Hemangioblastoma: Imaging findings

Poster No.: C-2543
Congress: ECR 2015
Type: Educational Exhibit
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Keywords: CNS, Neuroradiology brain, Neuroradiology spine, MR, CT, Diagnostic procedure, Pathology
DOI: 10.1594/ecr2015/C-2543

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Learning objectives

• To review the MRI features of intra and extra cranial hemangioblastomas.
• To discuss the differential diagnosis.
• Emphasize the role of the imaging techniques in the diagnosis and follow up of patients with hemangioblastoma within the von Hippel Lindau disease.

Background

The hemangioblastoma of the central nervous system is a low-grade highly vascularized tumor.

It is mainly in the cerebellum, brainstem and the spinal cord.

It may be sporadic or associated with Von Hippel-Lindau syndrome.

Findings and procedure details

I. MR Imaging Characteristics of cerebral Hemangioblastomas

Histologically and radiologically, hemangioblastomas are traditionally described as four types:

1. **Type 1**: 5% of posterior fossa HBs, is a simple cyst without abmacroscopic nodule (fig 1, 2)

2. **Type 2**: is a cyst with a mural nodule (60%) (fig 3, 4, 5)

3. **Type 3**: solid tumors (26%) (fig 6)

4. **Type 4**: solid tumors with small internal cysts (9%), are also seen in the cerebellum (fig 7, 8)

1. Behavior in conventional MRI sequences

*The solid component* is usually nodular hypo to isointense on T1, hyperintense on T2 and greatly enhanced after contrast injection. Occasionally, a heterogeneous hyperintense signal on T1 corresponding to lipids in the stromal cells or intratumoral hemorrhage.
**Cystic component**, its is pure fluid signal on all sequences: hypointense T1, hyperintense T2 and hypointense on FLAIR. It may be hyperintense on T1 and T2 due to protein content.

Cyst wall does not take the contrast, but if the cyst is surrounded by the tumor, its wall may enhance.

The association at the posterior fossa cyst containing a fleshy mural nodule fed by prominent vessels is pathognomonic of HB. These dilated vessels were identified in contact with the tumor in 37.5% of cases.

2. Behavior in new MRI sequences

**Diffusion:**

The solid component appears hyperintense on diffusion with low ADC while the cystic component has a low signal on DWI and high ADC.

**Perfusion:**

HB are benign vascular tumors. They have a very increased rVSCmax as well as disorders of the permeability. The vascular component of the HB is such that the rVSCmax is increased despite permeability disorders. This profile is found with lesions morphologically and histologically varied as glioblastomas, hypervascular metastases and meningiomas.

3. Differential diagnosis

**Solid lesion:**

- Metastases: the problem arises especially for multiple HB, which may be referred to the diagnosis of metastases.

The age of the patient and the clinical setting may help in the diagnosis. The diagnosis is further difficult in case of hypervascular metastases such as renal cell carcinoma, because in this case, although the contribution of perfusion MRI is limited. Indeed, in both cases it shows increased rVSCmax.

- Arteriovenous malformations: dilated appearance of flow void vessels in contact the tumor may suggest the presence of arteriovenous malformation but the contrast medium administration allows to highlight the tumoral mass.

- Other: medulloblastoma, lymphoma, ependymoma

*The lesions are most often cystic, so it is in this context that the diagnosis can be difficult.*
• The pilocytic astrocytoma: It is one of the main tumors to evoke especially if seat at the posterior fossa: this is a very low-grade tumors (WHO grade I) that contains frequently cystic portion.

This portion has a wall that raises more often, which is very unusual for HB.

The pilocytic astrocytomas and HB may have similar characteristics on conventional sequences T1, T2 and T1 with contrast. MRI perfusion "first pass" allows to advance in the diagnostic process by calculating the rVSCmax significantly longer with HB. The histological data can explain the behavior of tumor in perfusion MRI.

The other argument is the age of onset is in the vast majority of cases less than 20 years for pilocytic astrocytoma, which is a little less than 30 years on average for HB.

• The abscess: has a raised peripheral enhancement after injection, which helps differentiate it from the HB. In addition, the sequence of diffusion shows, more than nine out of ten times the "cystic" portion hyperintense with a low ADC in the case of an abscess, and hypointense with high ADC in the case of HB.

• Other cystic tumors, such as gangliogliomas or dysembryoplastic neuroepithelial tumors are much rarer.

II. MR Imaging Characteristics Of Spinal Hemangioblastoma

1. Positive diagnosis

When compared with the spinal cord, spinal hemangioblastoma is usually hypointense to isointense on T1-weighted sequences and isointense to hyperintense on T2-weighted sequences.

T1-weighted images after gadolinium administration show intense enhancement.

Large lesions may be visualized without contrast material, but small lesions are often isointense and thus difficult to differentiate from the spinal cord.

Therefore, gadolinium-enhanced T1-weighted images are essential for the evaluation of lesions suggestive of hemangioblastoma.

The 4 types of HB are seen and predominately type 2 and 3 (fig 9, 10).

Multiple hemangioblastomas have been described only in patients with von Hippel-Lindau syndrome.

Small hemangioblastomas may be observed in patients with no spinal symptoms or in relatives of patients with VHL.
Patients with VHL and spinal symptoms usually have one dominant lesion that causes symptoms, though other asymptomatic tumors may also be present.

Large hemangioblastomas may show flow voids resulting from prominent vessels. Rarely, spinal hemangioblastoma may hemorrhage in either the cord parenchyma or the subarachnoid space.

The majority of spinal hemangioblastomas originate in the thoracic region, with cervical lesions also common.

Lumbar and sacral hemangioblastomas are less frequent.

Hemangioblastomas may originate from any compartment of the spinal canal or within a vertebral body.

The spinal cord reacts to hemangioblastoma in a number of different ways; reaction depends on the size and location of the mass.

Sixty-six percent of spinal hemangioblastomas originate at or near the surface of the spinal cord (either intramedullary with extension beyond the surface of the spinal cord or exophytic with the center of the lesion external to the spinal cord).

Twenty-five percent of spinal hemangioblastomas are completely intramedullary and 8% are intradural extramedullary neoplasms.

**2. Descriptions of the different appearances of hemangioblastoma.**

**a. Exophytic Hemangioblastoma**

A hemangioblastoma that originates at the cord surface may incite little or no reaction in the adjacent spinal cord.

The tumor may grow outward in an exophytic manner.

When the tumor is large, the appearance may mimic nerve sheath tumors, meningioma, or ependymoma.

**b. Hemangioblastoma with Syrinx**

Hemangioblastoma is known to cause syringomyelia, a condition seen in 40% of patients.

The syrinx may be small and cystlike or may extend the length of the cord.
Syrinx is not specific for hemangioblastoma and is associated with other spinal tumors such as astrocytoma and ependymoma.

Because a small hemangioblastoma can cause a large syrinx, contrast-enhanced MRI is a valuable tool for the diagnosis of syrinx detected on unenhanced images.

**c. Hemangioblastoma with Associated Spinal Cord Enlargement**

A unique characteristic of hemangioblastoma is spinal cord enlargement beyond the margins of the enhancing tumor and distinct from syrinx. Spinal cord swelling may be caused by arteriovenous shunting, venous congestion, or an edema-promoting factor produced by the tumor. When spinal cord enlargement caused by hemangioblastoma is noted, surgical treatment should be directed at the enhancing nodule.

Spinal cord swelling will usually improve after the tumor is removed.

**d. Extradural Hemangioblastoma**

It is rare but may occur in the lumbar or sacral spinal canal; it may be associated with exiting nerve roots.

In this location the appearance is similar to that of meningioma or schwannoma. The key to preoperative diagnosis is the recognition of marked enhancement and associated enlarged vessels.

A hemangioblastoma originating in a vertebral body may be seen.

**e. Intramedullary Hemangioblastoma with No Spinal Cord Reaction**

A small intramedullary hemangioblastoma may not elicit significant cord enlargement or syrinx.

3. differential diagnosis

Spinal hemangioblastoma has a number of appearances. Specific appearances may help differentiate hemangioblastoma from other spinal tumors or vascular malformations.

Hemangioblastoma usually shows uniform contrast enhancement.
**Spinal astrocytoma** usually shows inhomogeneous enhancement with occasional central cavitation or hemorrhage.

**Spinal ependymoma** usually shows uniform intense enhancement with well-defined margins and occasional central cavitation.

**Spinal astrocytoma or ependymoma** may have associated syrinx, but an extensive syrinx suggests hemangioblastoma.

**Neither spinal astrocytoma nor ependymoma** is likely to have prominent vessels.

**Spinal cord thickening remote from the tumor and without syrinx** may be specific for hemangioblastoma.

**A spinal arteriovenous fistula** may show prominent vessels, cord thickening, and diffuse patchy enhancement caused by venous engorgement or infarction; however, spinal arteriovenous fistulas rarely show a well-defined enhancing mass lesion and are typically heterogeneous on unenhanced MR images.

Images for this section:
Fig. 2: MRI: (a) axial T1 (b) Axial and coronal T2 (c) Axial T2 FLAIR (d) Axial diffusion (e) axial T2 * (f, g, h) axial, coronal and sagittal T1 with gadolinium injection: intracranial expansive process developed in the vermis. It presents a cystic component hypointense on T1 hypointense on T2; and a posterior mural nodule enhanced homogeneously after injection. It is responsible of compression of V4 with a tri-ventricular hydrocephalus and a peri-lesional edema.
Fig. 3
Fig. 6: MRI: (a) coronal T2 (b) Axial T2 FLAIR (c) axial T2 * (d) axial T1 with gadolinium: intracranial expansive process of the posterior fossa, seating in the vermis. It exhibits tissue component on hyperintense on T2 and greatly enhanced homogeneously. It is crossed by tortuous vascular structures in flow void signal on T2 and T2 FLAIR W images. It is surrounded by a peri-lesional edema hyper intense on T2 and FLAIR.
Fig. 7
Fig. 8: MRI: (a) sagittal T1 (b) Coronal T2 (c) Axial T2 FLAIR (d) axial T2 * (e, f) axial slices diffusion and ADC mapping (g, h) axial and sagittal T1 with gadolinium injection: expansive intra cranial process of the right cerebellar hemisphere, heterogeneous hypointense T1 with hyperintense T1 zones related to hemorrhage. T2 and FLAIR w images the fleshy part is heterogeneous hyperintense on T2. Gadolinium, there is a socket ring and nodular intense contrast on the posterior inferior pole. Exists at this level multiple dilated vascular structures. It liable for tonsillar involvement and is associated with edema beach peri-lesional FLAIR hyperintense and T2.
Conclusion

Despite their rarity, MRI exam allows considering the diagnosis of haemangioblastoma preoperatively. That is essential to a better surgical setup as these lesions have a great propensity for bleeding. MRI allows the management and follow up of patients with hemangioblastoma associated with von Hippel lindau disease.

Personal information

References