Solid vascular tumor of the posterior fossa

Hemangioblastoma is a very rare occurring with incidence of 1–3% of all tumors of the CNS and 7.5% of posterior fossa tumors.5 Hemangioblastomas very rare in children: In children the prevalence is lower than 1/1,000,000, and the tumor generally is correlated with VHL.

Association with VHL nearly 50%: Hemangioblastoma can occur sporadically (50–65% of the cases).4 The prevalence of VHL is 1/35,000-50,000.8

Hemangioblastoma is typically diagnosed during early adulthood (third/fourth decades). The mean age at presentation is significantly lower in patients with associated VHL (33 years) than in patients with sporadic cases (43 years).11 In children, the diagnosis is typically made during adolescence rather than during early childhood. A slight male predominance is reported.7

CT scan, MRI and USG are important imaging method, which can demonstrate characteristic patterns of tumor and association with VHL disease. Nevertheless, the confirmed diagnosis still depends on histological examinations. This case of hemangioblastoma was operated and managed in our department of Neurosurgery.

Case Report

A 15-year-old right handed girl was hospitalized with a complaint of headache associated with vomiting and dizziness on/off for last six months. A physical and neurological examination was unremarkable. A computed tomography scan revealed ill-defined heterogeneous solid midline posterior fossa with obstructive hydrocephalus. MRI Brain with Contrast showed intensely enhancing lesion at midline posterior fossa which is located in the inferior cerebellar vermis with indistinct margin with the cerebellum. The patient underwent posterior midline suboccipital craniectomy with en bloc excision of tumor under general anesthesia. Tumor was supplied by the
branch from PICA, vertebral arteries and AICA and tumor was attached to dorsal surface of cervicomedullary area and 4th ventricle extending up to aqueduct. Macroscopically the resected tumor was about 6X7X6cm3 extraaxial cherry like reddish, vascular mass, non-capsular with cystic component. Histologically show two main components comprising of abundant numerous thin walled capillary vessels and stromal cells. However, atypical mitotic figures and necrosis are not seen. Morphological features are consistent with hemangioblastoma (WHO grade I). She was weaned off from ventilator on 4th postoperative day, she underwent nasogastric feeding for one week to prevent aspiration, rest of event was uneventful. At the time of discharge her GOS was five. On follow up CT scan was done showed resolving cavity hematoma.

Discussion

In 1928, Cushing and Bailey introduced the term hemangioblastoma that refers to a benign vascular neoplasm that arises almost exclusively in the central nervous system. According to the World Health Organization classification of tumors of the nervous system, hemangioblastomas are classified as meningeal tumors of uncertain origin. Hemangioblastomas are rare tumors that occur in blood vessels of the brain and spinal cord. They may appear anywhere in the brain but are most often found in the cerebellum and the brainstem. They can progress from solid to cystic. They occur either as a familial autosomal dominantly inherited Von Hippel–Lindau (VHL) disease in about 5–30% of cases, but also may manifest as a sporadic tumor.

Sporadic tumors appear in the 5th and 6th decades of life, whereas VHL-associated tumors manifest earlier, in the 3rd or 4th decades.

Histologically and radiologically, cerebellar HBs are traditionally described as four types. Type 1 (5% of posterior fossa HBs) is a simple cyst without a macroscopic nodule. Type 2 is a cyst with a mural nodule (60%). Type 3, or solid tumors (26%), and Type 4, or solid tumors with small internal cysts (9%), are also seen in the cerebellum and predominate in the spinal cord. They are histologically benign lesions that comprise 1–2% of primary nervous system tumors and 8–12% of all posterior fossa lesions in adults.

These are, however, only unusually encountered in the pediatric population. Hemangioblastomas begin as solid nodules and subsequently enlarging cysts, which cause pressure symptoms. Cysts increase more rapidly than the solid components. Tumors have multiple periods of growth separated by period of arrested growth, but spontaneous regression is not observed.

The majority of patients are asymptomatic at the time of being discovered incidentally by routine CT scan. But large lesions may cause compression of adjacent structures and patients may present some symptom like headache, nausea and vomiting.

The brain CT scan is preliminary choice of investigation, which can clearly identify the location and size of the lesion. It shows all the clinically significant features of hemangioblastoma, along with secondary features such as hydrocephalus and edema.

MRI with gadolinium enhancement is the best choice for diagnosis of hemangioblastoma, with the highest sensitivity and specificity compared with CT and nonenhanced MRI.

A cyst with a small mural nodule is the most common presentation. Cystic fluid surrounding the nodule is hyperintense on T1-weighted images and hyperintense on T2-weighted images.

Angiography might be helpful sometime to identify feeders.

Hemangioblastomas are benign tumors and usually not invasive in nature, so total surgical excision is the best treatment option.

They are benign tumors which presents as a part of VHL disease or as a sporadic lesions affecting the central nervous system.
nervous system. Cerebellar hemangioblastomas and surrounding tissues in the tight posterior fossa have extremely abundant blood supply and are prone to excessive intraoperative hemorrhage, which can hinder the complete resection of the tumor and increase the risk of postoperative morbidity, mortality, and recurrence. Some authors have reported spontaneous hemorrhage in some cases. Massive bleeding with major blood loss can occur during attempts at surgical resection, which resulted in aborting the procedure in many cases. Fukuda et al. reported that two cases of their series were partially resected due to blood loss and cerebellar swelling. They also mentioned that solid lesions with preoperative endovascular embolization were completely resected. In addition, Eskridge et al. reported that two cases of their series had undergone recent attempt at surgical resection at another institution. Liu et al. in their series reported that massive intraoperative bleeding prevented complete resection in eight cases of the control group and that blood loss reached 3240mL in some cases.

These lesions are surgically challenging cases due to their high vascularity and vicinity to neural and vascular structures in the tight posterior fossa, thus making total and safe surgical removal difficult. Dealing with these lesions basically requires different thinking. The surgeon has to circulate around the lesion to devascularize it 360° before its removal.

Such a technique may not be easy within the tight posterior fossa. Preoperative embolization may be helpful to excise tumour with less amount of blood loss.

Other therapeutic modalities include endovascular embolization and stereotactic radiosurgery. Antiangiogenic treatment of hemangioblastoma has also been recently described.

Conclusion

Hemangioblastoma is a purely benign lesion and complete cure is possible after complete tumour excision, however technical difficulties may arise during surgery due to high vascularity of lesion.

Acknowledgement

We would like to thank the pathology department of for cooperation during the histological evidence of disease

References

sporadic cerebellar hemangioblastomas. World Neu-
rosurg 82:815-821, 2014
5. Glasker S, Bender BU, Apel TW, et al. The impact
of genetic molecular analysis of the VHL gene in pa-
tients with hemangioblastomas of the central nervous
system. J NeurolNeurosurg Psychiatry 67:758-
762, 1999
6. Gonzales MF. Classification and pathogenesis of
brain tumors. Kaye AH, Laws ER, eds. Brain Tu-
1995, pp 31-45
7. Hussein MR: Central nervous system capillary haem-
gioblastoma: the pathologist’s viewpoint. Int J
ExpPathol 88:311-324, 2007
von Hippel-Lindau disease tumor suppressor gene.
Science 260:1317-1320, 1993
of preoperative embolization for cerebellar heman-
gioblastoma. Asian Pac J Cancer Prev 14:5179-
5183, 2013
dau disease: a genetic study. J Med Genet 28:443-
447, 1991
11. Maher ER, Yates JRW, Ferguson-Smith MA: Statis-
tical analysis of the two stage mutation model in von
Hippel-Lindau disease, and in sporadic cerebellar
hemangioblastoma and renal cell carcinoma. J Med
12. Niemela M, Lim YJ, Soderman M, Jaaskelainen J,
Lindquist C. Gamma knife radiosurgery in 11 he-
13. Rachinger J, Buslei R, Prell J, et al. Solid haemang-
gioblastomas of the CNS: a review of 17 consecutive
14. Rubinstein LJ. Atlas of Tumor Pathology: Tumors of
the Central Nervous System. Washington, DC: US
Government Printing Office; 1972, pp 235
15. Schuch G, de Wit M, Holtje J, et al. Hemangioblas-
tomas: diagnosis of von Hippel-Lindau disease and
antiangiogenic treatment with SU5416. J ClinOncol
23:3624-3626, 2005
16. Slater A, Moore NR and Huson SM. The natural his-
tory of cerebellar hemangioblastomas in von Hippel–
Lindaudisease. Am J Neuroradiol 24:1570-1574,
2003
ment of hemangioblastomas with presurgical endo-
vascular embolization. Neurol Med Chir 41:246-
251, 2001
treatment of hemangioblastomas with presurgical en-
dovascular embolization. Neurol Med Chir 41:246-
251, 2001