CASE REPORT

What lies beneath

Padraig O'Sullivan, Ebere Ogbonnaya, Chandrasekaran Kaliaperumal, Charles Marks

Department of Neurosurgery,
Cork University Hospital-HSE
South, Cork, Ireland

Correspondence to

Dr Padraig O'Sullivan,
padraigosullivan8@gmail.com

SUMMARY

Haemangiopericytomas are a group of aggressive soft tissue sarcomas that originate from the pericytes in the walls of capillaries. Local invasion of the surrounding structures is not uncommon. Symptoms depend on the location, size and grade of tumour. Coexistence with a benign tumour in the same location is very rare. We report an interesting case of occipital scalp lipoma with an underlying torcular haemangiopericytoma and skull defect.

BACKGROUND

A scalp lesion may rarely overlie more significant cranial pathology, and an awareness of this is important.

CASE PRESENTATION

A 59-year-old man was referred to our neurosurgical unit, having been initially seen by a surgical team in a peripheral hospital for an occipital lump. This lump was consistent with a scalp lipoma on examination. The patient had no other medical issues and underwent a simple lumpectomy. During the procedure a skull defect was noticed beneath the lump.

INVESTIGATIONS

Histology of the scalp lump confirmed a lipoma. A CT scan of the brain was requested by the referring surgeon to further evaluate the underlying defect. A space-occupying lesion of the torcula (confluence of the venous sinuses) was noted with some erosion of the skull. We decided to manage the patient conservatively. Within a year, significant occipital swelling had developed, becoming worrisome for the patient. A repeat CT of the head revealed an interval increase in the size of the torcular lesion to 4.5×6.5 cm, with progressive destruction of the overlying occipital bone (figure 1).

TREATMENT

The patient underwent elective partial resection of the midline occipital lesion under general anaesthesia. The tumour was found to be well-encapsulated and highly vascular. The extracranial component of the tumour was excised and significant intraoperative haemorrhage was encountered, requiring transfusion of four units of red cells to the patient. We believe the torcula was invaded by the tumour given the degree of bleeding, even with partial resection. Figure 2 shows residual tumour on post-operative MRI.

Histopathological analysis revealed a primary tumour with many spindle cells and characteristic 'staghorn' blood vessels. There was marked nuclear

pleomorphism, high mitotic rate and multifocal areas of dense, pleomorphic tumour corresponding to a WHO grade III anaplastic haemangiopericytoma.

OUTCOME AND FOLLOW-UP

The patient was referred for stereotactic radiosurgery to treat the residual tumour.

DISCUSSION

Intracranial haemangiopericytomas (HPC) are malignant vascular tumours arising from mesenchymal cells with pericytic differentiation.¹ They are aggressive lesions that tend to occur at an earlier age than other meningeal tumours, recur with high frequency and metastasise extracranially.^{2–4} Because

Figure 1 (A) An enhancing midline occipital lesion (4.5×6.5 cm) centred in the calvarium with overlying soft tissue mass, bony destruction and apparent invasion of the torcula. (B) Occipital bony destruction.

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Figure 2 (A) T1 sagittal postgadolinium showing residual tumour. (B) T1 axial postgadolinium showing marked enhancement originating from the torcula. There was no definite evidence of superior sagittal sinus thrombosis. However, there was a filling defect consistent with tumour infiltration to the transverse sinuses. (C) T1 sagittal without contrast demonstrating occipital bone erosion. The residual tumour mass measured 2.6 cm (AP)×4.2 cm (transverse diameter)×6.5 cm (craniocaudal diameter). (D) T2 axial without contrast.

of their rarity, representing less than 1% of all central nervous system tumours, few guidelines exist for their optimal management.

In a retrospective single centre study of 35 intracranial HPCs, earlier recurrence was associated with tumour sizes of greater than 6 cm and locations distant from the skull base. Only 20% of the patients developed extracranial metastases. Better overall survival was associated with greater extent of resection.³ A systematic review of predictors of death following treatment of intracranial HPCs showed that gross total resection alone (n=105) resulted in overall superior survival rates (median 13 years) than if subtotal resection (n=23) was performed (median 9.75 years). Subtotal resection plus adjuvant radiotherapy led to a median survival of 6 years, suggesting that radiation did not confer a survival benefit.⁴

Intracranial HPCs present a diagnostic challenge in that they are very often mistaken for meningiomas, owing to their radiological appearance.^{5–8} A review of the CT, MRI and angiographic findings in seven pathologically proven intracranial HPCs failed to distinguish them from meningiomas, in that none showed hyperostosis or tumour calcification and all showed marked enhancement.⁷ Another study of six intracranial HPCs showed that two displayed calcification and one was eroding bone. MRI was unable to distinguish any of them from angioblastic meningioma.⁹

Intracranial HPCs can potentially occur anywhere. Although they generally arise from the dura of the parasagittal and falx regions,¹⁰ there are case reports of HPCs arising primarily in the cerebral parenchyma,^{11–13} pituitary fossa,^{10 14 15} cavernous sinus,¹⁶ cerebellum,¹⁷ pineal region,¹ ventricular system¹⁸ and trigeminal nerve.¹⁹ In addition, they may present with acute subdural haematoma²⁰ and intracerebral haemorrhage.^{21–24}

Learning points

- ▶ To our knowledge, there is no similar case reported in the literature of a histologically confirmed lipoma overlying a skull defect attributed to a malignant haemangiopericytoma of the torcula.
- ▶ This case demonstrates the necessity for surgeons and trainees to be aware of the broad differential diagnosis for scalp lesions and osteolytic lesions involving the skull; these include metastatic cancer, plasmacytoma, giant cell tumour, haemangioma, epidermoid cyst, osteogenic sarcoma, eosinophilic granuloma, aneurysmal bone cyst, atypical meningioma and fibrous dysplasia.
- ▶ Although rare, malignant haemangiopericytoma should be excluded in an osteolytic skull lesion with an associated soft tissue component.

Contributors PO'S was responsible for the composition of the case report, the literature review, image selection and subsequent revisions. EO identified the case as a novel entity, oversaw the writing of the report in its entirety and edited the final draft. CK proofread the case report prior to submission. CM was the neurosurgeon who assumed overall care of the patient and oversaw the development of the case report. All contributors were involved directly in the care of the patient.

Competing interests None.

Patient consent Obtained.

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