

## Cerebral Hemangioblastoma: A Rare Case Report



### Medical Science

**KEYWORDS :** Hemangioblastoma, Temporo-parietal, Cerebral

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### ABSTRACT

*Hemangioblastomas are rare, benign tumors of which most common site is cerebellum and spinal cord. Here we report a rare case of hemangioblastoma in left temporo-parietal region which is the site. A 45-year-old female presented with history of headache, dizziness and vomiting. MRI demonstrated a well defined extra-axial lesion in left temporal lobe anteriorly at the base of skull. Surgically the single tissue mass was removed which was well encapsulated, firm, grey-brown in colour. On frozen sections, we had given the differential diagnosis of Astrocytoma and hemangioblastoma. But on histopathological examination and special stain study we reached to the final diagnosis of hemangioblastoma. Hence the final diagnosis should not be given on the frozen section study only*

### INTRODUCTION

Hemangioblastomas are rare, benign tumors occurring in any part of the nervous system. Most are found as sporadic tumors in the cerebellum or spinal cord. However, these neoplasms are also associated with von Hippel-Lindau disease. Hemangioblastomas (HBLs) are World Health Organization (WHO) grade I capillary-rich neoplasms of the central nervous system (CNS) accounting for 1-2.5% of all intracranial neoplasm(11). They can occur either sporadically (60-75% of cases) or in the context of von Hippel-Lindau (VHL) disease, the multi-organ neoplastic disorder that is inherited in an autosomal dominant manner (25-40% of cases)(11,12). The cerebellum is the most frequent location of these tumors; however, they may also occur in other regions of the CNS, medulla, and spinal region. We report a rare case of hemangioblastoma in left temporo-parietal region.

### CASE REPORT:

A 45-year-old female presented with a 2 month history of headache and dizziness and vomiting. Her past medical history was unremarkable. Her family history was not significant. Pituitary function was normal. There was no neurologic deficit.

### INVESTIGATIONS:

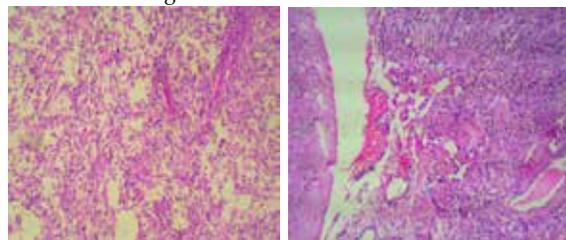
Brain magnetic resonance imaging (MRI) demonstrated a well defined extra-axial 0.9x2.2 cm lesion in base of skull in left temporal lobe anteriorly with buckling of adjacent grey matter cortex On the T2-weighted images.



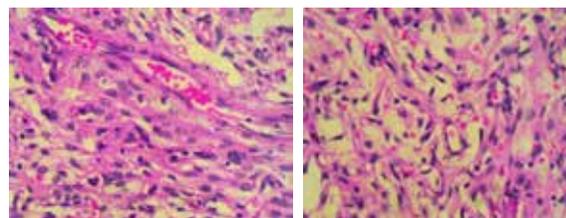
**GROSS FEATURES :** We received single, well encapsulated, grey-brown tissue mass, firm in consistency, measuring 2.6x2x0.5 cm.

**MICROSCOPIC FEATURES :** H and E stained sections studied show two components, Stroma and capillary network. Stromal cells arranged in cohesive nests and lobules. Individual stromal cells were large, vacuolated, round to oval with nucleus showing moderate degree of pleomorphism with coarse chromatin and some having prominent nucleoli with foamy cytoplasm. Numerous blood vessels forming anastomosing network of delicate capillary like channels are also seen.

Special stain study with Vimentin shows positivity which confirms the diagnosis.



(a) and (b) Low power view showing stromal and capillary rich network



(c) & (d) high power view(40x) showing large, vacuolated tumour cells with delicate capillary channels

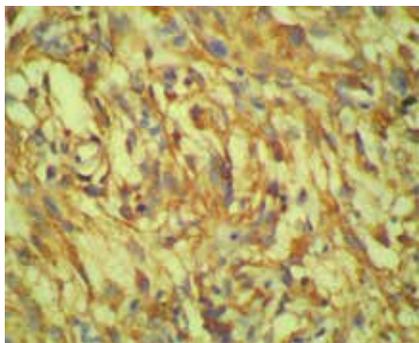


Fig. e) Section show positivity for vimentin

## DISCUSSION

Hemangioblastomas are tumors of the central nervous system that originate from the vascular system usually during middle-age. These are benign, highly vascular neoplasms of uncertain histogenesis(10). Sometimes these tumors occur in other sites such as the spinal cord and retina.[3] They may be associated with other diseases such as polycythemia (increased cysts and Von Hippel-Lindau syndrome (VHL syndrome). Hemangioblastomas are most commonly composed of stromal cells in small blood vessels and usually occur in the cerebellum, brain stem or spinal cord. They are classed as grade one tumors under the World Health Organization's classification system. While HBLs are frequently located in the posterior fossa, they may rarely be originated from their supratentorial counterpart(5,14). They occur sporadically or as a manifestation of VHL disease with an autosomal dominant inheritance(2,11,12). Generally, HBLs are distributed in a highly conserved, region-specific manner within the CNS that includes the retina, brainstem, spinal cord, and cerebellum regardless of genetic predisposition(10). Therefore, supratentorial HBLs including pituitary stalk HBLs are rare, accounting for only 2.1-2.9% of all HBLs(3,8,11,14)

Cushing and Baily inferred that hemangioblastoma do not occur in the cerebrum, while Russell and Rubinstein consider these tumors to be extremely rare in the cerebral hemisphere.(1)

We report a case of hemangioblastoma in temporoparietal region, of which site is rare. On frozen sections, we had given the differential diagnosis of Astrocytoma and hemangioblastoma. But on histopathological examination and special stain study we reached to the final diagnosis of hemangioblastoma. This is because of bursting of lipid during frozen section.

## Conclusion:

Hemangioblastoma are one of the rarest central nervous system tumors, accounting for less than 2% of which the site (Temporo-parietal) is also rare. Fibrillary astrocytoma should be kept in mind as a differential diagnosis and final diagnosis should not be given on the frozen sections. Despite the danger of potential complications due to excessive vascularity, the tumor in our case was successfully removed without a significant complication but patient died in the ICU within two week after surgery.

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