Rare disease

Skull base oncocytoma presenting as epistaxis: an unusual presentation of a rare tumour successfully managed with active surveillance

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Summary

Oncocytomas are rare tumours, usually occurring in the salivary glands, but may very occasionally occur in other sites. The authors present a skull base oncocytoma as a rare cause of spontaneous epistaxis. Following diagnosis through imaging and intranasal biopsy, the patient opted for annual surveillance instead of active treatment and made a full recovery nonetheless. Skull base oncocytoma is a rare tumour that may result in otherwise common symptomatology. While excision is the mainstay of management, active surveillance may be a viable alternative for select patients.

BACKGROUND

Epistaxis is one of the commonest otolaryngological emergencies. The majority of cases are idiopathic in nature, with very few being caused by neoplasm.¹ Inverted papilloma and juvenile angiofibromas are the most commonly cited benign tumours to cause epistaxis. We describe the case of an oncocytoma arising from the skull base which presented with epistaxis. These tumours, already rare in their own right, usually arise asymptotically in the major salivary glands, but have also been reported as occurring in other various head and neck locations, including the nose and paranasal sinuses.² What makes it exceptional is its rare location, and oncocytoma of the skull base primarily presenting as spontaneous epistaxis has not been previously described in the literature. Finally, our case is unique in that the patient was managed through active surveillance.

CASE PRESENTATION

A 78-year-old Caucasian male presented with a history of spontaneous, profuse bilateral epistaxis, which was controlled with nasal packing. His nosebleeds, previously only left-sided, had been a persistent problem for the past 4 years, and this was his fifth presentation to the hospital. He gave no previous history of nasal surgery or trauma and was not taking anticoagulant or antiplatelet medications. His medical history included hairy-cell leukaemia which was in remission postchemotherapy, hypertension and rheumatoid arthritis.

The packs were removed after 48 h and the nose was examined. Anterior rhinoscopy did not reveal an anterior bleeding point and rigid nasendoscopy showed only a left-sided generalised bloody ooze. No other abnormalities were noted, though views on the left were limited due to a septal deviation. Due to this continuing ooze and a significant history of unresolving, recurrent, unilateral epistaxis

the decision was taken to perform a left sphenopalatine artery ligation.

Following a septoplasty to allow further access, an examination under general anaesthetic revealed a polypoid mass arising from the left skull base, medial to the superior turbinate and involving an intersinus septal cell. This was deemed to be the cause of his persistent bleeding, and the ligation of his sphenopalatine artery was thus abandoned. There was no evidence of skull base dehiscence or a cerebrospinal fluid leak. Multiple biopsies were taken for histological examination.

INVESTIGATIONS

CT of the sinuses was requested postoperatively which showed opacification mainly of the left ethmoid and frontal sinuses and of a pneumatised crista galli. The lamina papyracea was seen to be intact (figures 1 and 2). MRI with contrast confirmed a relatively localised lesion involving the left superior meatus which abutted the cribriform plates and extended in continuity into, and slightly expanded the crista galli, but with no evidence of meningeal involvement or reaction in the underlying brain. A paraganglioma or an ectopic pituitary gland were both entertained as differential diagnoses, but an MRI without contrast revealed a normal pituitary gland and fossa.

Histology showed squamous mucosa with focal reactive changes and a proliferation of oncocytic cells with uniform round nuclei, prominent nucleoli and abundant granular eosinophilic cytoplasm within its stroma. The cells were found to be arranged in anastomosing trabeculae, separated by fibrovascular septa and, in places, interspersed between nests and strands of squamous epithelial cells (figure 3). Immunohistochemistry showed the oncocytic cells to be strongly positive for cytokeratin and chromogranin A. There was also focal positivity for markers NSE and S-100. Melan A was also weakly

Figure 1 Coronal CT scan of sinuses showing oncocyoma arising from left skull base.

positive, but was of unknown significance; CD 56, CD 10, adrenocorticotrophic hormone and HMB 45 were all negative. Ki67 was focally positive in the overlying epithelium, in keeping with reactive changes, but was negative within the oncocyctic cells. The morphological and immunohistochemical appearances were of a benign oncocyoma.

DIFFERENTIAL DIAGNOSIS

A paraganglioma or an ectopic pituitary gland were both entertained as differential diagnoses, but an MRI without contrast revealed a normal pituitary gland and fossa.

TREATMENT

After discussion at the regional head and neck multi-disciplinary team meeting, the management options presented to the patient were to either perform endoscopic resection or to conservatively follow the mass up annually. The patient opted for the latter.

OUTCOME AND FOLLOW-UP

At his most recent clinic appointment, he described no further epistaxis and rigid nasendoscopy showed only scarring between the septum and superior turbinate. A follow-up CT scan has confirmed resolution.

DISCUSSION

Oncocytomas are rare epithelial tumours, occurring most commonly in middle aged or older women. They are most frequently found in the parotid gland, but also rarely occur in endocrine system (thyroid, parathyroid and adrenal glands), buccal mucosa, breast, kidney, upper aerodigestive tract, ovary and the nasolacrimal system.^{3 4} Their occurrence within the nasal cavity is rare and they may exhibit either benign^{2 3 5 6} or malignant^{7 8} characteristics. Diagnostic criteria have been developed for malignant oncocytomas which include the invasion into and destruction of adjacent structures; perineural, intravascular or lymphatic invasion; local lymph node involvement of distant metastases.² Johns *et al* through ultrastructural study, consider all oncocytomas of the nose, paranasal sinuses and larynx to be grade 1 carcinomas.⁹

When occurring within the nasal cavity, locations described include the nasal septum,^{3 5} the postnasal space,⁶ the turbinates^{2 8} and the paranasal sinuses.¹⁰ They have also been described as arising from the skull base in rare cases.¹¹ Patients usually present with symptoms of nasal obstruction and/or epistaxis, although involvement of neighbouring structures may occur such as orbital invasion from an ethmoidal oncocytoma.¹² Presentation may also be in the form of distal metastasis in oncocytomas that have undergone malignant transformation.⁸ Our case is

Figure 2 Sagittal CT scan showing oncocytoma arising from left skull base.

Figure 3 Histology slide showing oncocytic tumour cells forming sheets beneath the ciliated respiratory-type epithelium (H&E x50).

the first to describe a benign oncocyoma arising primarily from the skull base primarily presenting with spontaneous epistaxis. Had it been malignant in nature, the patient would have likely been at risk of intracerebral spread and its associated complications.

Hampton *et al* describe an array of rare tumours arising from the sellar region that can be easily confused with pituitary adenomas. Although clinically and radiographically the tumours resemble non-functioning pituitary adenomas, their histologic, immunohistochemical and ultrastructural features differ and indicate a salivary gland origin, and include cellular pleomorphic adenoma, monomorphic adenoma, oncocyoma and low-grade adenocarcinoma of the salivary gland.¹¹ Indeed, the initial differential diagnoses of our case included an ectopic pituitary adenoma or a paraganglioma. While the former was ruled out radiologically, the latter was considered unlikely due to the absence of true S-100 positive sustentacular cells at histology. Cytokeratin and chromogranin A positivity would have favoured an ectopic pituitary adenoma, however pituitary function tests were within normal limits. All other histological and immunohistochemical findings were characteristic of a benign oncocyoma. However, Comin *et al* state that the biological behaviour of oncocyomas of the nasal cavity seems related to their local growth rather than to the cytological characteristics of the tumour.¹⁰

Management of oncocyomas differs depending on the nature, ranging from simple excision in benign cases, to adjuvant therapies such as intensity-modulated radiotherapy in malignant cases.⁷ From a surgical perspective, excision can be performed either intranasally or, where necessary, through craniofacial approaches such as a Le Fort I osteotomy and CO₂ laser excision as described by Colveary *et al*⁶ depending on the anatomical location. We biopsied our patient's lesion endoscopically, and made the decision to also excise it endoscopically with the use of a microdebrider instrument. However, based on the patient's wishes, annual surveillance was undertaken instead. Nonetheless, with subsequent visits, the lesion was seen to self-involute, leaving behind only scarring, presumably as a delayed result of the initial biopsies.

Learning points

- ▶ Oncocyomas are rare tumours and their occurrence in the nose is exceptional, where they present as obstruction or epistaxis and may behave in a benign or a malignant fashion.
- ▶ Nasal oncocyomas usually arise from the septum, but may arise in other areas of the nose and paranasal sinuses, including the skull base.
- ▶ Ideally, treatment is excision and, where malignancy is present, radiotherapy. However, in selected cases, active surveillance can be undertaken.

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Competing interests None.

Patient consent Obtained.

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