

CASE REPORT

INTRACRANIAL HAEMANGIOPERICYTOMA: A VERY RARE ENTITY HAVING A HIGH MALIGNANT/METASTATIC POTENTIAL

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Intracranial haemangiopericytomas are very rare tumours. Their radiological appearance resembles that of meningioma. Recommended treatment is total surgical excision, if possible, followed by radiotherapy. A vigilant, long term clinical and radiological follow up is very essential due to a high rate of late onset recurrence along with neural and extraneural metastases. A case report and review of literature is presented.

Keywords: Intracranial haemangiopericytoma, recurrence, radiotherapy, metastasis

INTRODUCTION

Haemangiopericytomas are uncommon central nervous system (CNS) tumors of vascular origin. These have only recently been included in a specific group of CNS tumors (WHO classification 1993) and subsequently as a group by itself (WHO classification 1997 and 2000), while before these were confused with meningiomas. Haemangiopericytomas may also arise in the neck, trunk, skin, retroperitoneum and oral cavity. These are very uncommon in childhood.¹⁻³

CASE SUMMARY

A 22 years old young, healthy male presented with three months history of headache, vomiting, ataxia and deterioration of vision. On examination, he had markedly deteriorated vision in both eyes and there was gross papilloedema. Cerebellar signs were positive on right side along with ataxia.

CT scan brain with contrast showed: "A large lobulated mass in the posterior fossa with cystic component. It is slightly eccentric to the right paramedian. There is dense contrast enhancement of the solid component and walls of the cystic areas. Mass effect with displacement of the fourth ventricle has resulted in obstructive hydrocephalus (Figure-1). Impression: Haemangioblastoma; Suggested to have an MRI."

MRI showed: "An extra-axial posterior fossa mass along the midline projecting against the vermi of the cerebellum, primarily originating from the dura in the vicinity of the torcula. It is inseparable from the distal part of straight sinus. It has quite a heterogeneous morphology with cystic component as well. However, it enhances quite strikingly, unlike any primary glial cell tumour of the cerebellum. The paucity of significant surrounding oedema despite the large size of the lesion is also supportive of extra-axial lesion besides the intervening prominent blood vessels. Marked mass effect on the fourth ventricle has resulted in significant proximal ventriculomegaly (Figure-2). The basal CSF spaces are appreciably obscured. I strongly suspect this

is a meningioma originating from the under surface of the tentum adjacent to torcula."

After adequate preparation (including sufficient blood for intra-operative transfusion), the patient was operated. With the patient in prone position, a right occipital burr hole was made to drain the ventricles. Midline posterior fossa craniectomy was performed. An encapsulated, soft, very vascular tumour having its attachment to the under surface of tentorium along the straight sinus was excised completely and the site of attachment was diathermised.

Postoperative course was smooth. CT scan brain done on third postoperative day showed resolved hydrocephalus and no local collections. His vision started improving in the early postoperative period and cerebellar signs improved as well.

Histology showed: "A cellular tumour composed of monotonous sheets of tumour cells interrupted by numerous slit-like vascular spaces lined by flattened endothelial cells. Individual tumour cells have oval nuclei with inconspicuous nucleoli and scant cytoplasm. Up to 7 mitoses/HPF are noted. No nuclear pseudoinclusion, psammoma bodies or necrosis seen. Features are suggestive of haemangiopericytoma."

This diagnosis was confirmed by immunohistochemistry, in which epithelial membrane antigen (EMA) and Chromogranin were negative while CD34 was positive.

Postoperatively, the patient received full course of radiotherapy. CT scan brain with contrast, done 3 months after surgery, did not show any residual/recurrent disease in the operative bed. The ventricular system was normal (Figure-3). Till now, nearly three years after surgery, the patient has been fine and on his preoperative job. Postoperative MRI brain with contrast, done at yearly interval, is normal. A vigilant, long term clinical and radiological follow up is planned to watch for any local recurrence and/or distant metastases.

Note: Due to the usual expected confusion in the diagnosis of haemangiopericytomas, I have intentionally 'copied' the radiological and histology reports so that the reader gets the exact idea of the problem while dealing with such lesions.

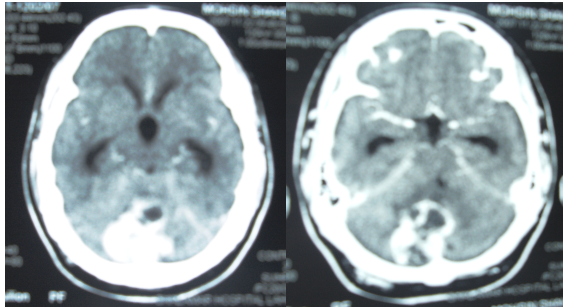


Figure-1: CT scan brain with contrast showing the posterior fossa haemangiopericytoma with hydrocephalus

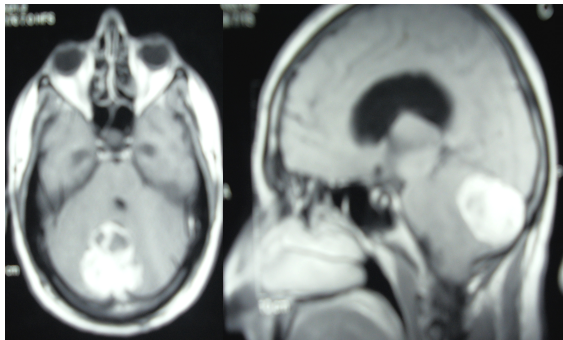


Figure-2: MRI brain with contrast showing the posterior fossa haemangiopericytoma with hydrocephalus

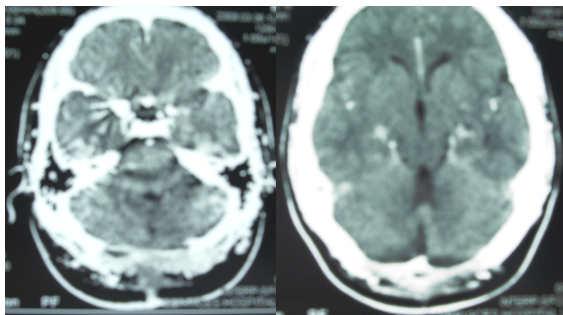


Figure-3: CT scan brain with contrast (done 3 months after surgery) showing no residual tumour and no hydrocephalus

DISCUSSION

The usual presentation of intracranial haemangiopericytoma is headache, vomiting, fits, visual deterioration or focal neurological deficits. Aggressive growth, tendency to local recurrence and relatively frequent metastases are significant features of this tumour. It can rarely present as intracranial haemorrhage which can be life-threatening.^{1,2,4}

Haemangiopericytoma arising within the sella turcica is very rare but till now 9 cases have been reported and it should be considered as very rare differential diagnosis of sellar and suprasellar masses.⁵

Plain x-rays and angiography may distinguish haemangiopericytoma from meningioma. A well-defined, lytic destruction of the adjacent skull favours meningeal haemangiopericytoma whereas hyperostosis supports meningioma. CT scan and MRI picture of haemangiopericytoma resembles that of meningioma (well-defined tumour with dural attachment and strong contrast enhancement), but unlike meningioma, meningeal haemangiopericytoma appears to lack calcification.¹

Haemangiopericytomas are highly cellular neoplasms composed of plump cells with scant cytoplasm. A dense reticulin network typically investing individual cells is one of the most characteristic features of the tumour. Haemangiopericytomas have a particular immunohistochemical profile which aids in its diagnosis. The tumour cells react with antibodies against vimentin and, in most cases, against CD34, but unlike meningiomas, they lack EMA. They also lack immunoreactivity for cytokeratins, carcinoembryonic antigen (CEA) and Von Willebrand factor.¹

Complete surgical excision of the tumor is usually the treatment of choice. Postoperative radiotherapy is strongly recommended even after an apparently complete tumour resection and must be given after subtotal tumour resections. Preoperative radiotherapy has also been used effectively in high risk surgical cases.^{1,2}

Stereotactic radiosurgery is indicated for recurrent intracranial haemangiopericytomas measuring less than 25 mm in greatest diameter.¹ In a study, Gamma knife radiosurgery provided local tumour control in 80% of recurrent haemangiopericytomas.⁶

Chemotherapy does not appear to have an established role in the primary management of these tumours. However, in patients with disseminated distant metastases, palliative chemotherapy has been tried.²

Prediction of patient outcome is difficult. However, rapid progression is correlated with increased mitotic rate (≥ 5 mitotic figures per HPF), high cellularity, nuclear pleomorphism, haemorrhage and necrosis. Ten year survival rate varies from 27–40%.^{1,2}

Intracranial haemangiopericytomas can recur locally or distantly in the neural axis or as extraneural distant metastases. In a report of 29 cases of haemangiopericytoma, 14% developed spinal and 55% extraneural metastases.^{3,7}

Local recurrence of intracranial haemangiopericytomas is almost inevitable (up to 91%) while extracranial metastases occur in 68% of cases after 15 years.¹

Metastases to bone, liver, lung, kidney and skin have been reported more than 10 years after the initial intracranial surgery. Moreover, a very rare case

of intradural metastasis to the cauda equina has also been reported. The prognosis becomes poorer and, combined with effective radiation therapy, surgical intervention should be carefully considered.^{2,4,8-10}

CONCLUSION

Intracranial haemangiopericytomas are very rare tumours and their management includes aggressive surgical resection and postoperative radiation therapy. Moreover, a life-long vigilant clinical and radiological follow up is mandatory due to very high malignant and metastatic potential of this tumour years after the initial diagnosis.^{1-3,6}

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