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Case report on cerebellar hemangioblastoma showing post contrast cyst wall enhancement on magnetic resonance imaging

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ABSTRACT

Background: Cerebellar hemangioblastoma is a WHO grade one tumor of vascular origin located in posterior cerebral fossa and is the most common primary intra-axial and infratentorial tumor in adults which usually contains a highly vascular mural nodule that shows avid post contrast enhancement on MRI (Magnetic Resonance Imaging). We present a case involving a 41-year-old adult male who came to our hospital with complaints of giddiness. **Objective:** To differentiate adult posterior fossa tumors from cerebellar hemangioblastoma on MRI. To establish histopathological correlation with imaging finding on MRI. **Material and Methods:** Magnetic Resonance Imaging (MRI) is the modality of choice for diagnosing cerebellar hemangioblastoma. We used Three Tesla (3T) MRI machine of GE healthcare system for our case study. A microscopic evaluation with haematoxylin and eosin staining sections in low and high magnification was done on histopathology. Subject of our case study is a 41-year-old Adult male patient presented to our institution. Patient was administered Gadolinium contrast agent and was subjected to the MRI scan. **Results and conclusions:** We diagnosed the patient with Cerebellar hemangioblastoma on MRI and also established a firm Histopathology correlation to strengthen our diagnosis. Thus, excluding other differentials of adult posterior fossa tumors of the brain. Also, post contrast MRI study revealed cyst wall enhancement in our case which is an uncommon finding in cerebellar hemangioblastoma.

Keywords: Cerebellar hemangioblastoma, Hemangioblastoma, Mural nodule, posterior fossa tumors, Pilocytic Astrocytoma, Ganglioglioma, Brain metastases.

1. INTRODUCTION

Hemangioblastoma is a WHO grade one tumour of vascular origin occurring in the central nervous system (CNS) (Slater et al., 2003). It has also been described in other parts of the body, such as liver, kidney and pancreas. This

tumour can present in isolation or as part of a syndromic association with other lesions such as in Von Hippel Lindau disease (Nadgir & Yousem, 2021). Cerebellar hemangioblastoma contains a highly vascular mural nodule that enhance readily post contrast administration (Osborn, 2021). It is mostly cystic lesion associated with solid enhancing mural nodule with prominent flow voids (Ordookhanian et al., 2017). Hemangioblastoma has an occurrence in young to middle aged adults (slight male predilection) (Abd Hamid et al., 2000). We present a case involving a 41-year-old adult male who came to our hospital with complaints of giddiness.

2. CASE HISTORY

40-year-old adult male patient presented with giddiness, swaying while walking and feeling of imbalance while getting up from bed since ten days. Patient also had complaints of diffuse, low grade, continuous worsening headache and difficulty concentrating since three days. Loss of appetite and nausea and one episode of vomiting since two days. Patient did not have hemiparesis, paraesthesia or visual disturbances. Patient was well oriented with surroundings. No signs of tremors, incoordination, dysmetria and dysdiadochokinesia. Speech, memory and extra ocular movements were normal. Power of bilateral upper and lower limbs were five by five (5/5). Pin prick test, kneel heel shin test and toe tapping tests were normal. However, Broad based ataxic gait was seen.

Pre contrast MRI sequences of Axial non-contrast T1-weighted image (Figure 1) shows a well-defined hypo intense cystic lesion (blue arrow) arising from the left cerebellum. Lesion is seen to compress the vermis and the medial aspect of right cerebellum (yellow arrow). Effacement of fourth ventricle is seen and lesion is seen to extend into the cisterna magna. Axial non-contrast T2-weighted image (Figure 2) shows a well-defined hyper intense cystic lesion (blue arrow) arising from the left cerebellum with mild surrounding edema (white arrows). Few flow voids (white arrowheads) representing feeding and draining vessels to the mural nodule is seen. Axial non-contrast FLAIR images (Figure 3) of the brain shows hydrocephalus with dilatation of bilateral temporal horns (yellow arrowheads). Axial non-contrast T2 Fluid Attenuated Inversion Recovery (FLAIR) sequence image (Figure 4) of the brain demonstrate trans ependymal flow of Cerebrospinal fluid (blue arrowheads).

After contrast administration, Axial T1- weighted pre and post contrast images (Figure 5) of the brain shows a cystic lesion (blue arrow) arising from the left cerebellar hemisphere with a posteriorly located enhancing mural nodule (yellow circle) measuring $\sim 1.4 \times 0.6$ cm in size. The mural nodule is seen to be in contact with the adjacent pia. No evidence of adjacent meningeal enhancement is seen. Mild cyst wall enhancement is also seen post contrast administration (white arrows). On DWI and ADC map of the brain shows lower DWI signal (yellow arrowhead) and higher ADC signal (blue arrowhead) within the mural nodule compared to the rest of the cerebellum (Figure 6). Sagittal T1 post contrast (Figure 7) with maximum intensity projection (MIP) image shows a feeding artery from a branch of the left posterior inferior cerebellar artery (yellow arrow) and a draining vein from the left transverse sinus (blue arrow) into the mural nodule. Histopathological imaging features demonstrate shows vacuolated tumour cells with increased number of blood vessels and foamy cytoplasm (Figures 8, 9).

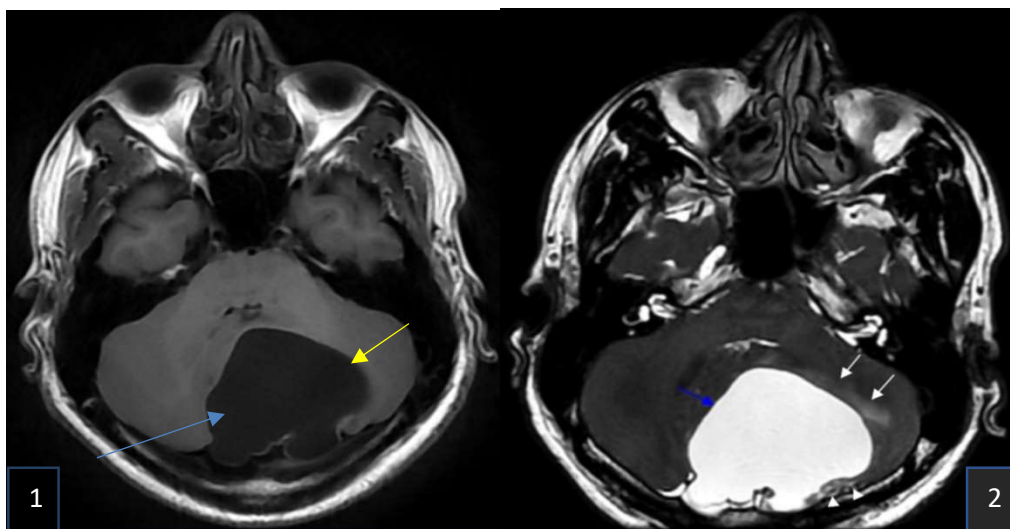


Figure 1 Axial non-contrast T1-weighted image

Figure 2 Axial non-contrast T2-weighted image

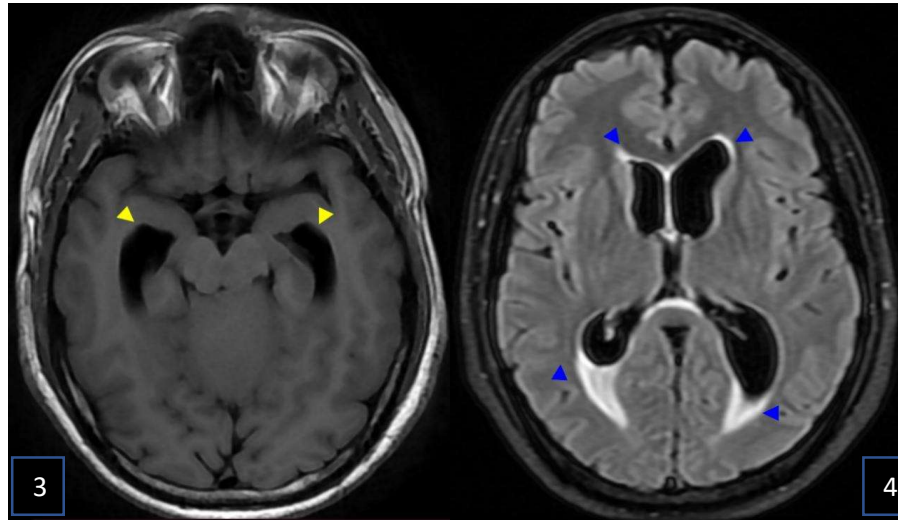


Figure 3 Axial non-contrast FLAIR images

Figure 4 Axial non-contrast T2 (FLAIR) sequence

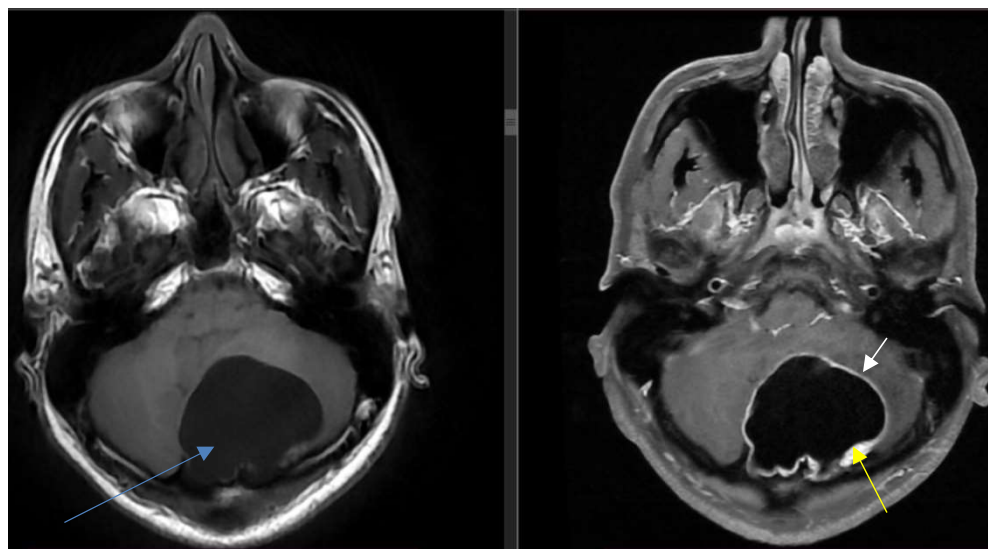


Figure 5 Axial T1- weighted pre and post contrast images of the brain

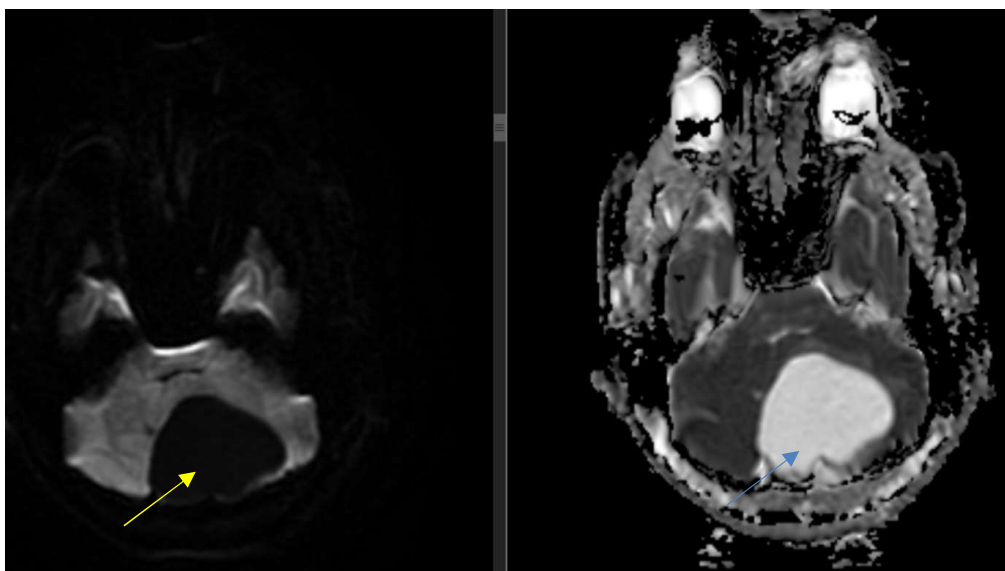


Figure 6 Axial diffusion weighted imaging (DWI) and ADC map of the brain

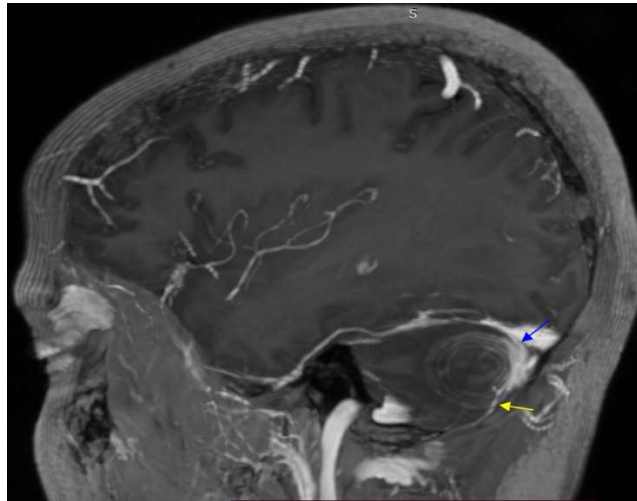


Figure 7 Sagittal T1 post contrast with maximum intensity projection (MIP) image

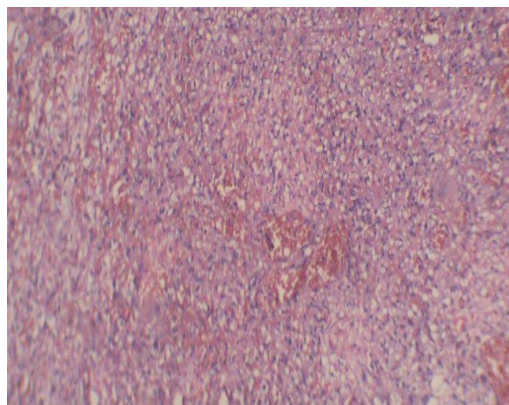


Figure 8 Low magnification image on microscopic evaluation (x40) shows vacuolated tumor cells with increased number of blood vessels and foamy cytoplasm.

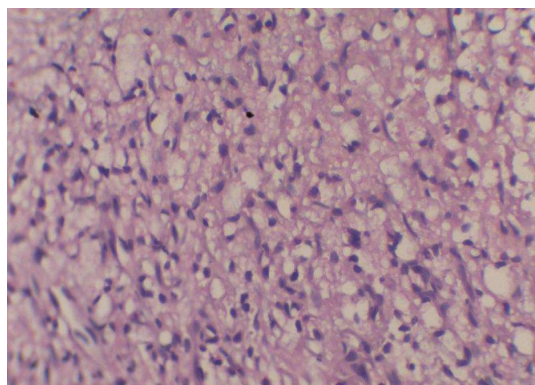


Figure 9 High magnification image on microscopic evaluation (x200) shows vacuolated tumor cells with increased number of blood vessels and foamy cytoplasm.

3. DISCUSSION

Hemangioblastoma is a WHO grade one tumor of vascular origin located in central nervous system (CNS). It has also been described in other parts of the body, such as liver, kidney and pancreas (Grant & Griffin, 2019). This tumor can present in isolation or as part of a syndromic association with other lesions such as in Von Hippel Lindau disease (Kuharic et al., 2018).

Cerebellar hemangioblastoma contains a highly vascular mural nodule that enhances readily post contrast administration. It is mostly cystic lesion associated with solid enhancing mural nodule with prominent flow voids. Hemangioblastoma has an occurrence in young to middle aged adults (slight male predilection) with estimated incidence of one percent to 2.5% of all intracranial tumors. Symptoms of cerebellar hemangioblastoma include headache, hydrocephalus, vertigo, ataxia, nystagmus,

altered mental state, nausea and vomiting (Bing et al., 2009). Cerebellar hemangioblastoma's are routinely reported as four types on the basis of their histology and radiological appearances (De san Pedro et al., 2010). Type 1 is a simple cyst without a macroscopic nodule, affecting 5% of posterior fossa. Type 2 is a cyst with a mural nodule (60%). Type 3, or solid tumors (26%). Type 4, or solid tumors with small internal cysts (9%), both of which are more common in the spinal cord than in the cerebellum. Type 1 is a rare entity as stated by some authors (Louis et al., 2021).

MRI is the modality of choice for diagnosing Cerebellar hemangioblastoma. We used 3 Tesla MRI of GE healthcare systems for our case study. T1, T1 post contrast and T2/FLAIR sequences of MRI showed a well-defined, cystic lesion noted arising from the left cerebellum with mild surrounding edema. Lesion was seen to compress the vermis and the medial aspect of right cerebellum. Effacement of fourth ventricle was seen with dilatation of third and bilateral lateral ventricles and mild periventricular seepage. Axial non-contrast FLAIR images of the brain demonstrate tran's ependymal flow of Cerebrospinal fluid. Lesion was seen to extend into the cisterna magna. Minimal cyst wall enhancement was seen post gadolinium contrast administration. However, focal nodular enhancement was seen in the left lateral aspect of the cyst corresponding to mural nodule of size~ 1.4 x 0.6 cm and with few flow voids were also seen surrounding it representing feeding and draining vessels. The mural nodule was seen to be in contact with the adjacent pia. No evidence of adjacent meningeal enhancement was seen. Axial diffusion weighted imaging DWI and ADC map of the brain shows lower DWI signal and higher ADC signal within the mural nodule compared to the rest of the cerebellum.

One additional sequence of highly maximum intensity projection (MIP) image Post contrast T1 was also included which showed feeding artery from the branch of left posterior inferior cerebellar artery and draining into left transverse sinus (Raz et al., 2012). Usually, patients diagnosed with cerebellar hemangioblastomas are also associated with von-Hippel Lindau disease. Screening ultrasonography of whole abdomen was done in our patient but it did not reveal any significant features or abnormalities associated with von Hippel-Lindau disease (Utsuki et al., 2011).

Differentiating points of Cerebellar hemangioblastoma from other posterior fossa tumors on MRI are as follows

- Juvenile pilocytic astrocytoma- As the name suggests occurs in patients under 20 years of age and involves an enhancing cyst wall with a hypo vascular nodule. Whereas, our patient is an adult and the mural nodule is enhancing post contrast administration revealing its highly vascular nature.
- Adult Pilocytic Astrocytoma- It is a very rare entity. The cases described have enhancing wall and mural nodule. But the mural nodule is in contact with the Pia in our case and is more in favour of hemangioblastoma.
- Ganglioglioma- Common in < 30 years of age group and it is a solid lesion maybe with a cystic component and calcifications, commonly located in temporal region.
- Brain Metastasis- Cystic component with enhancing mural nodule in brain metastasis is a quite rare presentation. Also, the patient's age, location of tumour, no evidence of calcification and no findings on whole abdomen ultrasonography screening excludes metastasis as our diagnosis.

Surgical resection of hemangioblastoma is the preferred treatment for the patient diagnosed with Cerebellar Hemangioblastoma. Our patient underwent surgical resection and tissue samples were sent for histopathology examination. On Histopathology examination by microscopic evaluation with haematoxylin and eosin staining sections in low and high magnification showed vacuolated tumor cells with increased number of blood vessels and foamy cytoplasm.

4. CONCLUSION

We diagnosed the patient with Cerebellar hemangioblastoma on MRI with contrast study and also established a firm Histopathology correlation to strengthen our diagnosis. Thus, excluding other differentials of adult posterior fossa tumors of the brain. Also, post contrast MRI study revealed cyst wall enhancement in our case which is an uncommon finding in cases of cerebellar hemangioblastoma.

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Authors' contribution

Nasin usman: Conceptualization, Supervision, Methodology, Resources, Data Collection, Writing and Formal analysis
 G Jayasri: Writing, Investigation, Resources, Analysis, Draft preparation, Review and Editing
 Lekha priya M: Writing, Investigation, Analysis, Review and Editing
 All authors have read and agreed to submit the manuscript.

Informed consent

The proper written consent obtained from the patient.

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Conflict of interest

The authors declare that there is no conflict of interests.

Data and materials availability

All data sets collected during this study are available upon reasonable request from the corresponding author.

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