

Case Report

A case of intraroot cauda equina cavernous angioma: clinical considerations

E Caroli*¹, M Acqui¹, G Trasimeni², D Di Stefano³ and L Ferrante¹

¹Neurosciences Department – Neurosurgery, St Andrea Hospital-Second Faculty of Medicine, La Sapienza University Medical School, Rome, Italy; ²Neurosciences Department, St Andrea Hospital-Second Faculty of Medicine, La Sapienza University Medical School, Rome, Italy; ³Department of Experimental Medicine, St Andrea Hospital-Second Faculty of Medicine, La Sapienza University Medical School, Rome, Italy

Study design: Case report and literature review.

Objective: Intradural-extramedullary cavernoma is a rare condition with only 23 cases reported in the literature. Most cases described adhere to spinal root or spinal cord. We report an example of this rare entity located within the sheets of one cauda equina nerve root.

Setting: Rome, Italy.

Methods: The authors report clinical, radiological, surgical, and pathological features of intraroot cavernoma with a literature review.

Results: Patient was operated. One year after surgery, he was healthy.

Conclusion: Lumbar intradural cavernous angioma may present with sciatalgia, low-back pain, neurological deficit, or, more rarely with subarachnoid hemorrhage or hydrocephalus. They are treated successfully with surgical resection but preoperative status is a predicting factor for outcome. Sometimes the sacrifice of the spinal root is inevitable because, as in present case, the cavernous angioma is included in the nerve root. These lesions are rare and benign lesions. It is important for neurosurgeons to be aware of the existence of this entity to avoid preoperative misdiagnosis with tumor.

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Introduction

Central nervous system (CNS) cavernomas are rare vascular malformations consisting of closely packed large sinusoid-like vascular channels with little or no intervening nervous tissue. They can occur anywhere in the CNS, but they favor cerebral hemisphere.^{1–4} In the spine, cavernomas are found far more frequently in the vertebral bodies and, when involve intradural space, they occur primarily within the spinal cord.^{5–7} Intradural-extramedullary location is very rare, with only 23 cases reported in the literature accessible to us by Medline Research.^{8–27}

We describe a case of cavernoma located within the boundaries of a lumbar nervous root and we discuss its clinical, radiological, and surgical findings.

Case report

A 71-year-old man presented to our institution with low-back pain radiating into the left leg. The pain was exacerbated by the supine position.

On physical examination, the patient was found to have hypoesthesia in the left L4 dermatome and absence of the rotuleus reflex.

Lumbar MRI showed a left sided intradural-extramedullary nodular mass at the L3 level. The lesion appeared slightly hyperintense on T1-weighted images without contrast enhancement and hypointense on T2-weighted images (Figure 1a–c).

An L3 bilateral laminectomy was performed and the dura was opened under microscopic magnification. A reddish lesion was identified incorporated into one nerve root of the cauda equina (Figure 2a and b).

Histopathological examination revealed typical features of a cavernous angioma within the sheath of a spinal nerve and presence of nervous radicle peripherally to the vascular malformation (Figure 3a–d).

The postoperative period was uneventful and the lumbosciatalgia resolved after operation. One year after surgery, the patient was healthy.

Discussion

Cavernous angioma has been described occurring throughout the entire CNS.^{1–4,6,15} The most common

*Correspondence: E Caroli, Department of Neurological Sciences, Policlinico S Andrea, Via Meropia, 85, Rome 00147, Italy

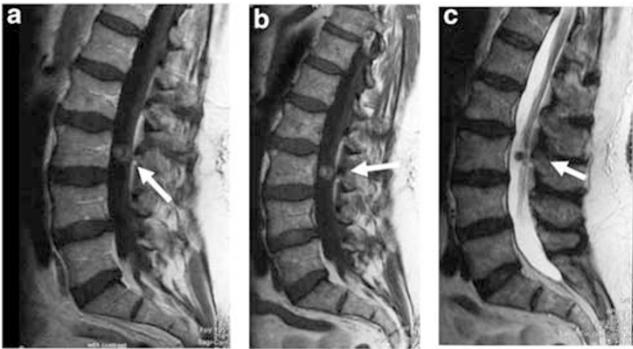


Figure 1 Lumbar MRI shows a nodular mass at the L3 level. The lesion is slightly hyperintense on T1-weighted images (a) without contrast enhancement (b) and hypointense on T2-weighted images (c)

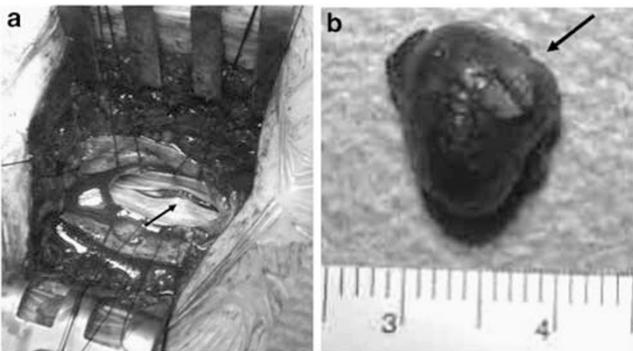


Figure 2 Intraoperative view: the dura has been opened; the arrow indicates the angioma included in a nerve root (a). The angioma has been removed; the arrow indicates the root stamp (b)

location of these vascular malformations is the supratentorial cerebral parenchyma. Cauda equina nerve root is a very rare location.^{4,9-13,15-19,21-26} Pagni *et al*¹⁵ reviewed all intradural cavernous angioma of the spine described in the literature. They concluded that subdural extramedullary cavernous angiomas are the most rare of the cavernomas with only 10 cases documented between 1903 and 1990. Although these vascular malformations are being discovered most frequently because of the widespread use of MRI, they remain still uncommon lesions. A Medline search identified just 24 cases, including the present case (Table 1).⁸⁻²⁷

The patients are in their third to seventh decades of life. The most common age at diagnosis is during the third decade. There is a male predominance (M:F ratio 2:1); and the lesions are mostly located in the lumbar region (13 cases – 54%), followed by lower thoracic (seven cases – 29%), cervical region (three cases – 12%), and thoraco-lumbar passage (one case – 4%). Low-back pain and radiculopathy are the main symptoms associated or not with motor deficit or neurologic symptoms secondary to spinal cord compression. However, 37% of patients developed a subarachnoid hemorrhage with acute onset of symptomatology. More rarely (12%),

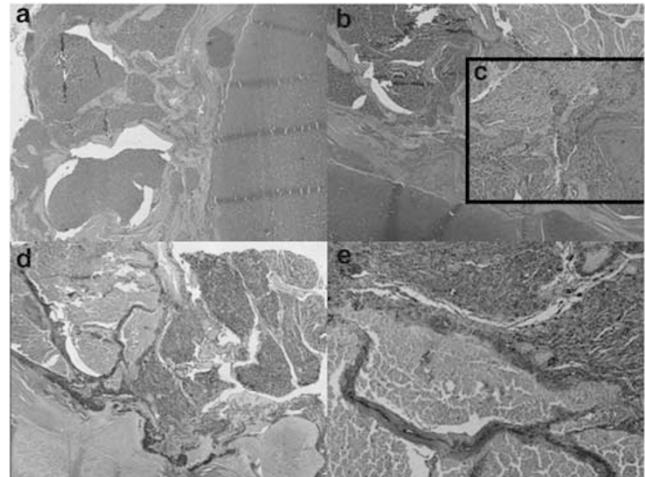


Figure 3 Thin-walled multiple vascular spaces of different diameter. H&E, 2.5 × (a). A large nerve appears at a margin of the lesion. H&E, 2.5 × (b). In the inset (c), a higher magnification of (b) evidences small nerve fibers compressed by hamartomatous vessels and the broader peripheral nervous component. H&E, 10 ×. Staining of nerves by immunohistochemistry for protein S-100 in the same field shown in Figure 2. PAP method, 2.5 × (d). Higher magnification evidences nerve fibers outside the lesion and entrapped in the wall of hamartomatous vessels. PAP method, S-100 protein, 10 × (e)

symptoms of intracranial hypertension caused by hydrocephalus have been found.

The preoperative radiological diagnosis of intraroot cavernous angioma is quite difficult, probably because of the rarity of this lesion.

Cavernous angiomas are lesions angiographically occult, but are easily diagnosed by MRI. MRI is the imaging modality of choice.^{5,29} The typical MRI features are those of a well-defined lesion with mixed signal intensity on both T1- and T2-weighted images. They are often surrounded by a hypointense ring on T2-weighted images owing to hemosiderin deposition.⁸ Enhancement with Gadolinium is variable.²⁸ In the present case, we found no significant contrast enhancement in the cavernous angioma, probably because the intralésional blood flow was too low.

The treatment of choice is surgical removal. The most frequent surgical finding is that the lesion adheres to the nerve roots, dura, or spinal cord extending extraphytically.²⁵ In the present case, cavernous angioma was within one spinal root. This finding was confirmed by histological examination that showed the disarrangement of radicles. A similar pattern of growth is reported only in another two cases.^{15,26}

Histologically, cavernous angioma must be distinguished from capillary hemangioma. Capillary hemangioma consists of a myriad of small vessels of capillary size. Cavernous angioma consists of large, dilated hyaline vascular channels arranged in diffuse patterns. They often show thrombosis, perivascular hemosiderin deposition, and calcifications.^{4,29,30}

Table 1 Surgically treated spinal intradural-extramedullary cavernous angioma

Author	Age/sex	Location	Presenting symptoms	Surface of origin	Extent of removal	Outcome
Roger <i>et al</i> ⁸	22/F	T11	Sciatic and back pain, M deficit	NR	T	Worse
Floris ⁹	57/M	T12	M deficit	NR	T	NR
Hirsch <i>et al</i> ¹⁰	20/M	L2–3	SAH (1), SM deficit, sphincteric dysfunction	Roots	T	Incomplete recovery
Pansini and Lo Re ¹¹	46/M	L2	Sciatic and back pain, SM deficit, sphincteric/erectile dysfunction	Root	Excision	Incomplete recovery
Ortner <i>et al</i> ¹²	22/M	C4–7	SAH (1)	NR	T	No improvement
Heimberger <i>et al</i> ¹³	24/M	T2–3	SAH (4)	Root	T	Excellent
Ueda <i>et al</i> ¹⁴	28/M	L1–2	Pain, SAH (2)	Root	T	Excellent
Pagni <i>et al</i> ¹⁵	46/M	T12–L1	Back pain	Root	T	Excellent
Ramos <i>et al</i> ¹⁶	67/F	L3	Hydrocephalus, sphincteric dysfunction, difficulty ambulating	Filum terminale	T	Excellent
Mastronardi <i>et al</i> ¹⁷	49/F	T4	SM deficit	Root	T	Excellent
Mori <i>et al</i> ¹⁸	65/M	T1	SAH (2)	Tumor adherent to the cord surface	T	Excellent
Acciarri <i>et al</i> ¹⁹	54/F	C2–3	SAH (2)	Dura mater	T	Excellent
Sharma <i>et al</i> ²⁰	63/M	T12	Back pain, SM deficit, sphincteric dysfunction	Root/spinal cord	T	No improvement
	43/M	T5	SAH (1)	Root/spinal cord	T	Excellent
Bruni <i>et al</i> ²¹	28/M	L2	SAH (1)	Root	T	Excellent
Cervoni <i>et al</i> ²²	26/F	L1–2	SAH (1)	Root	Total	Improved
	32/M	L5	Pain	Root	Total	Incomplete recovery
Makino <i>et al</i> ²³	67/M	L2	Hydrocephalus	Root	T	Excellent
Rao <i>et al</i> ²⁴	60/M	L1–3	SM deficit	Root	T	Excellent
	35/F	T12	SM deficit	Spinal cord	ST	No improvement
Duke <i>et al</i> ¹	49/F	L4	Sciatic and back pain, sensory deficit	Root	T	Excellent
Nozaki <i>et al</i> ²⁵	51/M	C5–6	SM deficit	Root/dentate ligament	T	Excellent
Falavigna <i>et al</i> ²⁶	44/F	L4	Lower extremity numbness, sphincteric dysfunction	Root	T	Excellent
Present case, 2004	71/M	L4	Sciatic and back pain, sensory deficit	Intraroot	T	Excellent

F, female; M, male; NR, not reported; SM, sensorimotor; SAH, subarachnoid hemorrhage; T, total; ST, subtotal
Number in parentheses indicates the number of SAH episodes

Most of the cases surgically treated had excellent results, but in patients with severe preoperative neurological deficits, such as sphincter dysfunction, recovery was not complete.^{1–27} Total excision was obtained in most of the cases. Subtotal removal was performed only in one case in which the cavernoma tightly adhered to the spinal cord.²⁴ In most cases, the lesion has been removed sparing the nerve root, but sometimes this is not possible because the lesion originates within the nerve root. This occurred in our case. However, our patient had a good recovery, even if it was necessary to cut the nerve root.

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