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Case Report

Cerebellar Hemangioblastoma: A cyst with a mural nodule – Case report with review of literature.

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Abstract

Hemangioblastomas are slow-growing vascular tumors of central nervous system. It can occur sporadically or can be seen as a part of Von Hippel Lindau (VHL) syndrome. It is usually seen in cerebellum, but might be found in brainstem or spinal cord as well. These are uncommon tumors, comprising only around 1-2.5% of intra-cranial tumors. It also comprises of around 10% of all the posterior fossa tumors. Here we discuss an instance of a 33 year old male presenting with complains of giddiness and headache for last 2 months. No significant past/medical/familial history. Laboratory investigations were unremarkable. Patient was then adviced to undergo MRI-Brain. Contrast enhanced MRI-Brain revealed a well-defined cystic lesion with vividly enhancing eccentric solid mural nodule in right cerebellar-hemisphere, suggestive of cerebellar hemangioblastoma. Surgical excision was done and histopathological evaluation confirmed the diagnosis. Therefore features of cerebellar hemangioblastoma and other differentials for cyst with mural nodule are discussed here.

Keywords: Hemangioblastoma, Cerebellum, Magnetic resonance imaging.

Introduction

Hemangioblastomas are slow-growing vascular tumors typically occurring in young adults or middle aged individuals. They demonstrate a male predominance with peak incidence between 30–60 years. Even though these are uncommon, comprising of only 1-2.5% of all intracranial tumors; they are also the most frequent primary intra-axial, infra-tentorial tumors. They are grouped under WHO grade 1 tumors. Approximately 85% of them are located in cerebellar hemisphere, followed by 10% of cases in vermis. These can be noted airing in spinal cord, brain stem as well.

Hemangioblastomas can either occur sporadically or can be a part of Von Hippel-Lindau (VHL) syndrome. Around 75-80% of them occur sporadically and the rest 20-25% are associated with VHL.[1] Solitary tumors are usually sporadic, but multiple lesions are generally with VHL. Sporadic tumors appear later by 5th to 6th decade of life, the tumors that are VHL-associated manifest earlier by 3rd or 4th decade. VHL is an inherited, autosomal-dominant condition which presents with multiple benign and malignant tumors, including hemangioblastomas in nervous system, retinal angiomas, endolymphatic sac tumors, renal cell carcinoma (RCC), pheochromocytomas of kidney, cysts of pancreas and neuroendocrine tumors.[1]

Here, we discuss a case of cerebellar hemangioblastoma in a 33 year old male with complaints of headache and giddiness and also discuss other significant differentials for cyst with a mural nodule in brain.

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Case History

A 33 year male patient came to outpatient department with complaints of giddiness & headache since 2 months. He gives a history of head injury following fall 3 years back. Rest there was no significant past / medical / family history. He was not a known diabetic or Hypertensive. On examination of central nervous system, no obvious neurological

deficits were noted. Laboratory investigations were unremarkable. Patient was then adviced to undergo MRI-Brain to exclude possibilities of any space occupying lesion causing patient symptoms.

Imaging findings

MRI brain (Plain and contrast enhanced)

MRI brain showed a large, well-defined, T2 hyperintense cystic lesion measuring ~ 5.2 x 4.6 x 3.3 cm (TR x AP X CC) showing suppression on FLAIR sequence with an eccentric T1 iso-slightly hyperintense compared to gray matter and T2/ FLAIR heterogeneously hyperintense solid mural nodule and few internal septations in right cerebellar hemisphere (Figure 1). There was no restricted diffusion on DWI/ corresponding ADC sequences (Figure 2A & 2B). Few foci of blooming were noted within the mural nodule on SWI & PHASE sequences (Figure 2C & 2D). On post contrast imaging, the lesion demonstrated avid heterogeneous enhancement of eccentric solid mural nodule and minimal enhancement of septations (Figure 3).

Mass effect was noted in form of compression and mild displacement of cerebellar peduncles and brainstem on right side and compression and effacement of 4th ventricle with resultant mild dilatation of supratentorial ventricles. Midline shift of ~ 1.1 cm was noted to left side (Figure 1). There was also crowding at foramen magnum with tonsillar-herniation of ~ 8 mm below level of McRae's line (Figure 4).

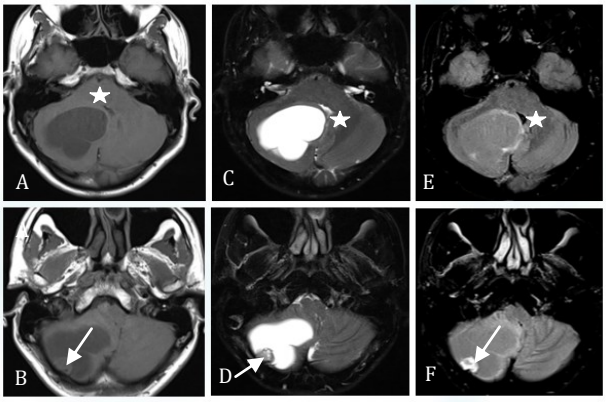

Figure 1: MRI Brain axial sections at the level of cerebellum showing a well-defined cystic lesion with few septations and an eccentric solid mural nodule in the right cerebellar hemisphere.

(A&B): On T1 weighted images cystic component is hypointense and solid mural nodule (white arrow) appears iso-to slightly hyperintense to adjacent grey matter.

(C&D):T2 weighted images showing a hyperintense cystic lesion with a heterogenously hyperintense eccentric solid mural nodule (white arrow).

(E&F): On FLAIR images, there is suppression of signal intensity within the cystic component and solid mural nodule (white arrow) appears same as that on T2-WI.

Also note the mass effect caused by the lesion in the form of compression and mild displacement of cerebellar peduncles & brainstem on right side, compression and effacement of 4th ventricle (star) and a midline shift of ~ 1.1 cm towards left side.

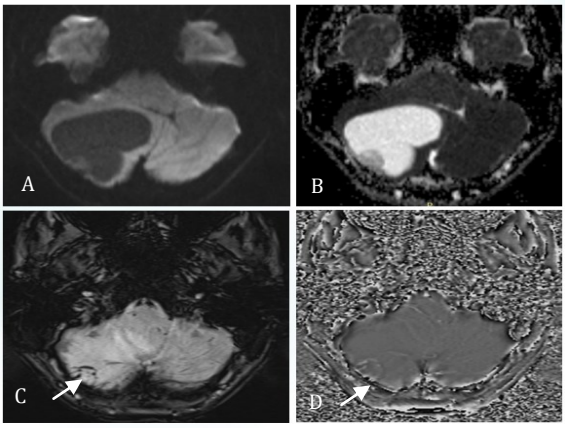

Figure 2: MRI axial sections

(A) DWI & (B) corresponding ADC sequences showing no evidence of restricted diffusion. (C) SWI and (D) corresponding PHASE sequences showing few foci of blooming were within the mural nodule (white arrows).

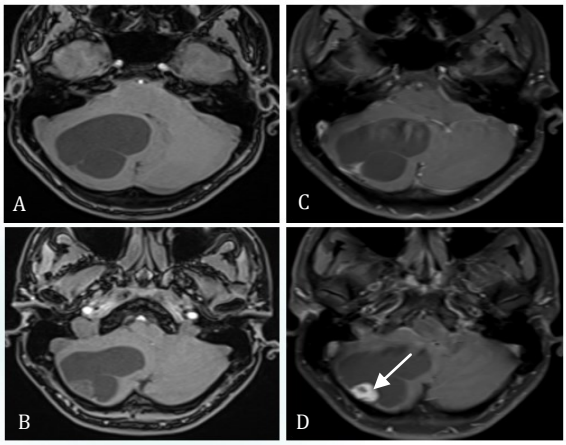

Figure 3: MRI axial sections at the level of cerebellum (A&B) T1 pre-contrast VIBE sequence and (C&D) T1-FS post contrast imaging, there is avid heterogeneous enhancement of the eccentric solid mural nodule (white arrow) and minimal enhancement of septations.

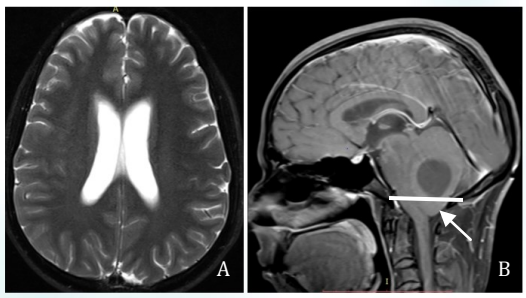

Figure 4: (A) Axial T2 image showing mild dilatation of supratentorial ventricles due to compression of 4th ventricle.
(B) Sagittal T1 FS post contrast image showing mass effect in the form of crowding at foramen magnum with ~ 8 mm tonsillar herniation (white arrow) below the level of McRae's line (white line).

Radiological Impression

The above features of cyst with an eccentric enhancing mural nodule on contrast enhanced MRI brain were suggestive of right cerebellar hemangioblastoma with its mass effect on adjacent structures & ventricles and midline shift towards left side.

Patient was subjected to surgical removal of the tumor and the excised specimen was addressed for histopathological examination.

Histopathological Impression

The impression was given as cerebellar hemangioblastoma which was in correlation with findings on contrast enhanced MRI brain as discussed earlier.

Discussion

Hemangioblastomas are slow-growing, vascular tumors of central nervous system, typically seen occurring in young adults or middle aged men with peak age of incidence between 30-60 years. Here patients usually presents with headache (80%) and uncommonly can also exhibit features of elevated intracranial pressure such as vomiting, drowsiness, altered mental status or with features of cerebellar dysfunction such as ataxia, imbalance, dysarthria or nystagmus.[1] Current patient was also a 33 year male presenting with complaints of headache & giddiness.

They occur in various anatomical locations of central nervous system. Most commonly in cerebellar hemispheres (83%-86% of cases). They can be seen in spinal cord (3-13% cases), medulla (2-3% cases) and cerebrum (~1.5% cases).[3]

There are basically capillary hemangioma which is WHO grade I lesion (low grade). Its a well-circumscribed, predominantly cystic tumor with an eccentric mural nodule. These mural nodules contains multiple high-flow vessels within them which causes their intense enhancement.[1, 4] The cystic components are said to possess no wall of their own, their content is due to exudation from the solid vascular component and the displaced cerebellar parenchyma by the fluid forms its boundaries.[5]

CT findings

On CT, they appear as cystic lesion with the eccentric mural nodule. The mural nodule appears isodense to adjacent neuro-parenchyma and shows intense homogeneous enhancement on contrast administration. The cyst wall doesn't enhance on contrast study. Calcification are usually absent.

MRI findings

On MR imaging, hemangioblastomas commonly appear as large, well-circumscribed, homogeneous cystic masses with a hypervascular small eccentric mural nodule; however can also have a varied appearance.[4] They are categorised into 4 types based on their components and appearance. Type I is a simple cystic lesion without any macroscopically visible solid mural nodule, seen in about 5% of cases. Type II is the most frequent form where lesion appears as a cystic lesions with a strongly enhancing eccentric mural nodule, seen in about 60% of cases.[2] These mural nodules are hypointense on T1 and hyperintense on T2-weighted images, demonstrating intense enhancement after contrast injection.[1,4] Type 3 are solid tumors without any macroscopically visible cystic component (26% of cases) and lastly Type 4 consists of solid tumors with internal small cysts (9%).[2]

Angiography (DSA)

Solid mural nodule demonstrates the feeding arteries and sometimes the dilated draining veins as well with presence of central dense tumor blush.[3]

Differential diagnosis

Main differential diagnosis for cyst with mural nodule includes Pilocytic astrocytoma, Pleomorphic Xanthoastrocytoma, Ganglioglioma or Desmoplastic infantile Gangliogliomas.[6]

Pilocytic astrocytoma is also a well-defined, usually cystic lesion with eccentric mural nodule defined under WHO grade I lesions. Commonest primary brain tumor amoung children with a peak age of presentation between 5-15 years and was previously called as juvenile pilocytic astrocytoma. It has a strong interrelation with NF1 (neurofibromatosis type 1). 60% of them are located in cerebellar hemispheres and other locations include optic nerve/chiasm/tract. Solid components appear T1 iso/hypointense and T2/FLAIR hyperintense to

gray matter. Cystic components appear isointense to mildly hyperintense to CSF (cerebrospinal fluid) on T1 and shows hyperintense signal on T2 with no suppression of signal on FLAIR sequence. Cyst wall may occasionally show enhancement.[6] However they can sometime also appear as a heterogenous or a solid lesion. Calcification can also be seen. Presentation in childhood, presence of calcification and wall enhancement differentiates it from hemangioblastoma.

Pleomorphic xanthoastrocytoma (PXA) belongs to WHO grade II. It is typically a supratentorial cortical mass, most common in temporal lobe. This nodule appears hypointense-isointense to the adjacent gray matter on T1 whereas cystic part appears isointense to the CSF. On T2 images, they appear hyperintense or heterogenous and shows suppression on FLAIR sequence. Presence of adjacent meningitis, dural tail, scalloping of overlying bone and supratentorial location differentiates it from hemangioblastoma.[6]

Gangliogliomas are well-differentiated, WHO I – II grade lesions that are slow-growing. Typically seen as partially cystic, cortically based, heterogeneously enhancing mass in young adolescent person/child with symptoms of temporal lobe epilepsy. Commonest location is temporal lobe where it appears as a well-circumscribed mass expanding the adjacent cortex with cortical dysplasia. Calcifications are common. Younger age of presentation, location of lesion, presence of calcification and heterogenous enhancement differentiates them from hemangioblastoma.[6]

Desmoplastic infantile ganglioglioma (DIG) is also a WHO I grade tumor affecting infants during first 2 years of life. Appears as huge cyst with cortical based enhancing nodule in frontal or parietal lobes. On contrast administration, solid nodule shows marked enhancement with adjacent reactive dural thickening and enhancement of pia.[6]

Other differentials includes medulloblastomas in adults, Tanycytic ependymoma, Intracerebral schwannoma and Papillary glioneuronal tumor (PGNT).

Treatment

Complete surgical resection is curative in these lesions. However a pre-surgical embolization in indicated in larger lesions to make the surgical procedure easy. If the lesion appears as a simple cyst, drainage of the cyst fluid can be done. In cases of insufficient resection and just simple cyst drainage, an adjuvant radiotherapy is indicated. Recurrence rate of these lesions is ~ 25%.[7]

Conclusion

Radiological imaging, specifically MRI plays a very important role in pre-operative diagnosis of hemangioblastoma. It also helps us in differentiating it from other differentials of cyst with mural nodule in brain. Hemangioblastomas appear as well-defined cystic lesions with an eccentric enhancing solid mural nodule within them, most commonly seen in cerebellum, especially in young and middle aged men. No calcifications/ enhancement of cyst wall will be seen in them.

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