

Spinal cord hemangioblastoma

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Usually non-infiltrating, well demarcated, may have cystic caps. 33% of patients with [spinal hemangioblastoma](#) will have [Von Hippel-Lindau disease](#). It cannot incise nor core because of vascularity. Requires microsurgical approach similar to [AVM](#), possibly with intraoperative [hypotension](#).

a) occur in 13–44 % of VHL patients

b) 90% are located cranially within the [cervical](#) and [thoracic cord](#). Almost all(96%) of the tumors are located in the posterior half of the spinal cord, 4% are located in the ventral half of the spinal cord. 1–3% are found in the lumbosacral nerve roots

c) by way of comparison, 80% of spinal cord HGB are associated with VHL, whereas only 5–31% of cerebellar HGB are associated with VHL

d) 95% of symptom-producing spinal HGBs are associated with [syringomyelia](#)

[Spinal hemangioblastoma](#) are intramedullary (75%) or extramedullary (25%); extramedullary tumors are mainly located at the [cauda equina](#), at the [filum terminale](#), and, less frequently, in the [spinal nerve root](#) ¹⁾

Cervical Spinal cord hemangioblastoma

see [Cervical Spinal cord hemangioblastoma](#)

Sacral hemangioblastoma

see [Sacral hemangioblastoma](#).

Extradural spinal nerve root hemangioblastoma

[Extradural spinal nerve root hemangioblastoma](#)

Epidemiology

see [Spinal cord hemangioblastoma epidemiology](#).

Pathology

Haemangioblastomas are vascular benign (WHO grade I) lesions and they do not undergo malignant degeneration.

Histologically, they consist of large pale stromal cells packed between blood vessels.

Clinical Features

Clinical presentation is similar to that of other [spinal cord tumors](#), with pain, weakness and sensory changes common. Rarely, spinal hemangioblastomas may cause subarachnoid haemorrhage or hematomyelia.

Radiographic features

The most common location is the thoracic cord (50%), followed by the cervical cord (40%).

The majority of haemangioblastomas have an intramedullary component with two thirds located eccentrically and having an exophytic component (most commonly along the dorsum of the cord 8). Only 25% percent are entirely intramedullary. A minority appear entirely extramedullary and only rarely are they extradural

Eighty percent of haemangioblastomas are solitary. If multiple lesions are present, Von Hippel-Lindau syndrome should be suspected.

The prevalence of syringomyelia due to SH is considerably high, and the initial clinical presentation of syringomyelia resulting from SH should be emphasized ²⁾.

Angiography

A densely enhancing nidus with associated dilated arteries and prominent draining veins is characteristic for a hemangioblastoma.

CT

On non-contrast CT They may be seen as a soft tissue nodule often with a prominent hypodense cyst-like component. Contrast administration results in vivid enhancement of the solid component.

MRI

Although they usually appear as discrete nodules, there can be diffuse cord expansion. An associated tumour cyst or syrinx is common (50-100%) 2,4. Additionally they may rarely be a source of subarachnoid haemorrhage or haematomyelia.

Reported signal characteristics of the solid components include:

T1: variable relative to normal spinal cord.

hypo-isointense most common, and difficult to identify

hyperintense (25%)

T2

iso-hyperintense

focal flow voids especially in larger lesions

surrounding oedema and associated syrinx are usually seen

hemosiderin capping may be present

T1 C+ (Gd): the tumour nodule enhances vividly

Care should be taken to image the whole neuraxis to ensure that no other lesions are present.

Differential diagnosis

The [differential diagnosis](#) can be thought of in two groups on account of the main features of these tumours: neoplasms of the spinal canal (enhancing component) and vascular malformations of the spinal cord (enlarged vessels)

Other enhancing masses to be considered include:

[intradural extramedullary tumors](#)

[spinal meningioma](#)

[myxopapillary ependymoma](#)

leptomeningeal [spinal metastases](#)

[neurogenic tumors](#)

[spinal schwannoma](#)

[spinal neurofibroma](#)

[spinal paraganglioma](#)

intramedullary spinal cord tumours

[spinal cord metastases](#)

[spinal astrocytoma](#)

[spinal ependymoma](#)

Vascular malformation to consider include:

other hypervascular cord neoplasms

[spinal arteriovenous malformation](#)

[spinal dural arteriovenous fistula](#)

[spinal cavernous malformations](#)

Treatment

[Spinal cord hemangioblastoma treatment.](#)

Outcome

Hemangioblastomas are slow growing.

Case series

[Spinal cord hemangioblastoma case series.](#)

Case reports

As vascular tumors, intramedullary [hemangioblastomas](#) are associated with significant intraoperative blood loss, making them particularly challenging clinical entities. The use of intraoperative [indocyanine green](#) or other fluorescent dyes has previously been described to avoid breaching the tumor capsule, but improved surgical outcomes may result from identifying and ligating the feeder arteries and arterialized draining veins.

To describe the use of combined preoperative angiography and intraoperative indocyanine green use for the identification of feeder arteries and arterialized draining veins to decrease blood loss in the resection of intramedullary hemangioblastomas.

A patient with [cervical myelopathy](#) secondary to a large C3 hemangioblastoma and cervicothoracic

syrinx underwent a C2-3 laminoplasty with resection of the lesion. To reduce intraoperative blood loss and facilitate safe lesion resection, the vascular architecture of the lesion was defined via preoperative digital subtraction angiography and intraoperative use of indocyanine green. The latter permitted ligation of the major and minor feeding arteries and arterialized veins prior to tumor breach, allowing for facile en bloc resection of the lesion.

The lesion was resected en bloc with minimal blood loss (approximately 100 mL) and without intraoperative neuromonitoring signal changes. The patient remained at neurological baseline throughout their stay.

Molina et al., presented a written and media illustration of a technique for intraoperative indocyanine green use in the en bloc resection of intramedullary hemangioblastoma ³⁾.

A 37-year-old female presented with bilateral lower extremity pain without a focal neurological deficit. The magnetic resonance (MR) image demonstrated an intradural spinal tumor at the L1 level, which was accompanied by peritumoral cysts. In addition, there were multiple serpentine flow voids (e.g., consistent with tortuous and convoluted vessels), which is typical for hemangioblastoma. At surgery, a spinal hemangioblastoma originating from the film terminale with peritumoral cysts at the L1 level was fully excised without producing a focal postoperative neurological deficit. Histological examination revealed stromal cells with vacuolated cytoplasm and small nuclei in a rich capillary network accompanied by several enlarged vessels. These findings were compatible with a hemangioblastoma.

Zaimoku et al. reported a rare case of a hemangioblastoma originating from the conus presenting at the L1 level. Complete surgical resection was accomplished without any motor deficit ⁴⁾.

References

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