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Original Research Paper

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SPORADIC SUPRATENTORIAL HEMANGIOBLASTOMA WITH SUB EPENDYMAL EXTENSION: A CASE REPORT

Dr Rajesh Kumar Barooah — Associate Professor Department of Neurosurgery, Gauhati Medical College & Hospital.

Dr. Nayan Jeet Deka* — Senior Resident Department of Neurosurgery, Gauhati Medical College & Hospital.*Corresponding Author

ABSTRACT

Objective: To report a rare case of supratentorial hemangioblastoma, its management and outcome. **Background:** A 23 year old female presented with progressively increasing headache for last 4 months. Clinical examination revealed hemiparesis on right side of the body with bilateral grade 2 papilledema. Patients' blood parameters and other investigation did not suggested vhl disease. **Conclusion:** Hemangioblastoma is a histologically benign tumor that most commonly occurs in the cerebellum. Extracerebellar location is rare. Since it's a benign lesion surgical cure is possible only with complete excision of the mural nodule.

KEYWORDS : supratentorial hemangioblstoma, hemipareisis

INTRODUCTION

Hemangioblastoma of central nervous system is a peculiar capillary reach neoplasm [1]. It comprise of approximately 1-3% of primary CNS tumors. Hemangioblastoma can occur from any region of the CNS, but are more common in the posterior fossa (5-15%). [2] The next common location is the spinal cord, other less common location is the brain stem and Supratentorial compartment.[3]

From 1902-2021 only 136 cases of Supratentorial Hemangioblastoma has been reported.[4] Hemangioblastoma can occur in sporadic or in association with von Hippel-Lindau disease (30-35%). [5, 6] Although this tumor is a benign slow growing tumor, its pathogenic action is derived from the mass effect of the tumor and peritumoral edema.

Report of a case

A 28 year old female was admitted in our hospital with progressively increasing intensity of headache with associated nausea and vomiting for 4 months. Headache was projectile and occurred mostly during the morning hours and vomiting use to decrease the intensity of headache. Patient also developed weakness of the right side of the body. Her power on the right upper limb was 4/5 and right lower limb was 4+/5 (on MRC scale). Right side plantar was Babinski positive. Fundus examination revealed bilateral grade 2 papilledema and rest finding was normal. Haematological studies showed; Rbc count 3.8×10^6 and haemoglobin level was 12.3 g/dl and haematocrit was 42.2% (all were within normal limits). Ultrasound of abdomen and pelvis was also normal. MRI revealed a well-defined loculated cystic lesion with an internal solid component in the left parasagittal high fronto-parietal lobe extending to sub ependymal region with mild diffusion restriction and post contrast enhancement of the solid component. (*Figure 1*)

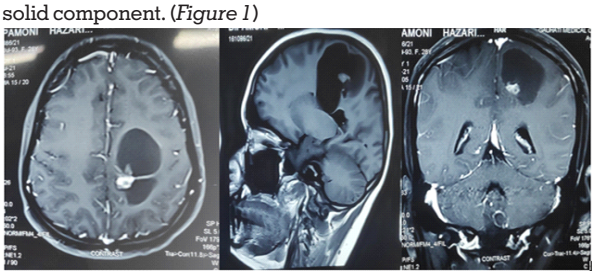

Figure1: MRI showing T1W1 plain and contrast imaging showing a cystic lesion in the fronto-parietal region with an enhancing mural nodule on contrast

During surgery a fronto-parietal craniotomy was made. Dura was incised in a u shaped manner and everted towards the sagittal sinus. Trans-sulcal dissection was carried out and xanthochromic fluid was drained, the mural nodule was identified and was excised careful as a whole. Blood loss minimal and the mural nodule was send for histological examination. (Figure 2)

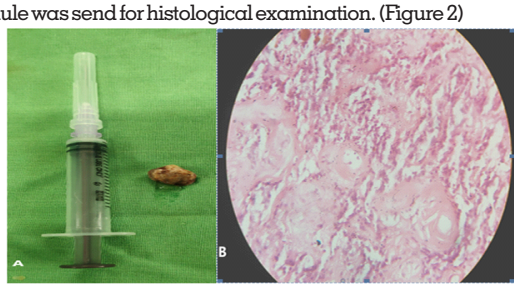

Figure 2: *(A) Showing excised mural nodule of size approximately 2.5× 2 cm. (B) H & E stain showing neoplastic stromal cells arranged between numerous small vessels*

During the post-operative period the patient was given anti-epileptics and the post-operative period was uneventful. Immediately after operation there was was no improvement of power however on follow up of two months the power in the right upper limb is 4=/5 and the lower limb is 5/5 and no papilledema present. Headache improved immediately after the surgery and the patient was discharged on day 7 post-operative period.

Pathological finding

For definitive examination, the tissue was fixed in 10% buffered formalin, routinely processed, and embedded in paraffin. Paraffin-embedded sections were stained with hematoxylin-eosin. There were numerous thin walled, fairly closely packed small blood vessels lined with plump endothelial cells. The vessels were separated by numerous vacuolated stromal cells, no mitotic activity was found. (*Figure 2*)

DISCUSSION

Hemangioblastoma is a histologically benign tumor that most commonly occurs in the cerebellum. Extracerebellar location is rare. Patients with VHL disease generally present with more than 1 tumor and are young and may present with Hemangioblastoma in unusual location. However VHL disease must be ruled out in each patient with CNS Hemangioblastoma.

There are several theories that have been proposed for this tumor, including vasoformative elements like endothelium/ pericytes/ astrocytes precursor cell which leads to its genesis.

However all theories are controversial. [7]

Hemangioblastoma frequently presents as a mural nodule projecting into a cyst. The cyst trends to be well demarcated. In general when we find a cyst with mural tumor nodule the primary diagnosis that comes to our mind is pilocytic astrocytoma and other tumors like ependymomas & chroid plexus papilloma [8].

During surgery surgeon must always try for complete excision of the mural nodule. Direct biopsy of the nodule must be avoided as it may lead to massive bleeding from the neocapillaries. This mural nodule must be approached from the brain tumor interface and all feeding vessels must be coagulated circumferentially and also from the base.

The tumor in this patient is one of the rare examples of Supratentorial Hemangioblastoma. Although rare in the supratentorial location, Hemangioblastoma must be considered in the histopathologic differential diagnosis of especially cystic tumors with mural nodule.

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