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Endoscopic sinus surgery for sinonasal haemangiopericytomas

E. Serrano, A. Coste*, J. Percodani, S. Hervé*, L. Brugel*

Abstract

Haemangiopericytomas are rare peri-vascular tumours with variable malignant potential. The nasal cavity and the paranasal sinuses are most often involved in the head and neck. Five cases of haemangiopericytomas treated by a strict endonasal endoscopic approach are presented. Bleeding and nasal obstruction are the most frequent symptoms. Computed tomography (CT) scan and magnetic resonance imaging (MRI) allowed pre-operative assessment. Angiography with embolization was needed in two of the five cases. None of our patients presented with malignant histology. Our five cases were operated on, and a total tumour excision was performed through the endoscopic endonasal approach. We had one recurrence with a mean follow-up of 4.5 years. We suggest that when the tumour is purely intranasal or strictly located in the ethmoid or sphenoid sinus, it can be removed via an endonasal approach under endoscopic guidance in experienced hands.

Key words: Haemangiopericytoma; Paranasal Sinus Neoplasms; Surgical Procedures; Operative; Endoscopy

Introduction

Haemangiopericytomas (HPs) are uncommon mesenchymal tumours that represent approximately one per cent of vascular tumours.[1,2] They were first described by Stout and Murray in 1942.[3] They are thought to be derived from extra-capillary cells, the pericytes, which, in these tumours, surround normal vascular channels. They are found throughout the body[4] with less than one third occurring in the head and neck.[1,5,6] Of these, only five per cent are located in the nasal cavity and paranasal sinuses,[2,7] with a predilection for the nasal cavity, and the ethmoid and sphenoid sinuses.[8] Their aetiology remains unknown. A past history of trauma or long-term corticosteroid treatment have been suggested without confirmation.[8] These tumours occur in all age groups with no sex predilection, and a peak incidence between the 5th and 6th decade.[9] HPs are tumours of uncertain evolution, generally slow-growing, with local infiltration.[6,10] They slowly invade the surrounding tissues. Generally they are considered to have malignant potential[10] with late recurrences reported in almost half of patients and metastasis in up to 10 per cent. Moreover, mortality can be as high as 50 per cent, within five to 20 years after the first appearance.[11] Wide local excision and long-term follow-up whatever the histology are therefore mandatory. When the tumour is purely intranasal and especially if attached to the nasal septum, or when the tumour is strictly located in the ethmoid or sphenoid sinus, it can be removed via an endonasal approach under endoscopic guidance.[12]

We report five cases of non-malignant HPs (two located in the nasal cavity and three in the ethmoid sinus) treated by a strict endonasal endoscopic approach which allowed us to perform a complete and secure resection, thus avoiding an external scar and the secondary complications of lateral rhinotomy.

Case report

Our five cases are summarized in Table I.

A pre-operative biopsy was performed in three cases. A CT scan was performed in all cases, confirming the vascular nature of the tumour and showing that it was strictly located in the nasal cavity, or the ethmoid and/or sphenoid sinuses, thus according a removal via an endonasal approach. The MRI, available in two cases, confirmed the CT scan findings. Carotid angiography with a selective embolization was performed in two cases.

All patients were operated on by endonasal endoscopic surgery with a complete resection. No post-operative complication was noted. The pathological reports were in all cases in favour of a benign HP.

One of our patients presented at five years follow-up with a local recurrence that could still be treated by further endoscopic endonasal resection.

Discussion

Most often, HPs arise in the musculoskeletal system, the skin of the limbs, trunk and from the retroperitoneal area. In the head and neck, they usually develop in the soft tissues of scalp, face or neck.[6,10] The tumours are twice as common in the nasal cavity as in the paranasal sinuses. The sphenoid and ethmoid sinuses are involved four times more often than the maxillary sinuses.[2]

Reports of case series point out equal sex distribution. No ethnic prevalence has been demonstrated. HPs have been reported at any age, from birth to old age, but there appears to be a slight incidence peak occurring in the 6th and 7th decades.[13] In our series, the patients were all more than 60 years old with one exception (51 years).

From the Department of Otorhinolaryngology Head and Neck Surgery, Rangueil Hospital, Toulouse, and the District and Henri Mondor Hospitals*, Créteil, France.
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TABLE I
PRESENTATION OF OUR SERIES

	Symptoms	Duration before diagnosis	Endonasal location	CT scan	MRI	Arteriography	Embolization	Follow up	Recurrence
Case 1 Female 72 years	Epistaxis Nasal obstruction	?	Pedicle to the nasal septum between middle turbinate and nasal septum	Left nasal cavity, nasopharynx and ethmoid cells	–	Marked tumour blush with early draining veins	Maxillary artery	4 years	–
Case 2 Male 65 years	Epistaxis	11 years	Pedicle to the nasal septum below the cribiform plate	Left nasal cavity, nasopharynx and ethmoid cells	–	Important and bilateral vascularization from the left anterior and posterior ethmoidal arteries, the right posterior ethmoidal arteries and both spheno-palatine arteries	Posterior ethmoidal arteries Sphenopalatine arteries	1 year	–
Case 3 Female 65 years	Epistaxis	4 months	Superior meatus, extending to the floor of the nasal cavity	Left posterior ethmoidal cells and spheno-ethmoidal recess	+	–	–	8 years	Recurrence at 5 years in the posterior ethmoid cells operated on endonasal approach
Case 4 Male 66 years	Epistaxis Nasal obstruction	?	Superior meatus	Left posterior ethmoidal cells	+	–	–	4 years (died of intercurrent event)	–
Case 5 Female 51 years	Nasal obstruction	1 year	Superior meatus, filling the nasal cavity, implanted on the upper area of the medial face of the middle turbinate and on the olfactory recess	Right posterior ethmoidal cells and sphenoid sinus	–	–	–	6 years	–

The aetiology of HPs remains unknown. Previous trauma has raised the possibility that this may stimulate proliferation of pericytes following damage to the capillary network and long-term steroid therapy and arterial hypertension have been suggested, although none of these theories has been proved.[14] None of these possibilities occurred in our cases.

Clinical aspects

No specific symptomatology exists for sinonasal HPs. Bleeding (spontaneous or provoked) and nasal obstruction are the most frequent symptoms. Pain is unusual but can occur when lesions are large enough to invade surrounding tissues or when they are confined in unyielding spaces such as the paranasal sinuses.[8] Their natural history is uncertain but they are generally slow-growing, solitary and indolent with local infiltration.[15,16] In our series, isolated nasal bleeding was the first symptom in two cases, associated with nasal obstruction in two cases. One patient presented only with isolated nasal obstruction.

Macroscopically, these tumours are described as soft and tan-coloured, bleeding easily and as a consequence biopsies should be carefully performed, because of the risk of haemorrhage.

Pre-operative check-up

Pre-operative examination must include fine detail axial and coronal CT scans of the nose and paranasal sinuses with contrast to show a soft-tissue mass which enhances on post-contrast scan.[17,18] (Figure 1).

Usually, MRI demonstrates a mass with an isotense signal on T1-weighted images, an iso- or low signal intensity on T2-weighted and enhancement when gadolinium is injected.[17,18] (Figure 2). Moreover MRI helps to differentiate the tumours from mucus.

Conventional angiography confirms the vascular nature of the tumour and enables pre-operative embolization but is of less importance in small tumours. This can obviously facilitate surgical resection of tumour, by diminishing the peri-operative bleeding.[19,20] Pre-operative angiography

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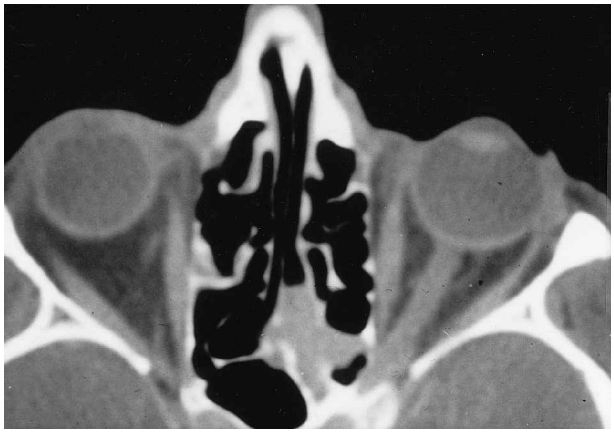

FIG. 1
CT scan: soft-tissue mass filling the left posterior ethmoidal cells.

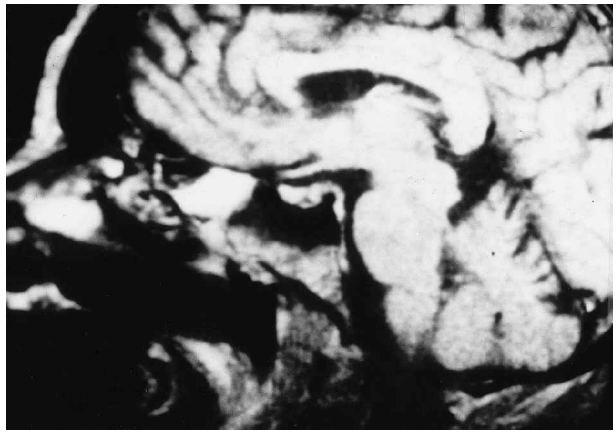

FIG. 2
MRI: Enhancement of the tumour after injection of gadolinium on T2-weighted sequence.

was performed in two of our patients with a tumour extending to the nasopharynx. A CT scan was performed in all five patients and MR imaging in two cases.

Histological diagnosis

Histological examination needs to be carried out on the whole surgical specimen, as a simple biopsy is frequently insufficient.

Macroscopically, the tumour appears as a red, polypoid mass. HPs are frequently encapsulated, although this is often a discontinuous pseudo-capsule, composed of a large accumulation of peritumoral connective tissue.[21]

Microscopically, HPs are typically constituted from sheets of relatively uniform ovoid or spindle-shaped cells with an indistinct cytoplasm and large nuclei, distributed around medium-sized and small vascular channels. These vascular channels often exhibit an abnormal branching 'staghorn' pattern. The tumour cells are invested in a fine network of pericellular reticulin and usually show immunohistochemical positivity for vimentin, with a proportion also expressing CD34, smooth muscle actin and desmin.[22] Histochemical and immunohistochemical techniques are essential, mainly to exclude other morphologically similar soft tissue tumours, including some patterns of malignant fibrous histiocytoma, synovial sarcoma, mesenchymal chondrosarcoma, haemangiomas and glomus tumours.[23]

The pathological criteria for HPs malignancy are: tumour size, increased cellularity, cellular pleomorphism, increased mitotic activity, necrosis and haemorrhage.[22] Nevertheless, none of these histological criteria have been shown to be predictive of the behaviour of HPs.[24]

None of the tumours in our patients fulfilled the histological malignant criteria above.

Cytometric flow analysis of DNA[23] have shown that diploid DNA content correlates with a long S phase, and therefore, slow proliferation and a good prognosis; on the other hand, aploid DNA content corresponds to a high percentage of cells in the S phase, indicating increased proliferative activity, and a poor prognosis.

Treatment

Due to its latent potential for malignancy, wide local excision, is still considered the gold-standard treatment.[13] In paranasal sinus localizations, an external procedure is most frequently chosen to achieve total tumour excision, but when the tumour is purely intranasal or strictly located in the ethmoid or sphenoid sinus, it can be removed via an endonasal approach.

Conclusion

HPs are uncommon vascular tumours, rarely located in the nasal cavity and paranasal sinuses. They appear as indolent reddish-grey masses, bleeding easily and profusely when traumatized. CT scan and MRI allowed pre-operative assessment. Angiography with embolization was not required in all cases. Tumour excision must be complete due to the latent potential for malignant behaviour of these tumours. This may be achieved by an endonasal endoscopic approach. If this is not possible, an external approach is needed which might be combined with endonasal endoscopic control.

Our five patients received no complementary treatment, as the excision was macroscopically complete and no histological criteria for malignancy were found. Only one recurrence occurred with a mean follow-up of 4.5 years. However, life-long follow-up is recommended.

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Address for correspondence:
Professor Elie Serrano,
Department of Otorhinolaryngology – Head and Neck Surgery,
Rangueil Hospital,
31403 Toulouse Cedex 4,
France.

Fax: +33(0) 561322078
E-mail: serrano.e@chu-toulouse.fr

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