Case Report

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A Giant Solitary Solid Hemangioblastoma: A Case Report and Review of The Literature

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We present a case of solitary vascular tumor on posterior cranial fossa in a 15-year-old female who had a complaint of headache on/off for six months with associateddizziness,nausea, vomiting and vertigo. Well circumscribed solid vascular mass was localized on posterior fossa by computed tomography along with magnetic resonance imaging scan. Furthermore, the diagnosis was confirmed by surgical findings and histological examinations.

Key Words: posterior fossa, solid hemangioblastoma, total excision, tumor

Solidvascular 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.[6]

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 tumorand association with VHL disease. Nevertheless, the confirmed diagnosis still depends on histological examinations. Thiscase ofhemangioblastoma 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-definedheterogenous solid midline posterior fossa with obstructive hydrocephalous. 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 suboccpitalcraniectomy with enbloc excision of tumor under general anesthesia. Tumor was supplied by the

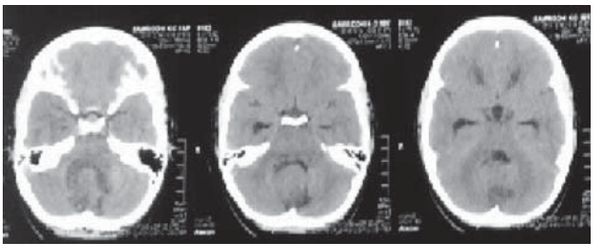

Figure 1 : Contrast-enhanced CT scan revealedill-defined heterogenous solid-cystic in midline posterior fossa. The mass is loacated in the inferior cerebellar vermiswithindistinct margin with the cerebellum.

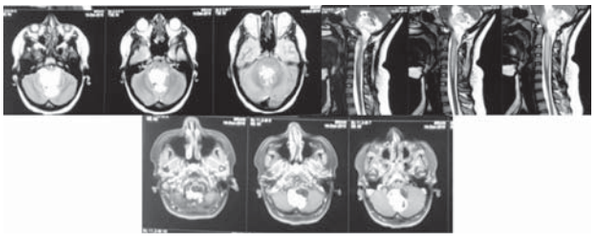

Figure 2 : Contrast –Enhanced MRI Brain revealed Intensely enhancing lesion noted at midline posterior fossa arising from cerebellar vermis. There is intensely enhancing solid component appearing hypo to isointense on T1 and hyperintense on T2 and few cystic areas within the lesion.

branch from PICA, vertebral arteries and AICAandtumor was attached to dorsal surface of cervicomedullary area and 4th ventricle extending uptoaqueduct.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[7] it 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.[1,14]

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.Theycan 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 oflife, whereas VHL-associated tumors manifest earlier, in the 3rd or 4th decades.

Histologically[17] and radiologically,[16] 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 inadults.

These are, however, only unusually encountered in the pediatric population. Hemangioblastomas begin as solidnodules 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 sametime 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

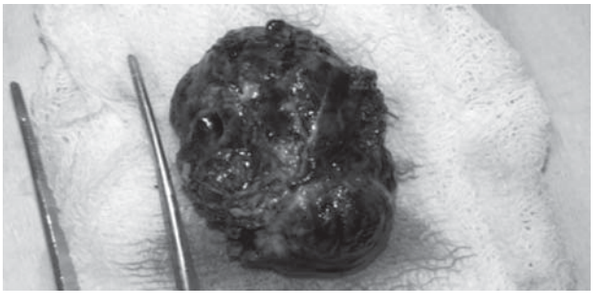

Figure 3 : The pathological specimen 6X7X6cm3in size appeared encapsulated with extensive vessels beneath.

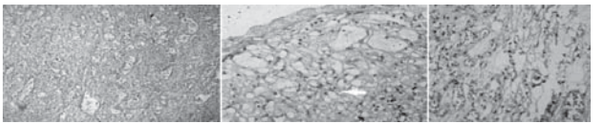

Figure 4 : Histological.:- Single piece of greyish black tissue measuring 4.5X3X2cm.On cut section black homogenous areas are seen AEB-17. Lesion show two main components comprising of abundant numerous thin walled capillary vessels and stromal cells. The stromal cells have abundant clear cytoplasm. The nuclei are of variable sizes being round to oval in shape and hyperchromatic.Areas of hemorrhage, congested blood vessels are also seen. However, atypical mitotic figures and necrosis are not seen. Morphological features are consistent with hemangioblastoma(WHO gradeI).

nervous system.[13,16]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.[2,9] Massive bleeding with major blood loss can occur during attempts at surgical resection, which resulted in aborting the procedure in many cases.[1,3,5,13] Fukuda et al.reported that two cases of their series were partially resected due to blood loss and cerebellar swelling.[4]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 sat surgical resection at another institution.[3] 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.[9]

These lesions are surgically challenging cases due to their high vascularity and their vicinity to neural andvascular structures in the tight posterior fossa, thusmaking total and safe surgical removal difficult.Dealing with these lesions

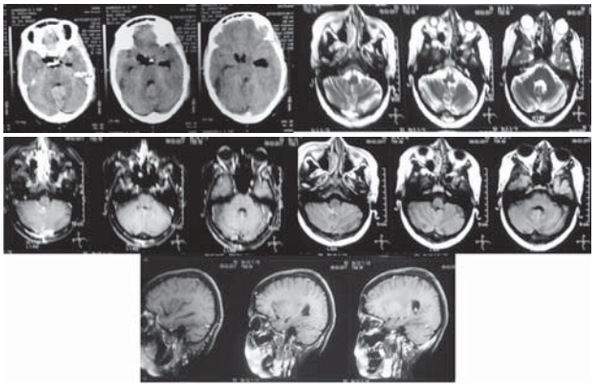

Figure 5 : Post-Operative:-CT and MRI reports showed no residual tumour and resolving hematoma.

basically requires differentthinking. The surgeon has to circulate around thelesion 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.[18]

Other therapeutic modalities include endovascular embolization and stereotactic radiosurgery. Antiangiogenic treatment of hemangioblastoma has also been recently described.[12,15]

Conclusion

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

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