Unusual presentation of more common disease/injury

Haemangiopericytoma with no dural attachment

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The authors describe a case of a gentleman in his 40s who presented with a space-occupying lesion in the right precentral gyrus which showed no dural attachment and was well circumscribed, both on imaging and at surgery. This lesion was histologically challenging to diagnose but favoured to be a haemangiopericytoma. The lack of dural attachment is unusual for this tumour type but is typically associated with metastases to the brain, and less commonly a solid primary central nervous system tumour. The authors propose that haemangiopericytoma should be regularly considered as a differential diagnosis of cerebral metastases so that inadequate preoperative anticipation and planning is not encountered.

BACKGROUND

There are very few reported cases of haemangiopericytomas not showing an attachment to the dura, whether this be broad or narrow based.¹ We describe a case where the tumour was not attached to the dura, mimicking a cerebral metastasis or a solid primary central nervous system (CNS) tumour. The diagnosis of haemangiopericytoma should always be considered when imaging shows lesions similar to cerebral metastases, allowing optimum surgical management.

CASE PRESENTATION

A right-handed gentleman in his 40s presented to his local hospital with a 1 week history of dysarthria, left facial weakness and worsening fatigue. Seven months previously he had had headaches, and rapid onset left facial numbness with left hand weakness which gradually resolved and were attributed to stress, hypertension and fatigue.

INVESTIGATIONS

A CT brain and MRI brain demonstrated a 2×2 cm well-circumscribed relatively homogeneously enhancing space-occupying lesion in the right precentral gyrus. A CT chest, abdomen and pelvis did not show any evidence of primary neoplastic disease. The preoperative MRI brain scan showed no attachment to the dura (figure 1).

DIFFERENTIAL DIAGNOSIS

The principal diagnosis considered at the preoperative stage based on the imaging characteristics was an intrinsic space-occupying lesion, most likely to be a cerebral metastasis from an occult primary neoplasm. This view was unchanged on the basis of the operative findings where the lesion was found to be entirely intrinsic and distinct from the dura matter.

However, histopathological evaluation confidently excluded cerebral metastasis and intrinsic neuroepithelial tumours because of the absence of the battery of relevant

immunohistochemical markers. Instead the differential diagnoses suggested were a haemangiopericytoma and a sarcoma. The histological morphology of the tumour was more in keeping with haemangiopericytoma, and while the lesion did express epithelial membrane antigen (EMA) which is more likely to be expressed by high grade sarcomas than haemangiopericytomas, a third of anaplastic haemangiopericytomas (WHO Grade III) also express EMA.² Furthermore, a whole body positron emission tomography scan showed no evidence of fluorodeoxyglucose avid lesions, excluding a diagnosis of sarcoma.

Figure 1 Preoperative MRI brain showing a lesion with no attachment to the dura.

Figure 2 Postoperative MRI brain showing complete surgical excision of the lesion.

TREATMENT

The patient underwent a neuronavigated surgical resection of the lesion approached through a short corticotomy. At surgery no dural attachment was encountered. There was a clear plane between the tumour and surrounding brain. A complete en bloc macroscopic excision was achieved. The postoperative MRI brain is shown in figure 2.

OUTCOME AND FOLLOW-UP

The patient made a good postoperative recovery with all prior neurological deficits recovered but complicated by transient left faciobrachial focal motor seizures. He received postoperative radiotherapy. Unfortunately, the patient died 1 year after surgical resection of the tumour. His cause of death was not established at postmortem.

DISCUSSION

This case is informative because atypically the haemangiopericytoma was not attached to the dura. Previous descriptions of this rare tumour have described a clear attachment to the dura, both on imaging and at surgery.³ Both the preoperative imaging and findings during the surgery showed that this tumour was subcortical, making it more likely to have been a metastasis to the brain.

Haemangiopericytomas are rare intracranial tumours which resemble meningiomas. The WHO Classification of CNS tumours (2007) graded diffusely infiltrative haemangiopericytomas with cytological atypia as WHO grade II (diffuse haemangiopericytomas) and those showing anaplasia and mitotic activity were classified as WHO

grade III (anaplastic haemangiopericytomas).⁴ They are highly vascularised tumours thought to originate from pericytes and account for less than 1% of intracranial tumours. Patients with haemangiopericytomas usually present in their fourth and fifth decades. Headache is the most common symptom caused by haemangiopericytomas; seizures are rare.

Identifying this tumour early is important as haemangiopericytomas show a high rate of recurrence, often with distant metastases. Studies have suggested mean recurrence rates of haemangiopericytomas following treatment at one, five and fifteen years as 3.5%, 46% and 92% respectively.⁵

Differing surgical planning is needed to remove a tumour that is not attached to the dura compared to a dural-based neoplasm. By considering the diagnosis of haemangiopericytoma when preoperative imaging suggests a cerebral metastasis, surgery can be planned more carefully and a complete surgical excision of the lesion is more likely to be achieved. Evidence suggests that complete surgical excision of haemangiopericytomas is a crucial factor in prolonging survival.⁶ Postoperative radiotherapy also improves prognosis. It remains to be seen whether a haemangiopericytoma not attached to the dura has a different prognosis to one with a dural attachment.

Learning points

- ▶ Haemangiopericytomas are rare intracranial tumours, similar to meningiomas, and usually show an attachment to the dura.
- ▶ Haemangiopericytomas are aggressive tumours with a high rate of recurrence, often with distant metastases.
- ▶ Rarely haemangiopericytomas may not be attached to the dura so will resemble a cerebral metastasis or solid primary CNS tumour.
- ▶ Always consider the diagnosis of haemangiopericytoma in lesions resembling cerebral metastasis so that effective surgery can be planned.

Competing interests None.

Patient consent Not obtained.

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