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CASE REPORT

Cerebellar Hemangioma: Advanced Imaging

Phukan Pranjal[1], Handique Akash[2], Kakati Arindom[3], Khonglarh Yookarin[4], Sarma Kalyan[5]

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ABSTRACT

The hemangioblastoma is a rare benign (WHO grade I) vascular tumour. It may difficult to differentiate from other posterior fossa tumour. The conventional MRI showed cystic lesion with markedly enhancing mural nodule. There is paucity of literature of advanced MRI imaging of cerebellar hemangioblastoma. We report a case of cerebellar hemangioma with advanced MRI imaging and reviewing the literature

***Keywords:** Cerebellar hemangioblastoma, Von Hippel-Lindau disease, Cyst with mural nodule*

Address for correspondence and reprint:
[1]Associate Professor
Email: pphukan10@gmail.com
Mobile: 9856928350.
[2]Associate Professor (Corresponding Author)
Email: drahandique@gmail.com
Mobile: 9436114332.
[3]Assistant Professor
Email: arindom.kakati@gmail.com
Mobile: 8014485569.
[4]Associate Professor.
Email: yookarink@gmail.com
Mobile: 9436336490.
[5]Resident, Department of Radiology and Imaging, NEIGRIHMS, Shillong, India- 793018

INTRODUCTION

The hemangioblastoma is a rare benign (WHO grade I) vascular tumour. It may be difficult to differentiate from other posterior fossa tumours. The conventional MRI showed cystic lesion with markedly enhancing mural nodule. There is paucity of literature of advanced MRI imaging of cerebellar hemangioblastoma. We report a case of cerebellar hemangioma with advanced MRI imaging and reviewing the literature.

CASE HISTORY

A 20 years old male presented with progressive holocranial headache for 2 months associated with vomiting and gait disturbances for 2 weeks. The boy was in severe distress due to headache. Fundal examination showed papilledema. Motor and sensory examinations were normal. Evaluation of the cerebellar system revealed truncal ataxia.

MRI Findings:

On MR, a well defined rounded, sharply defined cystic lesion was arising from the right cerebellar tonsil that showed prolongation of both T1 and T2 relaxation times with multiple flow voids. A solid nodule was seen within the wall of the cyst which showed strong enhancement on contrast study, whereas the cyst wall does not. The nodule abuts the cerebellar surface. On MR perfusion, the mural nodule showed increased rCBV (not shown). Mean perfusion curve showed rapid fall followed by rapid upstroke not reaching the base line. On MR spectroscopy, there was decreased NAA with lipid peak.

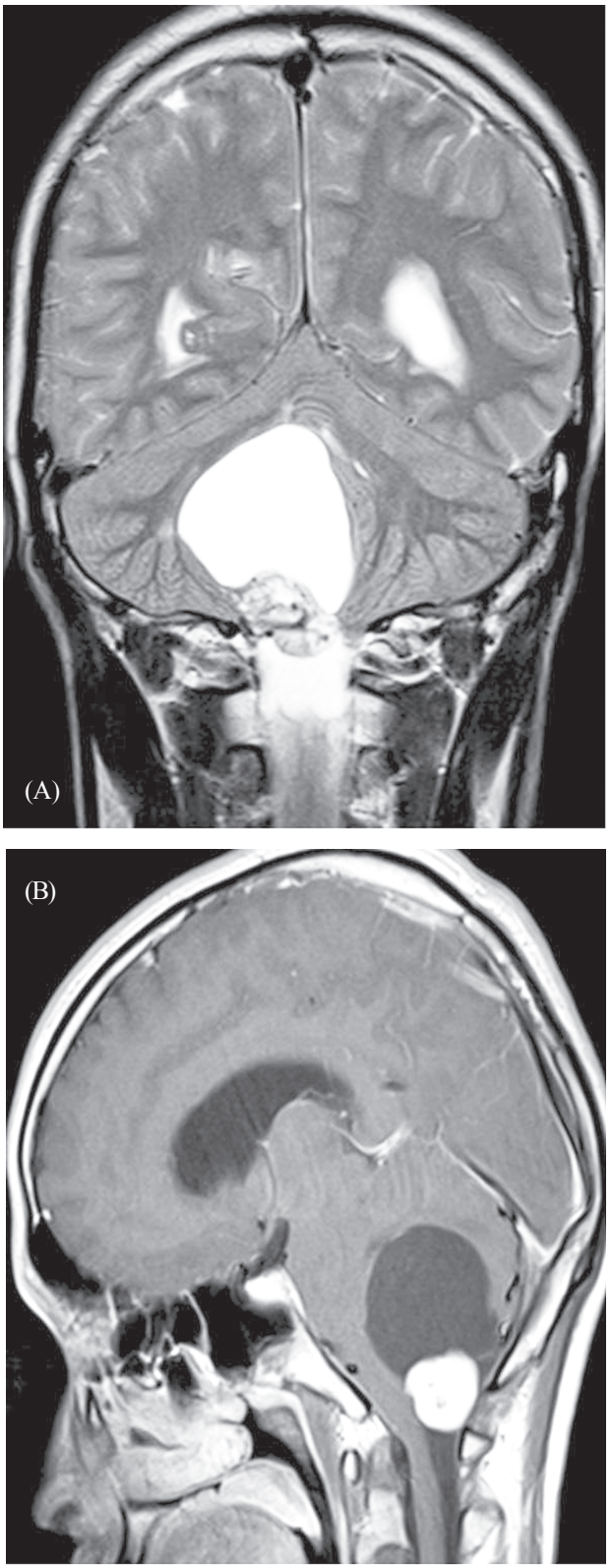

Figure 1 On MRT2WI, coronal images and contrast enhance images (A and B) showed a well-defined, sharply marginated cystic lesion arising from the right cerebellar tonsil with multiple flow voids. A strongly enhancing nodule was seen within the wall of the cyst abuts the cerebellar surface. (C) Mean perfusion curve showed rapid fall followed by rapid upstroke not reaching the base line. (D) On MRS, there was decreased NAA with lipid peak.

Cerebral Angiography

Digital angiography showed tangles of tightly packed vessels opacified in the early arterial phase with feeding artery from right AICA with draining vein through the superficial cerebellar vein to transverse sinus.

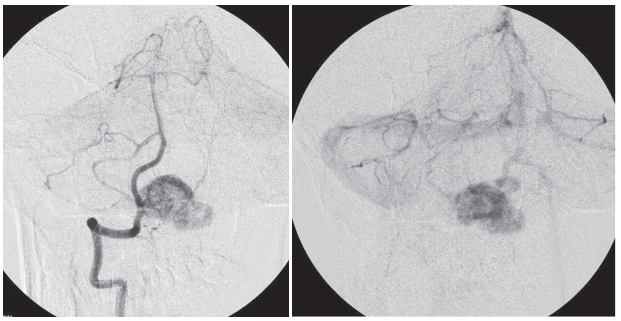

Figure 2 Digital angiography (A and B) showed tangles of tightly packed vessels opacified in the early arterial phase with feeding artery from right AICA with draining vein through superficial cerebellar vein to transverse sinus

Surgery

The patient underwent a sub occipital craniotomy and excision of the lesion. Per operatively there was a cystic mass involving the right cerebellar tonsil, with a mulberry coloured smooth walled nodule, it was highly vascular with a very prominent draining vein.

Histopathology

Histopathological section showed stromal vacuolated cells with interspersed blood vessels.

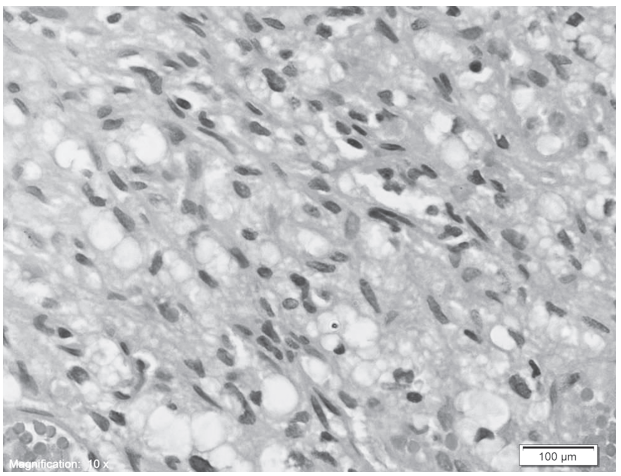

Figure 3 Histopathological section (1E) showing stromal vacuolated cells with interspersed blood vessels (H and E 40x:)

DISCUSSION

The hemangioblastoma is a benign (WHO grade I) vascular tumour. It accounts for 1%–2.5% of all intracranial tumours in the population.[1] Ninety-five percent of hemangioblastomas located in the posterior fossa of which 70 to 80% are located in the cerebellar hemispheres. Hemangioblastomas are linked to von Hippel-Lindau (VHL) disease, although isolated cerebellar hemangioblastoma is the commonest presentation.[2]

Hemangioblastomas are originating from the cerebellar surface. Therefore, they are constantly connected to a pial surface. [3] Macroscopically, the tumour has a bright yellow colour as a result of its lipid content.[1,4] The histological features are consisting components of capillary network lined by hyperplastic endothelial cells and the stromal cells, which have pleomorphic or lobulated nuclei and lipid containing abundant, pale cytoplasms.[1,4] Mitoses are usually inapparent.[1, 4]

The expression of highly angiogenic growth factors and their receptors might be responsible for high vascularity of the tumor. [5]

CEMR images are useful to detect additional small hemangioblastomas, which are often not visible by baseline MRI. Because of the significant likelihood of multiple lesions, the whole neuraxis should be imaged. On MRI, cerebellar hemangioblastoma typically appear as large, rounded, sharply marginated cystic lesions that show prolongation of both T1 and T2 relaxation times.[6] Variable intracystic signal intensity may occur, depending on protein or hematic content. A solid nodule may be seen within the wall of the cyst.[6] After gadolinium administration, the mural nodule enhances strongly, whereas the cyst wall most commonly does not enhance unless lined by neoplasm.[7] The nodule consistently abuts the cerebellar surface and is usually smaller than the cyst unlike cystic astrocytoma, which tends to have a larger nodule. A diagnostic pitfall is the hemangioblastoma with a small central lucency, which can be interpreted as a necrotic metastasis. In this case, the ring like enhancement of the necrotic nodule is thick and irregular.[8]

Large feeding and draining vessels in the periphery and within the solid component appear as tubular flow voids on T2-weighted images and MRA. However, digital angiography better shows tangles of tightly packed, wide

vessels, opacified in the early arterial phase.[6] Sometimes, the typical finding of a "cherry attached to its stalk" may be recognized. Angiography is needed to identify the vascular pedicle before surgery, and preoperative embolization may be useful in order to reduce the risk of intraoperative bleeding.[9,10] Angiography also is useful for differentiating between pilocytic astrocytoma and hemangioblastoma, because only the latter shows a typical blush**.**

MR Spectroscopy

Proton MRS reveals a high mobile lipids (Lip) peak between 0.9 and 1.4 ppm, which was compatible with histologically proven lipids in the tumor. The creatine/phosphocreatine peak is low. Choline-containing compounds may increase. The N-acetylaspartate peak is absent, which indicate nonneurogenic origin of the tumor. An oxaloacetate at 2.37 ppm is a characteristic feature of hemangioblastoma. These unique results of proton MRS can play an important role in the differential diagnosis of intracranial hemangioblastoma.[11, 12] In our case we did not find oxalate peak. From our point of view, the signal of 2.37 ppm in their reported case might have been due to a noise level, not oxaloacetate. The high lipid peak is also seen in other tumors, especially high-grade tumors such as high-grade gliomas, metastatic brain tumors, and anaplastic meningiomas due to presence of intratumoural necrosis. The high lipid peak on proton MRS without the necrotic component on MRI can be a characteristic finding of hemangioblastoma.

MR Perfusion

The rCBV map is also useful in differentiating cystic astrocytoma from hemangioblastoma. At conventional MR imaging, both cerebellar hemangioblastomas and astrocytomas often appear as small, enhancing nodules within a well-circumscribed, thin-walled cyst, as in our cases. Despite some differential features such as an intratumoral signal void, differentiation by conventional MR imaging alone is difficult, especially where tumors are smaller than 1cm.[13] Quantitative analysis indicated that the rCBV ratio of hemangioblastomas was significantly higher than that of cerebellar astrocytomas, and it was also found that compared with that of gray matter, the signal intensity of hemangioblastomas was much higher, while that of cystic astrocytomas was slightly higher.[14] These tumours show arterio venous shunting on angiography [15] and are usually lesions with high rCBV. They can show a rapid steep fall in signal intensity with rapid return to baseline; and a rapid steep fall in signal intensity with rapid return to baseline, followed immediately by a second, smaller dip. It has also been shown that, depending on rCBV values, hemangioblastoma and pilocytic astrocytoma can be differentiated with confidence.[16] In our case there is rapid fall of signal intensity with rapid return towards baseline but not reaching the base line. The 1st halves of the curve represent high leakiness of the blood brain barrier and 2nd half of the curve represent high arterio venous shunting of the tumor bed.

CONCLUSION

Advanced MR images are useful in differentiating hemangioblastoma from other lesion and detect additional small hemangioblastomas. The high lipid peak without NAA or Lactate peaks on proton MRS and the absence of a necrotic component on MRI may be the characteristic radiological findings of hemangioblastoma. These unique results of proton MRS can play an important role in the differential diagnosis of intracranial hemangioblastoma. Hemangioblastoma demonstrate relatively predictable patterns of mean perfusion curve on T2* DSC PMRI. Along with perfusion maps and conventional cross-sectional imaging, can help in the characterization of intracranial tumors.

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