Introduction

Haemangiopericytoma (HPC) accounts for less than 1% of all vascular tumours.\(^1\)\(^2\) It originates from Zimmerman’s cells, the pericytes around capillaries.\(^1\) Although the most common sites of HPCs are the pelvis, retroperitoneum and lower extremities,\(^1\) head and neck involvement occurs in 15-25% of cases.\(^3\) Only 5% of HPCs originate from the nasal cavity and sinuses\(^3\)\(^-\)\(^5\) and the most common signs of presentation are nasal obstruction, epistaxis, and polypoid nasal mass.\(^6\)\(^-\)\(^8\) Pain, visual disturbance, and headache are infrequent symptoms.\(^1\) In 1942, Stout and Murray published the first report on HPC. However, in 1976 Compago and Hyams described a specific type of it as sinonasal haemangiopericytoma-like tumour (SHPCL).\(^3\) HPCs are commonly thought to behave in variable biological ways.\(^1\) Although sinonasal haemangiopericytoma is generally considered to be a less aggressive tumour with a more favourable prognosis,\(^3\)\(^4\)\(^8\) there are some reports of destructive forms of HPCs located in the nasal cavity.\(^2\)\(^3\) Since this tumour is relatively radioresistant and has a major tendency to bleed, wide surgical resection in an open approach and with pre-operative embolisation is the main management option for large sinonasal haemangiopericytoma. We report on a case of haemangiopericytoma with massive unilateral involvement of the maxillary, anterior and posterior ethmoids, sphenoid, and frontal sinuses with extension to the ipsilateral orbit causing proptosis and diplopia. The tumour was completely removed endoscopically, and the dura exposed in the posterior part of the frontal recess was covered with a septal mucosal graft.

Case report

A 34-year-old man was referred to our hospital with an 8-month history of proptosis, intermittent epiphora and pain in the right eye and ear. He did not complain of any nasal obstruction or epistaxis. Physical examination of the right eye revealed a limitation of movement to medial, causing diplopia without any change in visual acuity. A nasal examination with endoscope showed that the lateral wall of the right maxillary sinus, the superior part of the septum, and the anterior wall of the right sphenoid sinus were destroyed and the right maxillary, sphenoid, and frontal sinuses were partially involved by the tumour.
There was some erosion in the cribiform plate of the ethmoid bone and fovea ethmoidalis, but no intracranial invasion was detected. Other paraclinical studies revealed no local or distant metastasis. The stage of the disease according to the scale of the American Joint Committee on Cancer (AJCC) was estimated at T4a, N0, and M0. An endoscopic incisional biopsy of the mass was performed under general anaesthesia and profuse bleeding occurred during the procedure. A histological study and the immunohistochemical staining of the tumour led to a diagnosis of haemangiopericytoma-like sinonasal-type tumour or glomangiopericytoma. We planned the total endoscopic removal of the tumour. At first, a hemitransfixion incision was made in the posterior part of septum behind the keystone area; the submucoperiosteal plane on the contralateral side was then dissected to remove the septal mucosa on the tumour side, the perpendicular plate of the ethmoid, and the vomer up to the articulation with the sphenoid rostrum. The mucosa on the contralateral side was removed separately and preserved in serum as a material for graft over the skull base. By removing the posterior part of the septum, two surgeons were able to work simultaneously in the operation field and successfully overcome profuse bleeding. Trasnasal sphenoidotomy was performed initially so that the skull base was found in the most posterior part and a mass measuring 4 cm × 5 cm × 6 cm was dissected from posterior to anterior. The right frontal sinus and frontal recess were the last parts to be treated. After the removal of agar nasi cells, the frontal recess was sufficiently wide and the mass which had just occupied the inferior part of frontal sinus and had not attached to any wall except the lateral wall of the frontal recess was pulled down and resected. Since the cribiform plate and fovea ethmoidalis were dehiscent just behind the frontal recess, the exposed dura was covered with a septal mucosal graft after the resection of the mass. The early post-operative period was uneventful and the patient was discharged after 48 hours. The control CT scan in the fourth week after the operation showed some enhancing residual tissues in the nasal cavity. In the second session of surgery, the remnant of the tumour, which was located submucosally over the inferior conchae and looked like a hypertrophied conchae, was excised. The patient was followed closely and remained tumour-free for about six months post-operatively (Figure 2).

**Discussion**

Haemangiopericytoma is an uncommon vascular tumour of unknown aetiology.\(^1\) Its incidence is equal among genders\(^2,3\) and more common between the third and fifth decades of life.\(^2\) HPC has been categorised as benign, borderline, and malignant according to the degree of cellular atypia, mitosis, haemorrhage or necrosis, and the size of the tumour. There is no reliable correlation between tissue characteristics and the clinical course of the disease.\(^3\) Sinonasal haemangiopericytoma-like tumour (SHPCL) can be classified into three subtypes.\(^10\) The first is similar to soft-tissue HPC in other parts of the body, with various degrees of cellular atypia, pleomorphism, necrosis, and no myoid differentiation.\(^5,10\) This histological subtype is usually aggressive and sometimes fatal. The second subtype, or true haemangiopericytoma, is composed of uniform cells with myoid differentiation, a little atypia and mitosis, but no necrosis. Some local recurrence may be seen in this type; nevertheless, prognosis is generally favourable. The third subtype of HPC is very similar to glomus tumour, with benign properties.\(^10\) The overall rate of
local recurrence in SHPCLs is 7% to 40%, and 4.2% of cases suffer from remote metastasis. Factors associated with worsening prognosis are large tumour volume (≥ 6.5 cm) and incomplete removal. Wide surgical resection is the main management modality, and chemotherapy and radiotherapy are used less commonly for the management of local and distant metastasis or unresectable primary tumours. The usual approach for the surgical removal of HPCs involving paranasal sinuses is lateral rhinotomy. Endoscopic resection has been recently introduced as an option which affords not only aesthetic results, but also a better view of the surgical field. Endoscopic visualisation of the tumour with straight and angulated lenses provides better exposure in all directions with adequate magnification, so complete excision of the mass may be possible. This technique also affords the opportunity for the endoscopic repair of the dura in the case of cerebrospinal fluid leakage, with an acceptable success rate and less morbidity than with open techniques. Although the piecemeal removal of a tumour is the usual trend in endoscopic surgery, it seems that it has no adverse effect on survival. According to the literature, the endoscopic approach is not sufficient when there is involvement of the skin, frontal sinus, orbital soft tissue, lachrymal system, hard palate, nasal pyramid, bony wall of maxillary sinus except medial wall, nasopharynx beyond the pharyngobasilar fascia, and massive invasion of the dura or lateral extension to the optic canal. Other studies suggest an endoscopic approach for HPC only when it is confined to the nasal cavity and the ethmoid or sphenoid sinus. This report appears to be unique because a tumour categorised as stage 4 with an extension to the frontal sinus was completely removed endoscopically without pre-operative embolisation. Since the positive effect of pre-operative embolisation in HPC has been not statistically proven, the patient did not undergo pre-operative embolisation and avoided the probable complications. This case was followed for a six-month period, which is too short to predict the outcome, but local or distant recurrence of HPC may occur any time during the life of the patient even after several years with no tumour. Regular endoscopic examination should be performed every three months during the patient's lifetime. A baseline CT with contrast one month after operation is essential and should be repeated annually for at least ten years and if endoscopic examination results in the suspicion of recurrence.

Conclusion

It appears that endoscopic techniques can be helpful in patients with massive haemorrhagic tumours of the paranasal sinuses, especially HPC. HPC is an intermediate tumour with an unpredictable prognosis, even after the complete removal of the tumour with open approaches. It therefore seems logical for the surgeons to abandon the more destructive external approaches in favour of endoscopic resection.

References


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