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Pathology in Focus

Haemangiopericytoma of infratemporal fossa

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Abstract

Haemangiopericytomas (HPCs) are rare vascular tumours that commonly involve the soft tissues of the trunk and lower extremities. In the head and neck, the most common sites are the nasal cavity and the paranasal sinuses, and unusually, the orbital region, the parotid gland, and the neck. We report a patient with HPC that originated in the infratemporal fossa and involved the pterygopalatine and the middle cranial fossae, apparently the first such case to be reported. Although the patient has undergone resection on three separate occasions, the tumour recurred. We then performed an extended resection using the infratemporal fossa approach type D. The patient has shown no recurrence in the past five years.

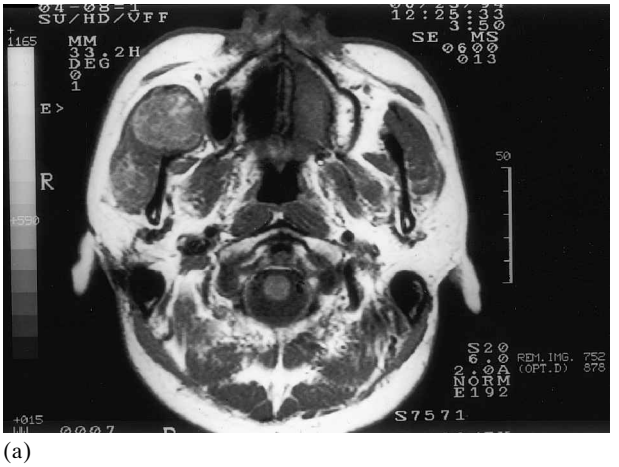
Although histopathologic confirmation of this malignancy may be difficult, extensive resection remains the most effective treatment in such cases.

Key words: Haemangiopericytoma; Skull Base, Neoplasms

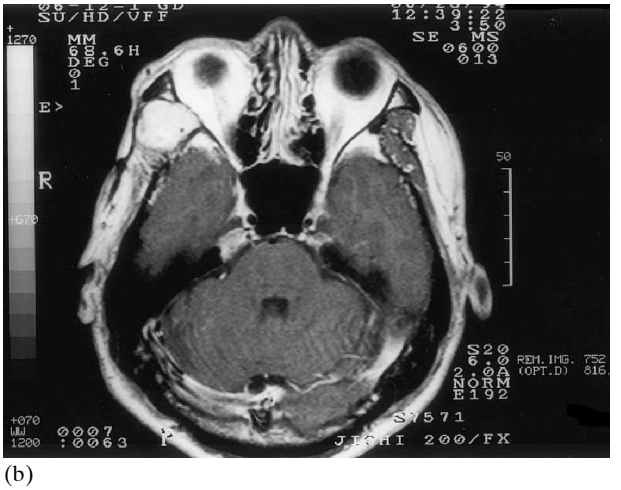
Introduction

Haemangiopericytoma (HPC) is an uncommon mesenchymal tumour. It comprises only one per cent of all vascular neoplasms[1] and approximately three per cent of all soft tissue sarcomas.[2,3] It is thought to demonstate differentiation toward the pericytes of Zimmermann, that lie externally to the endothelial cells of the capillaries and whose function involves changing the size of the vascular lumen.[4] The majority of HPCs occur in the trunk and lower extremities, with the head and neck being involved in only about 7.5 to 16 per cent of cases.[5,6]

This is a report of a rare case of recurrent HPC that originated in the infratemporal fossa and required surgery on the base of the skull.



(a)



(b)

FIG. 1

Magnetic resonance imaging (T1-weighted, gadolinium-enhanced). A gadolinium-enhanced mass was revealed in the right infratemporal fossa (a). There is destruction of the right pterygoid plates with extension through the right skull base into the middle cranial fossa (b).

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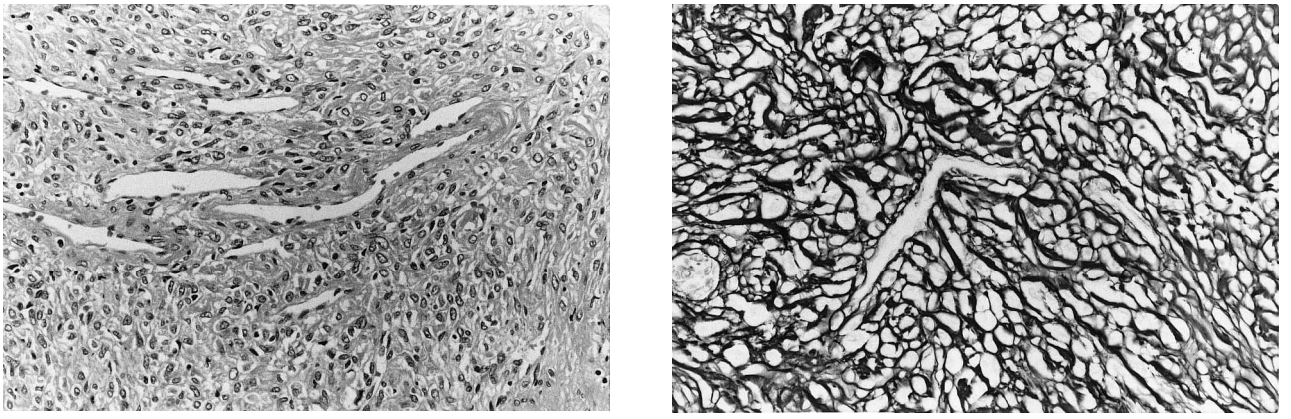

(a) (b)

FIG. 2

Histological appearance of haemangiopericytoma. (a) Microscopic view shows marked proliferation of oval to spindle-shaped cells of perithelial appearance without mitotic figures. Gaping vascular channels linked by a single layer of attenuated endothelial cells are surrounded by tumour cells (×30). (b) Reticulin preparation reveals a reticulin meshwork surrounding ramifying vessel and individual tumour cells. (×100).

Case report

A 42-year-old Japanese man presented at the Jichi Medical School Hospital with swelling in the right temporal area and a history of three prior resections for recurrent HPC of the infratemporal fossa. The first resection was performed in 1985. All prior resections were performed as simple resection without osteotomy of the zygomatic arch. Results of physical examination were unremarkable except for the swelling of the right temporal area and paralysis of the frontal ramus of the facial nerve. A magnetic resonance imaging (MRI) scan revealed a gadolinium-enhanced mass measuring 27×25 mm in the right infratemporal fossa, with destruction of the right pterygoid plates (Figure 1a) and with extension through the right base of the skull into the middle cranial fossa (Figure 1b). Angiography confirmed the tumour's vascular nature. The right internal maxillary artery was observed to be the major feeding vessel. Pre-operative embolization of this vessel was performed followed by a wide resection via the infratemporal approach type D.[7] A hemicoronal skin incision was made. The facial nerve was identified within the parotid gland and was dissected to expose the frontal ramus, that was sacrificed because it was involved with tumour. Osteotomy was carried out to release the zygomatic arch. The masseter muscle was resected and the temporal muscle was exposed. The tumour was seen to originate in the right infratemporal fossa and to involve the pterygopalatine fossa. The tumour was removed en bloc with the masseter muscle, the temporal muscle, the coronoid process of the mandible, and part of the middle cranial fossa. The photomicrograph in Figure 2a shows a marked proliferation of oval to spindle-shaped cells of perithelial appearance without mitotic figures. Gaping vascular channels linked by a single layer of attenuated endothelial cells were seen to be surrounded by tumour cells (Figure 2a). Reticulin preparation revealed reticulin meshwork surrounding ramifying vessel and individual tumour cells (Figure 2b). The diagnosis of haemangiopericytoma was confirmed by immunohistochemical tests. The patient's post-operative course was uneventful, and he remains free of disease five years after this resection.

Discussion

HPC is a rare vascular tumour that can occur anywhere in the body, but that commonly involves the soft tissues of the trunk and lower extremities. The majority of the cases involving the head and neck occur in the nasal cavity, paranasal sinuses, orbit, parotid gland, and the neck.[5,8–11] Although Buchanan[12] described a case with direct extension into the infratemporal fossa from an adjacent site, this is the first report of HPC originating in the infratemporal fossa.

Several authors have addressed the histologic grading of HPC as benign, borderline, low-grade malignant, and overtly malignant.[13–15] The mitotic rate has been used to distinguish between these categories, with lesions demonstrating over four mitoses per high power microscopic field being defined as 'malignant'.[16] However, Campango and Hyams[17] stated that most of the cases of intranasal HPC should be defined 'benign hemangiopericytoma-like tumour'. Despite the lack of mitoses in our patient's specimen, we regarded his recurrent tumour as malignant and performed a wide resection.

The surgical approach to the infratemporal fossa is difficult, as this area includes the lower cranial nerves, the sympathetic plexus, the internal carotid artery, and the internal jugular vein.[18] In previous reports, various surgical approaches to the infratemporal fossa region have been described, such as the infratemporal fossa approach,[7] the transmaxillary approach and the transfacial approach.[19] The transmaxillary approach and transfacial approach avoid the risk of damage to the facial nerves and are preferable cosmetically. However, these approaches have the disadvantages of access through potentially contaminated areas, and a lack control of the petrous portion of the internal carotid artery. The infratemporal fossa approach provides the surgeon with adequate wide access for the safe removal of massive tumours.[19] A broad surgical field should be exposed in performing a radical resection of the tumour. Even when a histological diagnosis of malignancy is not obtained, as in this case, we recommend the present approach to resecting the HPC. Absence of recurrence further supports the use of wide resection as in the present case.

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