Cerebellar Hemangioblastoma: An Unusual Cause Of Syncope

Shuaib Mohamed
Advanced Radiology Clerkship
Sri Ramachandra Medical College & Research Institute, India

Gillian Lieberman, MD
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Overview

- Patient history.
- Patient CT, MRI & CTA discussion.
- Hemangioblastoma literature review.
- Differential diagnosis.
- Imaging modalities.
- Von Hippel-Lindau disease.
- Follow-up recommendations.
- Treatment.
- Summary.
• 55 year old Male with history of syncope followed by a head trauma presents to the ER.
• He is conscious and oriented.
• He denies history of any drug abuse.
• History of Hyperlipidemia and mild L5-S1 disc herniation.
• He is on Simvastatin and Esomeprazole therapy.
• Mother has history of breast cancer.
On Clinical Examination

• Vital signs were unremarkable.
• Pupils, Mental status exam, Motor, Sensory, Co-ordination and Cranial Nerves were tested and were unremarkable.
• Blood and Urine tests were within normal limits.
• A Head CT was taken to evaluate the cause of syncope and extent of trauma.
NonContrast Head CT

- Axial view showing the Cerebellar tonsils appear to extend into the foramen magnum

- No signs of fracture or hemorrhage
History Contd..

• After the CT was analyzed, patient was sent home with no further intervention or follow up.
• Patient was apparently asymptomatic for Eight months.
• Following which, he developed multiple syncopal episodes.
• A Head CT was taken in the ER again.
Head CT Without Contrast

• Anterior displacement of Medulla Oblangata and Edema

• Focal area of High attenuation posteriorly at Foramen Magnum

• The Ventrices and Sulci appeared normal
A MR brain with and with out Gadolinium contrast was ordered for further workup.
MR Brain Sagittal T1 Image

- Solid lesion producing mass effect on Medulla
- The Ventricles appear normal
MR T1 Sagittal View

- **Cystic portion** measuring 12 X 39 mm
- **Solid portion** of the Tumor measuring 18 X 20 mm
MR Sagital T1 image With Contrast

- Contrast Enhancing lesion at the level of Foramen Magnum
MR T1 Axial View

• Medulla Oblangata.

• Cerebellar Hemispheres
• Medulla Oblangata

• Cystic portion of the tumor
MR T1 Axial View

• Cystic portion

• Solid portion of the tumor
MR T2 Axial View

• Medulla Oblangata

• Cystic portion of the tumor
MR T2 Axial View

- Cystic portion
- Solid portion of the tumor
MR T1 Axial Postcontrast

- Medulla Oblangata
- Cystic portion of the tumor
MR T1 Axial Postcontrast

- Medulla Oblangata
- Cystic portion
- Solid portion showing contrast enhancement of the tumor
MR Axial FLAIR Images

- Medulla Oblangata
- Cystic portion of the tumor
MR Axial FLAIR Images

- Solid portion of the tumor
MR Axial Susceptibility Image

Image showing Tumor
MR Axial Diffusion Image

• No diffusion abnormalities/ acute/ subacute ischemic changes or restricted diffusion abnormalities were observed
MR Axial Diffusion ADC Image

• Despite the large size of the tumor, no diffusion abnormalities were noted in the surrounding structures
MR T1 Coronal View

- Cystic portion
- Solid portion of the tumor
• Solid portion of the tumor showing contrast enhancement of the tumor
• Cystic portion of tumor showing multiple septations versus small vessels
• Image showing solid and cystic portion of the tumor
CTA Reconstruction

- Anterior half of Circle of Willis
• Posterior circle with avidly enhancing vascular tumor.
• Feeding vessels are the Right & Left Posterior Inferior Cerebellar Arteries (PICA).
CTA - Complete Circle Of Willis

- Anterior Communicating artery
- Middle Cerebral artery
- Internal Carotid artery
- Basilar artery
- Vertebral artery
Surgical Resection

• The patient underwent Suboccipital Craniotomy and resection of the posterior fossa tumor.
• Post operative the patient was stable.
• Biopsy analysis of the resected tumor showed it to be a Hemangioblastoma.
Post-operative non-contrast CT images showed trace postoperative extra-axial hemorrhage and pneumocephalus.
Hemangioblastoma- Literature Review

- Uncommon, slow growing tumors of CNS.
- Commonest in the cerebellum, brainstem and spinalcord.
- Symptoms depend on the location and size of tumor within the CNS.
- May be sporadic or associated with Von Hippel Lindau (VHL) syndrome.
Hemangioblastoma- Symptoms and complications

- Cerebellar ataxia.
- Headache, Radiculopathy, Quadriplegia.
- Oculomotor nerve dysfunction.
- Motor weakness or sensory deficits.
- Pain if in the spinal cord.
- Intracerebral bleed.
- Obstructive hydrocephalus.
- Cerebellar tonsillar herniation.
- Brainstem compression.
Hemangioblastoma - Macroscopic Appearance

Hemangioblastoma- Differential Diagnosis

- **Astrocytoma**- Non enhancing on T1 Gadolinium sequences. Usually at very young age.
- **Ganglioma**- Similar to Astrocytoma in appearance and presentation.
- **Metastatic tumors**- Usually multiple, grey and white junction, large amount of vasogenic edema and ring enhancing appearance postcontrast administration.
Imaging Modalities: Computed Tomography

• Demonstrates as an enhancing cystic and/or solid lesion.

• Ideal for patients who can’t undergo MRI due to a metal implant.

- Pitfalls:

• Bone artifacts may obscure small tumors in posterior fossa and spinal canal.

• Can’t detect a cancer within a renal cyst in VHL patients.

• High exposure to radiation.
Imaging Modalities: Magnetic Resonance Imaging

• Most preferred
• Appears as well demarcated cystic lesion with a vascular mural nodule.
• No radiation so ideal for screening VHL patients and relatives and follow-up.
Magnetic Resonance Imaging Pitfalls

• Gadolinium contrast agents have recently been linked to the development of nephrogenic systemic fibrosis (NSF) or nephrogenic fibrosing dermopathy (NFD).

• Not for patients with metallic implants or claustrophobic.
Imaging Modalities: Ultrasonography

• Useful for screening abdomen of VHL patients and relatives for cystic lesions or tumors.

• Useful for screening retinal hemangiomas in VHL, as they are too small to be picked up by CT or MRI.

• No radiation, low cost, quick.

  - Pitfalls:

• Can’t differentiate cyst from a cancer.
Imaging Modalities: Nuclear Imaging & Angiography

• **Nuclear Imaging:**
  - Limited use.
  - Detect bone metastases in VHL.
  - Assess renal function before resecting a renal mass in VHL.

• **Angiography:**
  - Preoperative evaluation of feeding vessels of the Hemangioblastoma.
Hemangioblastoma and VHL

• Hemangioblastoma can be sporadic or associated with Von Hippel Lindau.

• Hemangioblastomas are the most common lesions associated with VHL disease, affecting 60 to 84% of patients.

• Von Hippel Lindau is a rare autosomal dominant disorder of chromosome 3.

• A VHL gene abnormality is present in about 1 in 36,000 newborns.
Features of VHL

- Multiple hemangioblastomas in Brain and Spinal cord.
- Retinal hemangiomas.
- Endolymphatic sac tumors.
- Renal Cell Carcinoma.
- Pheochromocytomas.
- Cysts in epididymis and testis.
- Liver and spleen cysts.
- Café au lait spots on skin.
Reasons To Identify VHL

• Hemangioblastoma is most common lesion of VHL.
• In VHL newer lesions can develop over time.
• Lesions can recur after treatment.
• Early detection and treatment of lesions help reduce morbidity and mortality.
• Patients and relatives of VHL need life long follow up.
Recent studies

• In a cohort of 188 consecutive patients presenting with a seemingly sporadic hemangioblastoma, no family history of VHL, and no other evidence of the disease, VHL germline mutations were present in 5 percent of cases.

• Of those who tested negative, 5 percent developed a VHL-related lesion in the ensuing years, which may result from being mosaic for a VHL mutation.

Woodward et al
Recommendation For Future Follow-up

- It is therefore recommend that all patients with either a retinal or CNS hemangioblastoma be tested for VHL germline mutations, even in the case of a single lesion* and be considered for routine follow-up to detect occurrence of new tumors.

*Gayun Chan-Smutko et al
Follow up of VHL

- Regular **physical** exam and **plasma catecholamines** to screen for pheochromocytoma.
- Regular **ophthalmoscopie** exam for retinal hemangiomas.
- Regular **abdominal Ultrasonographic** and **CT** exams for Renal/Liver/spleen cysts or tumors.
- Regular **MRI** or **CT** scanning of the **brain**.
- Regular **fluorescein angiography**.
Companion Patient 1 With VHL

- Fundoscopic exam showing dilated blood vessels and Retinal Hemangiomas.

From: http://www.ncbi.nlm.nih.gov/pmc/articles/PMC2430573.
• CT showing mass lesion in the right kidney in a VHL patient.

Treatment Of Hemangioblastoma

• **Surgical resection** of tumor.

• Stereotactic radiosurgery and conventional fractionated **radiation therapy** (RT).

• Vascular Endothelial Growth Factor (VEGF) **inhibitors** have been used at experimental levels.

• Patients and relatives should be referred for genetic counseling and testing for VHL mutation.

• Lesions can recur in VHL so need life long follow-up.
Summary

• Hemangioblastomas are well-circumscribed, capillary-vessel-rich benign neoplasms.
• Most commonly occur at cerebellum or brainstem.
• Symptoms depend on location of tumor.
• Can be associated with VHL and need lifelong follow-up of patients & relatives for early detection and treatment of tumors.
• Treatment is surgery or radiotherapy.
• Over lifetime newer lesions can occur or older lesions can recur despite treatment.
References (1)


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