival rates of up to 76 per cent.[20]

Conclusion

Craniopharyngiomas are rare, benign but locally aggressive tumours. Complete excision is the treatment of choice. We describe only the second case in the literature involving the maxillary sinus. We advocate the Denker's approach for excision of an infrasellar craniopharyngioma as it provides excellent visualization of the tumour at the time of excision, and improved aesthetic results.

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