

Supratentorial Hemangioblastoma: A Case Report and Review of Literature

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ABSTRACT

Hemangioblastomas are benign lesions and very vascular. More common in posterior fossa and young people and constitute 2% of all intracranial tumours. They can be rarely found in the supratentorial compartment also. Till date 131 cases of supratentorial hemangioblastomas have been reported in the literature. They carry good prognosis and some are associated with von Hippel Lindau disease i.e Pancreatic tumours, Renal cell carcinoma or cysts, Retinal hemangioblastoma and intracerebral hemorrhage. And sometimes they are not associated with vHL disease. Clinical radiological and histological characteristics have been discussed and literature has been reviewed. Surgical excision is the only option followed by long term follow up using MRI and USG abdomen to detect newer lesions in other organs.

Keywords: Hemangioblastoma, supratentorial, vHL disease, posterior fossa.

INTRODUCTION

Supratentorial hemangioblastomas are exceedingly rare. Because of the rarity of hemangioblastomas in this location, the literature is primarily composed of case reports and small series. Little information is available regarding the clinical features of these tumors, particularly in comparison with lesions found in other locations. We report a case of Supratentorial hemangioblastomas of left parietal lobe not associated with VHL disease^[1].

CASE REPORT

A 50 year male patient presented with headache, vomiting, giddiness and weakness of right upper limb of 10 days duration and no other neurological defects. CT scan of brain showed a cystic lesion of 5x4.5cms size with a solid contrast enhancing nodule at the periphery. There is no

mass effect or perilesional oedema. MRI was not done due to financial restraints. Blood picture was normal, ultrasound abdomen was normal. After complete work up, left parietal craniotomy was done and the tumour was completely excised after aspiration of the cystic part. Histological report confirmed capillary hemangioblastoma. Immunohistochemistry was not done due to non availability in the institute.

DISCUSSION

Hemangioblastomas are rare in the supratentorial compartment. So far only 131 cases have been reported in the literature. We report one such case which is not associated with vHL disease in a 50 year male patient. The incidence is 2:1 (male: female). Clinically they cannot be differentiated from other space occupying lesions like Astrocytomas, Meningiomas and Ependymomas. Clinical presentation mainly depends upon the location of the

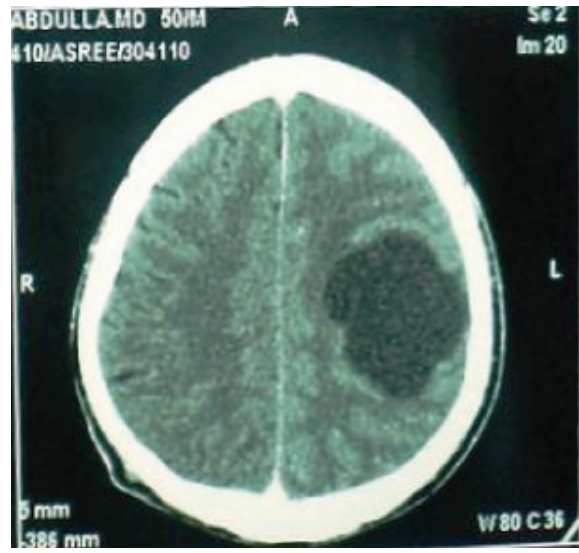


Figure 1: CT scanshowing brain plain showing a cystic lesion in the left parietal lobe.

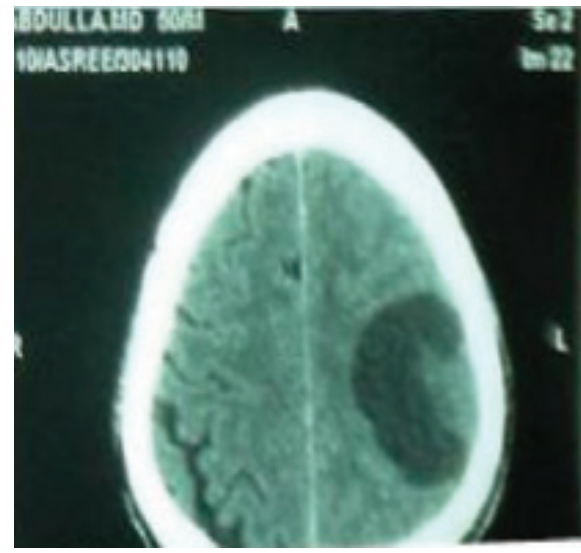


Figure 2: CT scan brain contrast showing enhanced mural nodule at periphery.

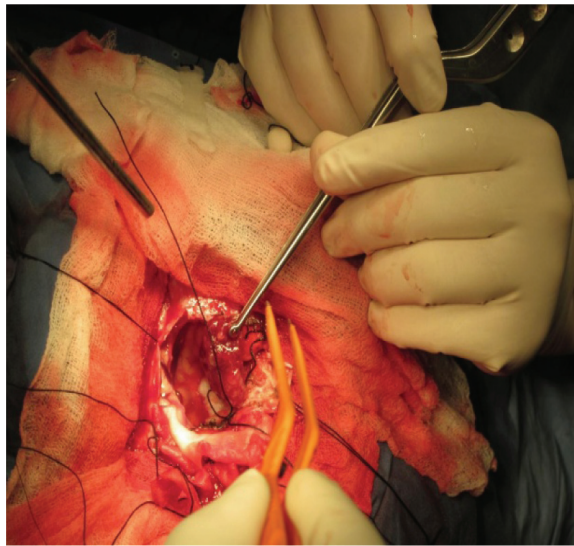


Figure 3: showing gross lesion on the table

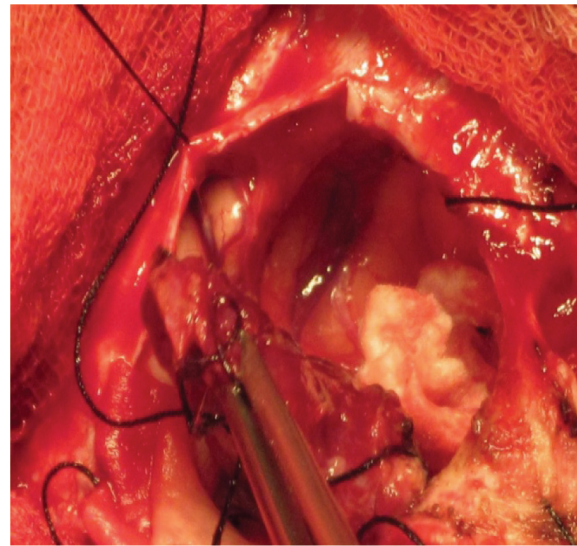


Figure 4: magnified view of the tumour mass

lesion. Memory disorders, motor and sensory defects, increasing head circumference in infants and spontaneous intracerebral bleed are less common. If associated with vHL disease the renal cell carcinomas or cysts, pancreatic tumours or cysts can be noted on the USG abdomen.

Histologically, they are composed of rich network of

blood vessels with a single layer of plump uniform endothelial cell and polygonal stromal cells with swollen foamy cytoplasm. The blood space may extend into the large sinuses. Immunohistochemically the stromal cells of hemangioblastomas react positively to inhibin alpha CD34.

Radiologically they can be single or multiple, cystic or solid lesions, with or without meningeal involvement. Cystic hemangioblastoma can present with large cyst and a small nodule. Epidermoid cysts and arachnoid cyst or cystic astrocytomas should be thought of in differential diagnosis. Surgical excision is the only treatment. Regular follow up with MRI brain and USG abdomen to detect new lesions in other organs on long term basis is recommended.

CONCLUSION

Supratentorial Hemangioblastomas (HBLs) are very rare. Only 131 cases have been reported in the literature so far. Prognosis is good, one has follow up the patients with MRI brain and USG abdomen to detect newer lesion in other organs. Lifelong follow up is recommended more so if associated with vHL disease.

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