

# CAUDA EQUINA HEMANGIOBLASTOMA

## Case report

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**ABSTRACT** - Hemangioblastomas of the spinal cord are rare lesions, and those located at the cauda equina are even rarer. Most commonly these tumors are present in patients with von Hippel-Lindau (VHL) syndrome. We describe here the case of a 48 years old woman with a pure radicular hemangioblastoma, not associated with VHL, presenting with radicular pain, diagnosed with magnetic resonance imaging (MRI) and submitted to total resection with a very good outcome. To our knowledge, this is the second report to describe the MRI aspect of histologically proved hemangioblastoma of the cauda equina in a patient without clinical criteria for VHL.

**KEY WORDS:** cauda equina, hemangioblastoma, Von Hippel-Lindau syndrome.

### **Hemangioblastoma da cauda equina: relato de caso**

**RESUMO** - Hemangioblastomas da medula espinhal são lesões raras, sendo sua localização na cauda equina ainda mais incomum. Estes tumores são diagnosticados com mais frequência em pacientes portadores da síndrome de Von Hippel-Lindau. Descrevemos o caso mulher de 48 anos hemangioblastoma radicular na cauda equina, diagnosticado com ressonância nuclear magnética, não associado à síndrome de Von Hippel-Lindau, tratado cirurgicamente com ótimo resultado. Em nossa revisão, este é o segundo relato de diagnóstico com ressonância nuclear magnética de hemangioblastoma da cauda equina em paciente sem critérios clínicos da síndrome de Von Hippel-Lindau.

**PALAVRAS-CHAVE:** cauda equina, hemangioblastoma, síndrome Von Hippel-Lindau.

Hemangioblastomas represents about 1.6 to 2.1% of all spinal cord tumors. Extramedullary-intradural hemangioblastomas are rarer, representing only one fifth of all spinal hemangioblastomas. One recent report suggested that the number of reported cases of hemangioblastoma of the cauda equina may be as low as 40. Most commonly, these lesions are associated with VHL disease, an heredo-familial autosomal dominant genetic disease with incomplete penetrance<sup>1-6</sup>.

We relate here a case of an hemangioblastoma of the cauda equina, diagnosed in a patient without clinical criteria to VHL disease, and show the aspects of MRI imaging of this rare disease.

### **CASE**

A 40-years-old woman presented with left leg pares-

thesia for twenty days prior to hospital admission. Her physical examination showed a discrete weakness for left leg extension, patellar arreflexia and hypoesthesia in the medial side of the same leg. The CT scan and eletromyography were unremarkable, and MRI showed an intradural, hyperintense lesion, with brightly enhancing after gadolinium infusion, at the aproximated level of the L2-L3 disc space (Figs 1 and 2). The patient was submitted to a L2-L3 laminectomy and complete resection of a reddish-brown, highly vascular lesion. The tumor was attached to some radiculae of the fourth left lumbar root, but a total removal was accomplished. In the immediate pos-operative period she showed a little worsening in the weakness of the left quadriceps, which improved over time. Six months after surgery she was asymptomatic, and showed no neurologic deficit. Histopathological examination demonstrated hemangioblastoma (Fig 3). Von Hippel-Lindau clinical screening was negative.

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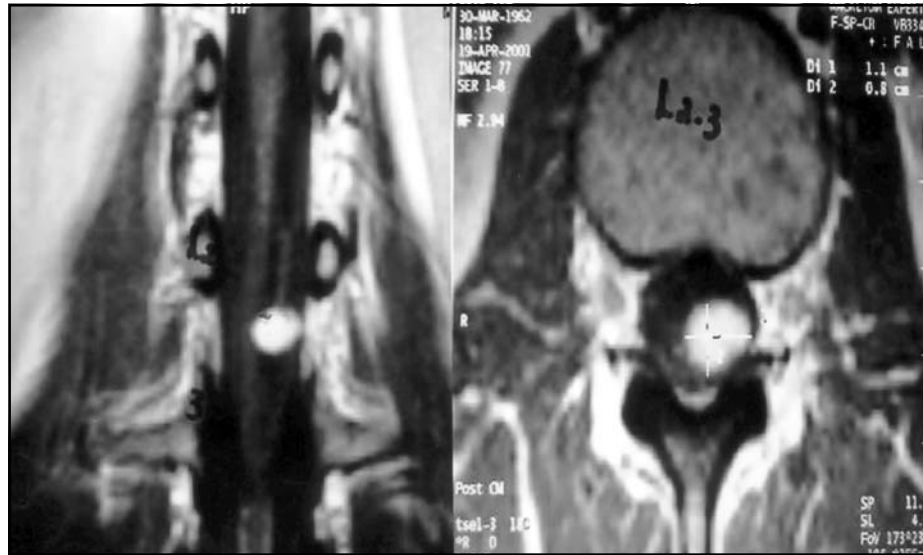


Fig 1. Coronal and axial T1 MRI with gadolinium infusion, showing enhancing intradural lumbar lesion.

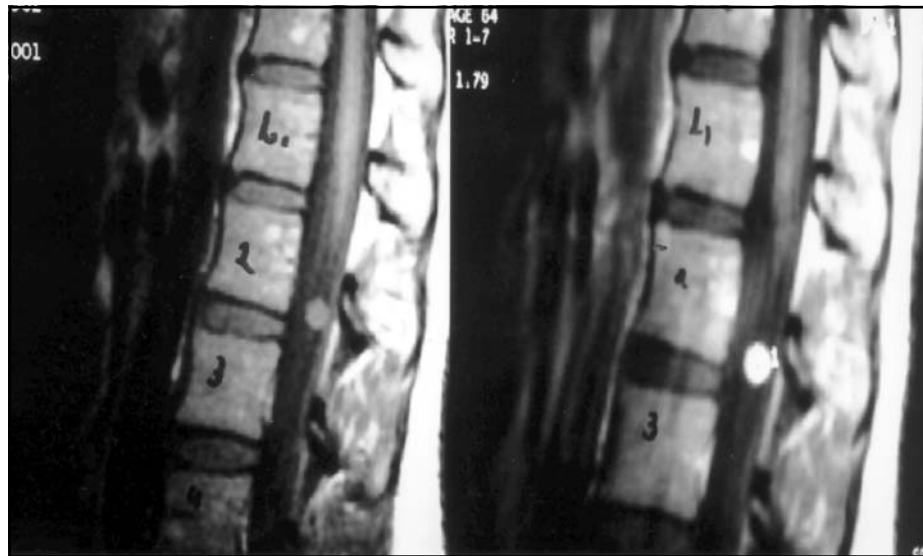


Fig 2. Sagittal T1 MRI with and without gadolinium infusion, showing tumor enhancement at L2L3 level.

## DISCUSSION

Spinal cord tumors account for about 15% of all central nervous system neoplasms, and only about 10% of these are found in the cauda equina. Primary tumors of the cauda equina are rare lesions. Most commonly, they are ependymomas and schwannomas. Hemangioblastomas account for 1.6% to 2.1% of all spinal cord tumors. The majority of these lesions are intramedullary, but hemangioblastomas have been described in all spinal levels and compartments, with cervical and thoracic lesions predominating. Extramedullary disease is distinctly uncommon, representing approximately one fifth of all spinal hemangioblastomas<sup>1,2,6,7</sup>.

Histologically, hemangioblastomas are benign lesions, partly cystic, characterized by stromal cells containing foamy cytoplasm in endothelium-lined vascular channels. The central nervous system lesions are located mainly in the retina and cerebellum; less often they occur in the medulla and spinal cord, and very seldom supratentorially. Medullary and spinal cord lesions are more common in patients with VHL disease, and sporadic lesions are mostly cerebellar. Von Hippel-Lindau disease is an hereditary autosomal dominant disease with incomplete penetrance, characterized by intracranial and intraspinal hemangioblastoma, often multiple, retinal hemangioblastoma, cystic lesions in the kidneys, liver, pancreas and epididymis, benign and malignant re-

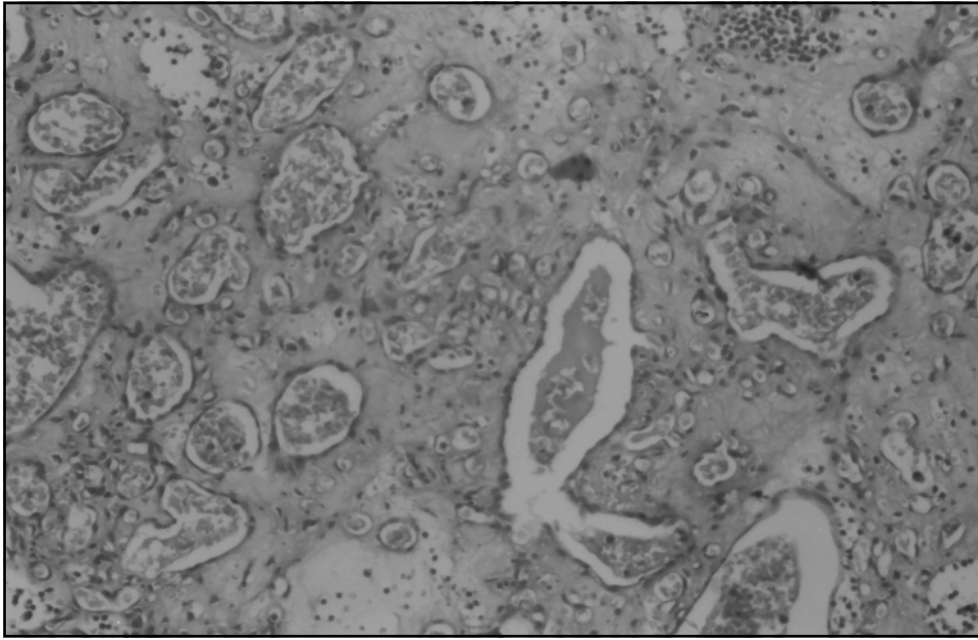


Fig 3. High magnification showing large stromal cells with hyperchromatic nuclei and abundant capillary channels (HE, 200X).

nal cell tumors. The vascular tumor may be the sole manifestation of this syndrome<sup>8</sup>. Although histologically benign, in the central nervous system these lesions may be devastating, especially if presenting in the posterior fossa or intramedullary.

In our review, we found only three cases of intradural hemangioblastomas of the nerve root in patients without the stigmata of VHL syndrome, and only one was documented with magnetic resonance imaging, as ours own<sup>1,4,9</sup>. Some authors recommend the superselective spinal angiography to distinguish between intradural hemangioblastoma and spinal arteriovenous fistulae<sup>1</sup>, but in this case, the MRI was quite sufficient for surgical planning. MRI showed a small nodular lesion, attached to lumbar root, hyperintense in T1 and isointense in T2, markedly enhanced with gadolinium infusion. This is not the typical imaging finding in intracranial lesions, that usually appears as cystic lesions with enhancing intramural nodules. Nevertheless we agree that angiography could be very useful in planning the surgical management of these lesions. Myelography could also suggest the diagnosis, showing the serpiginous vasculature and the mass interfering with the contrast filling, if MRI and angiography are not available.

The treatment of choice for these tumors is total surgical resection, which, if accomplished, is usually curative. At surgery, intralesional debulking should not be performed. These tumors should be dissected and removed en bloc, once the intralesional resection, even with the smaller lesions, will be associated with profuse bleeding<sup>1,2,5,7,10</sup>.

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