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Hemangioblastoma of The Fourth Ventricle: A Case Report

KEYWORDS: Hemangioblastoma, Fourth ventricle, posterior fossa.

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ABSTRACT *Hemangioblastomas (HMB) are one of the most frequently occurring vascular tumors of the Central nervous System (CNS). Histologically they are benign tumors representing 1-2% of all intracranial neoplasms but when posterior fossa tumors are considered alone, the percentage rises to 7.3. Most commonly they develop in the cerebellum of male adults in the third through fifth decades of life. Here we report a case of 60 years old male who presented with headache for past 2 years and weakness in lower limbs for last 8 months; his neurological examination revealed 12th cranial nerve involvement, power in lower limbs 3/5, superficial reflexes diminished & deep reflexes exaggerated. MRI showed a well defined mass showing peripheral enhancement, cystic in nature involving left cerebellar hemisphere & vermis which was causing compression of fourth ventricle. A sub occipital craniotomy was done and a diagnosis of hemangioblastoma was made on histopathological examination.*

Introduction

Hemangioblastomas are exclusive to the central nervous system (CNS) accounting for 2% of all primary intracranial tumors and approximately 10% of adult posterior fossa tumors. [1] They are benign, slow growing tumors that often contain both solid and cystic components. [2] They occur predominantly in the third to fifth decades of life and are more common in males than in females. 90% of Hemangioblastomas begin in the posterior fossa, of which 70 to 80 % are located in the cerebellar hemispheres, 10-15 % in the cerebellar vermis and 10 % in the brain stem. Hemangioblastomas appear in multiple sites in about 12 % of cases. [2] Other sites for the formation of hemangioblastomas include the cervical spinal cord and cerebrum.[2] Capillary hemangioblastomas may be associated with retinal hemangioblastomas or a variety of extra-central nervous system lesions including cysts of the liver, kidney or pancreas, renal cell adenomas, renal cell carcinomas, epididymal papillary cystadenomas and pheochromocytomas.[1] They occur either sporadically or in association with Van Hippel-Lindau (VHL) disease. VHL-disease is a heritable systemic syndrome that manifests in the CNS with multiple intracranial and retinal Hemangioblastomas. Here we report a case of capillary Hemangioblastoma of fourth ventricle which was diagnosed in the department of Pathology and the histopathological and clinical features are discussed.

CASE HISTORY

A 60-years-old male presented with headache for the past two years along with weakness in lower limbs for the last 8 months. Headache was sudden in onset and progressive in nature. Weakness in the lower limb was also sudden in onset and progressive in nature causing inability to walk. Patient also lost the bladder and bowel control for the last two days. His higher CNS functions were normal. There was 12th cranial nerve involvement. Power in the lower limb was 3/5, superficial reflexes were diminished and deep reflexes were exaggerated. The rest of his neurological and general physical examinations were unremarkable. There were no signs of meningeal irritation.

Investigations

Hemoglobin – 16.2gm %

All other routine haematological & biochemical parameters were within normal limits.

Radiological investigations

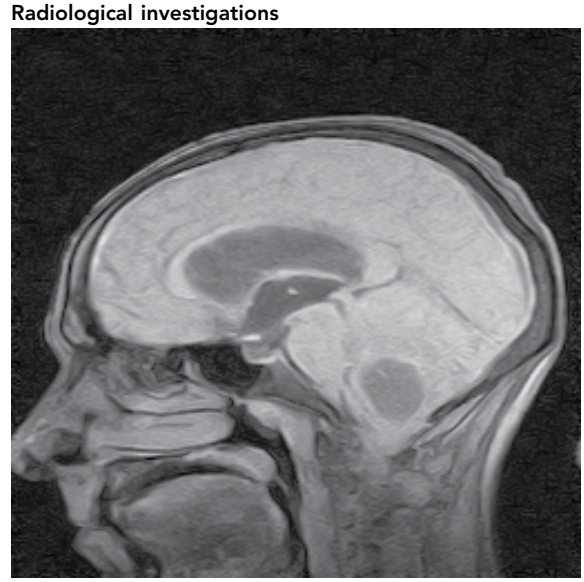

Fig-1

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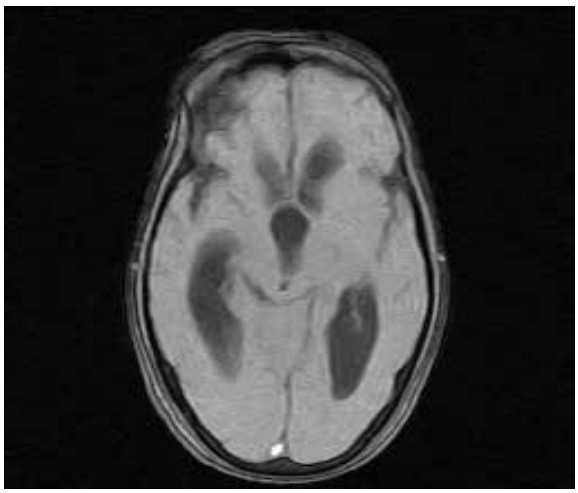

Fig-2

The CT scan was unremarkable, and MRI showed a well defined posterior fossa mass showing peripheral enhancement, cystic in nature and involving left cerebellar hemisphere and vermis which was causing compression of fourth ventricle. (Fig 1, 2)

MRI brain showed a well defined posterior fossa mass showing peripheral enhancement, cystic in nature, involving left cerebellar hemisphere and vermis which was causing compression of fourth ventricle

A sub occipital craniotomy was performed. Tumor was soft in consistency and highly vascular.

Neuropathology
Sections from tumor tissue displayed proliferation of both stromal and capillary like channels comprising vascular elements. There is proliferation of many capillary like channels of small to medium caliber blood vessels with plump, hyperplastic endothelial cells. The stromal cells had pleomorphic nuclei and lipid containing abundant pale cytoplasm along with few lipoblast like cells. Mitoses were not seen, and necrosis was absent. Scattered reactive inflammatory cells were present. [Fig 3&4]

Postoperative Course.
Postresection MRI revealed a small region of nonehancing residual tumor within the optic canal.

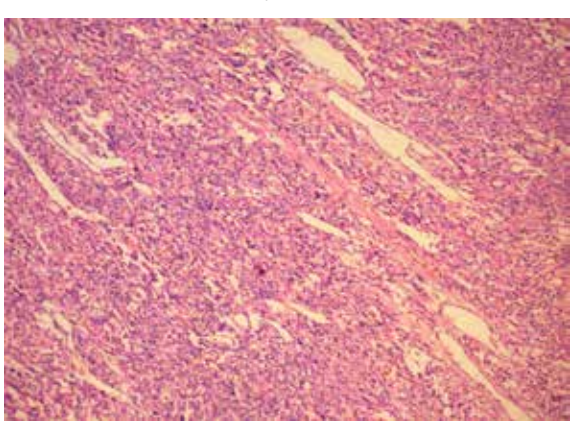

Fig-3 (10X, H&E stain) Photomicrograph shows section at low magnification, a highly vascular lesion

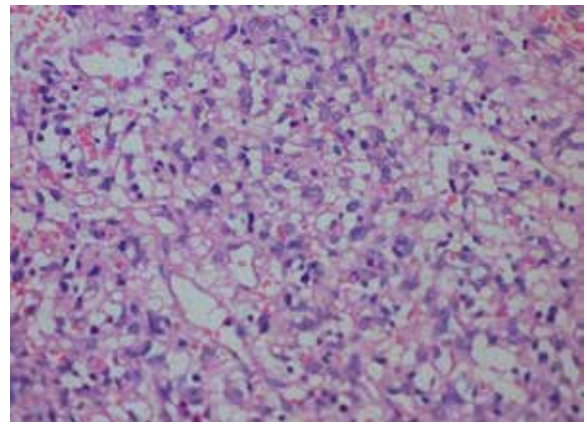

Fig-4 (40X, H&E stain) Photomicrograph at higher magnification shows numerous vascular channels lined by endothelial cells & stromal cells. Stromal cells are oval to round with clear cytoplasmic vacuoles

DISCUSSION
Hemangioblastomas [HMB] are the infrequent benign tumors of the CNS and account for 1.1-2.4% of all intracranial growths. They occur in any age group, but the young & middle- aged adults are most commonly affected, with a peak incidence occurring in fourth decade. Around 80% of Hemangioblastomas arise in the posterior fossa. Frequency of Hemangioblastoma among primary tumors of the posterior fossa is 7.3% in adults. [3] Most common location is in cerebellum; where their classical presentation is cystic with a mural tumor nodule. Non cystic, solid tumors are less common in this location. Cystic lesion in the cerebellum need to be carefully examined which may cause sudden death due to compression of brain stem. [4]Other less common location is vermis which constitutes around 13-14% but these may deform & obstruct the cavity of the fourth ventricle. In the present case there was a well defined posterior fossa mass involving left cerebellar hemisphere and vermis which was causing compression of fourth ventricle. Other rare sites are tonsils & cerebellopontine angle, medulla and spinal cord.

Signs & symptoms depend on the location of the lesion, whether the tumor is unifocal or multifocal and presence or absence of a significant cystic component. [5] Macroscopically, Hemangioblastoma is a well defined tumor having both solid & cystic components. Solid component known as mural nodule has a bright yellow color due to its lipid content, which shows prominent vascular features on angiography & contrast enhancement on CT scan.

Microscopically, two major elements of HMB are endothelial cells & stromal cells which may show variation in distribution. In the classic case, delicate capillaries and larger blood spaces form a fine meshwork while cords & clusters of stromal cells fill the intervascular areas known as reticular pattern. The stromal cells are rounded, oval or polygonal and characteristically contain clear, cytoplasmic vacuoles giving foamy appearance, which is due to cytoplasmic vacuoles. Mitoses are rare; the origin of stromal cells is controversial. According to most of the literature reviews, the stromal cells have both glial and vascular origin. [1] The expression of growth factors & growth factor receptors have been studied by Bohling et al. [6] and it has been found that the stromal cells express abundant epidermal growth factor receptor (EGFR) and some platelet-derived growth factor receptor –α (PDGF-α). [7] Although stromal

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cells display a staggering variety of antigenic reactivities, some of which may be useful in diagnosis but none of which has resolved the elusive histogenesis of these cells. CXCR4 and VEGF may collaborate to induce angiogenesis in hemangioblastomas. [8] Hemangioblastoma of the fourth ventricle are associated with higher mortality and postoperative morbidity than those in unusual locations, but several cases of successfully removed lesions have been reported. [9]

In differential diagnosis of Hemangioblastoma metastatic renal cell carcinoma(RCC) to brain or spinal cord must be considered due to cellularity, vascularity, clear cell appearance and nuclear atypia displayed by some Hemangioblastoma which may create difficulty in their distinction. Renal cell carcinoma shows mitotic activity, prominent nucleoli with the formation of glands and papillary structures. The epithelial character of metastatic renal cell carcinoma is revealed by positive immunoreactivity for EMA, cytokeratins and negative reactions for NSE & inhibin A, while HMB will show opposite immunoprofile.

Cellular variant of HMB may mimic a paraganglioma when the former presents as an extra axial spinal cord lesion. Paragangliomas are usually strongly immunoreactive for chromogranin while HMB are negative.

Vascular tumors which are dura based such as hemangiopericytomas and vascular meningiomas are sometimes included in the differential diagnosis. The former is a radiographically aggressive appearing dura based tumors, having characteristic staghorn vascular pattern of tumor cells and no vacuolization on histology while vacuolization is present in vascular meningiomas and they also show characteristic meningothelial whorls & psammomatous calcifications.

Conclusion

In conclusion, hemangioblastomas are richly vascular, slow growing benign lesions having both cystic as well as solid (mural) component morphologically. Signs & symptoms depend on the location of the lesion, whether the tumor is unifocal or multifocal and presence or absence of a significant cystic component. Hemangioblastomas show excellent prognosis with sufficient surgical excision, but recurrences may occur with insufficient excisions.

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