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Cauda Equina Intradural Extramedullary Cavernous Haemangioma: Case Report and Review of the Literature

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Abstract

Cavernous haemangioma (cavernoma) is a benign vascular lesion, exceptionally located in cauda equina. We report a case, diagnosed and operated in the Department of Neurosurgery from Pitesti County Emergency Hospital, of a 60-year-old woman with history of lumbar region distress, who presented with low back pain, paravertebral muscle contracture, and bilateral lumbar radiculopathy, with sudden onset after lifting effort. The preoperative diagnosis was done using computed tomography (CT) and magnetic resonance imaging (MRI), and the patient underwent surgery—two level laminectomy, dural incision, and tumor dissection from the cauda equina nerve roots under operatory microscope. Histopathological examination confirmed the positive diagnosis of cavernoma of cauda equina. The patient's outcome was favorable, without postoperative neurological deficits.

Key words: cauda equina, cavernoma, cavernous haemangioma

Introduction

Vascular tumors are some of the few groups of tumors showing an extremely wide spectrum of morphologic appearances and clinical behavior in which the line between neoplasia and malformation (or so called hamartoma) remains undefined, resulting in classification problems.[2] Of these, hemangiomas occupy a gray zone between hamartomatous malformation and true neoplasm, being frequently designated and regarded as tumors because of their usually localized nature and mass effect.[19,20]

Cavernous angiomas (or cavernomas) are a variant of haemangioma consisting of red-blue, soft, spongy masses 1 to 2 cm in diameter, sharply defined but not encapsulated, and composed of large, cavernous blood-filled vascular spaces, separated by a modest connective tissue stroma.[16] They can be found in all locations within the central nervous system, with an incidence of 0.02–0.9% (depending on the type of study: clinical, radiological or autopsy study) but quite rarely in the spine where they account for 5–12% of all vascular lesions.[4] The spinal intradural extramedullary space is a very rare location of cavernomas.[3] If we take into consideration only cauda equina section, we can discuss about the rarest location of a cavernoma, with only 25 cases reported as surgically treated in the literature accessible to us by Medline Research and Google Scholar (full articles but also abstracts, not always fully relevant, as in the two cases of Khalatbari et al.[13] where only general location and symptoms were mentioned), its incidence increasing in the last two decades because of its easy identification on magnetic resonance imaging (MRI) scans.[6] Therefore, we describe here the case of a cavernoma we found in this unusual site and which was treated. Here, we try

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to define a clinical, imagistic, and therapeutic profile starting from all cases reported till now in the literature.

Case Report

I. Clinical data

We report a case of a 60-year-old woman, admitted in the Department of Neurosurgery with agonizing low back pain, lumbar paravertebral muscle contracture, and marked bilateral radiculopathy relatively poorly systematized, symptoms with sudden onset after strenuous lifting exercise, 4 days before admission. The patient had no significant family or personal pathological history. Neurological examination revealed only bilateral L4-S1 mild paresthesia and hypoesthesia, deep tendon reflex changes, consisting of bilateral diminished ankle jerk reflexes and paravertebral muscle contracture with severe limitation of thoraco-lumbar spine anteflexion. All these data oriented the positive clinical diagnosis towards lumbar disk pathology.

II. Imagistic data

Lumbar X-rays oriented the diagnosis towards lumbar disc degenerative pathology, highlighting a plucked L5-S1 disk space, and posterior osteophytes at this level. Lumbar computed tomography (CT) scan disclosed a spontaneous hyperdense area, located at L4, apparently intradural. Lumbar MRI examination revealed a well-defined lesion situated in cauda equina with high signal in T_2-weighted images and discrete peripheral deposits of hemosiderin (Fig. 1a–c).

III. Therapeutical management

During hospitalization the patient received analgesics, muscle-relaxant drugs, and cortisone. The lack of symptoms improvement made us consider surgery. We performed a two-level laminectomy. After median opening of dura mater, a 1.5 cm in diameter, purple-reddish, mulberry-like, well-circumscribed lesion was found, growing between cauda equina nerve roots at L4 level (Fig. 1d). It was located strictly extraneural, posterior to cauda equina nerve roots in an arachnoid double fold. The lesion was suitable for total removal, favored by the lack of contact

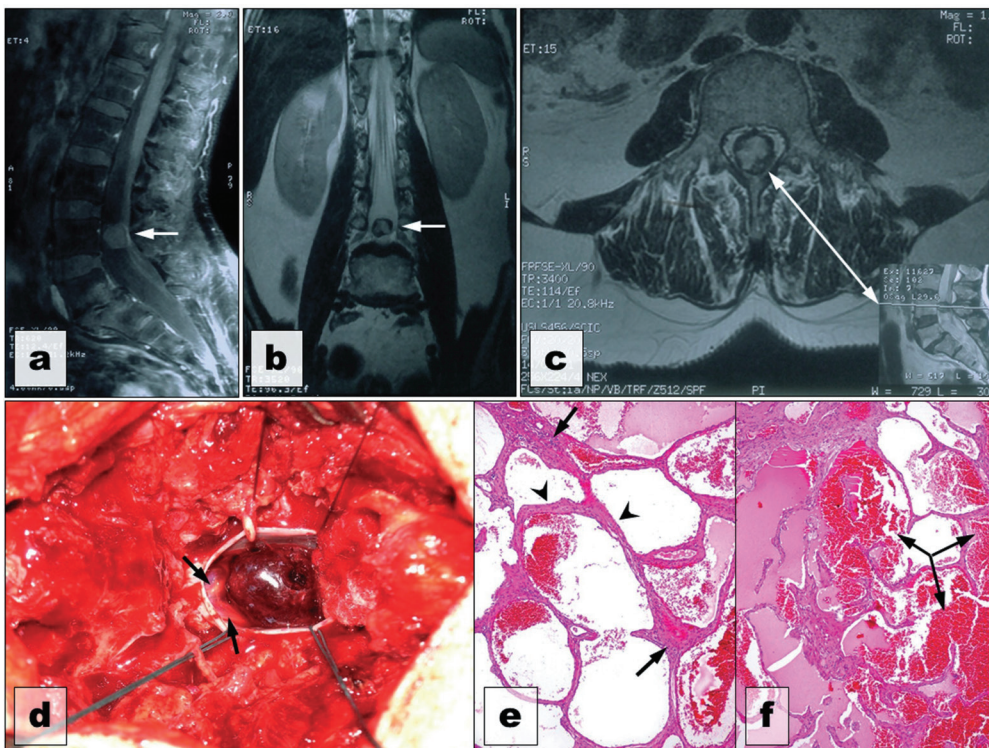

Fig. 1 a: Sagittal lumbar spine, T_1- weighted: well-defined, high signal lesion, located into the spinal canal behind L4 vertebral body. b: Coronar lumbar spine, T_2-weighted: well-circumscribed, mixed-signal, intradural lesion, located between cauda equine nerve roots. c: Axial lumbar spine, T_2-weighted: welldefined, intradural, round, occupying the spinal canal almost completely. d: Intraoperative view: well circumscribed tumor located between cauda equina nerve roots (*black arrows*). e: Irregularly dilated and packed vascular spaces separated by connective tissue walls, thickened by fibrosis (*black arrowheads*) and housing in many places a visible inflammatory cell population. f: Enlarged vascular spaces by the rupture of the thin connective tissue walls, with intratumoral hemorrhage and clot tendency (*black arrows*).

to nerve roots or filum terminale. Filum terminale can be easily districted from the nerve roots, because of its sinuous vessels pattern on the surface. We dissected cavernoma from cauda equine nerve roots, under operatory microscope. No nerve root was injured during dissection. After careful hemostasis, we proceeded to dissect dura mater closure with simple continuous suture (surjet) and followed by wound closure in anatomical layers, without aspirative drainage.

Intraoperative electrophysiological monitoring was not used, and intraoperative findings showed clearly that the cavernoma was not originating from the nerve roots.

IV. Histopathological examination

The low power magnification revealed the characteristic feature of large, irregularly dilated blood-filled vessels, arranged in a diffuse haphazard pattern, lined by flat endothelium walls made of connective tissue stroma, of varying thickness, usually thin, but sometimes enlarged by an adventitial fibrosis. Inflammatory cells could be observed, scattered throughout the stroma (Fig. 1e). In some tumoral areas, the walls laceration resulted in the formation of cystic spaces, which determined intratumoral hemorrhage with the tendency to form clots (Fig. 1f).

V. Patient outcome

The immediate postoperative outcome consisted of complete clinical remission, with no motor or sensory deficits, nor sphincter dysfunctions. Clinic and imaging follow-up at 1 and 2 years after surgery revealed no clinical or radiological recurrence.

Discussions

I. Location

According to the data collected up to now (Table 1), location of cavernoma in cauda equina is exceptionally rare, our case being the 26th reported in the literature. Cauda equina is indeed the least frequent site of occurrence of spinal cavernoma but the most common location for its intradural extramedullary variant.[15] When we designed our review, we took into account only the reported cases with tumors placed starting from L1 level, excluding the cases of Pagni et al., cited by Er et al.,[10] Moreno Rojas et al. cited by Miyake et al.,[17] Er et al.,[10] and Jin et al.,[12] where the tumor originated at T12 level with subsequent caudal extension to L1 level.

II. Clinical profile

It seems that this type of cavernoma has no gender predilection, the distribution being equal (12/12), except the two cases of Khalatbari et al.[13] The patients' mean age at diagnosis is 49 ± 18.1 years, with a wide range, between 18 years and 75 years.

The tumors had clinical expression over the age of 30 (only 5 cases under this age), with more than 1/3 of cases aged over 60 years, suggesting a slow but progressive increase of their volume in time.[4,9]

The origin point (not mentioned in 4 cases) was located, in a decreasing hierarchy, at L2 level (7 cases), then L1 and L5 levels (with 5 cases each), and L3 level (3 cases). Almost half of the tumors originating between L1 and L3 (7/15 cases) were extended to more than one level.

The most frequent clinical syndrome is the adjacent nerve roots compression syndrome, known as "cauda equina syndrome" (back and/or sciatic pain, lower motor neuron deficit and/or sensory loss, sphincter and/or sexual dysfunctions),[11] present in 19 cases, in 14 of which being the only expressed clinical syndrome. In only 2 cases the symptomatic triad was complete, the patients usually presenting (11/19 cases) only one symptom (either pain—7 cases or sensorimotor/sensory deficit—4 cases). However, pain (back pain or/and sciatic pain) is the most frequent symptom (13 cases), followed by sensorimotor/sensory deficit (12 cases). Sphincter dysfunction was present in only 4 cases.

Subarachnoid hemorrhage (SAH) syndrome (headache, nuchal rigidity, and vomiting)[11] is mentioned in 6 cases, in 2 of which being the only expressed clinical picture. Symptoms of intracranial hypertension (ICHT) caused by hydrocephalus were present in only 2 cases (Table 1, cases 4 and 10). Only one case revealed all three syndromes (Table 1, case 10).

The onset (not mentioned in 5 cases) is usually subacute-chronic (days, months, or years), with progressive accentuation of symptoms over time. It was, however, acute in 9 cases, and very important, in 4 of them being determined by SAH syndrome. SAH signs could mask other clinical picture in the acute presentation, and thus, make the diagnosis difficult.[18] Our case had an acute onset, but with an incomplete compression syndrome, lacking sphincter/sexual dysfunctions.

In our case, the onset was acute, despite the lack of subarahnoid hemorrhage. We presume that it was the result of a sudden mass extension, secondary to intralesional hemorrhage, without breaking the capsule, as revealed by the histopathological examination (Fig. 1f).

Cavernomas may be sporadic or multiple. Our case was most probably sporadic. The spinal MRI (cervical, thoracic, and lumbar) showed no other lesions. Unfortunately, the patient refused a cerebral MRI. She had no family history for cavernomas. No genetic studies were conducted.

III. Imagistic diagnosis

Cavernomas are occult lesions on angiography. CT scan is usually inconclusive (hyperdense spots caused by calcium deposits, in some cases). MRI is the imaging technique of choice, given the characteristic appearance of cavernomas

Table 1 Synopsis of reported cases with surgically treated intradural extramedullary cavenous haemangiomas

No	Author, year	Age (yrs)/ Sex	Presentation symptoms/ symptomatic period-onset	Level	MRI	Origin	Surgery extent	Follow-up/ Outcome
1	Hirsch et al. 1965 cited by[10]	20/M	SAH Sd + Cs Sd (SM deficit, Sph-dys)/10d	L2–3	No	Root	Total (+RR)	2 yrs/IR
2	Pansini and Lo Re, 1966 cited by[10]	46/M	Cs Sd (Sciatica and back pain, SM deficit, Sph-dys)/6 mo	L2	No	Root	Total	1 yr/IR
3	Ueda et al. 1987 cited by[10]	28/M	SAH Sd + Cs Sd (Pain)/SO	L1-2	Yes	Root	Total	3 wks/Excellent
4	Ramos et al. 1990 cited by[10]	67/F	ICHT Sd (Cognitive dysfunction) + Cs Sd (Sph-dys, Gait dis)/3–4 mo	L3	No	FT	Total	3 yrs/Excellent
5	Ahn et al. 1992[1]	NM/F	NM	NM	Yes	NM	Total	NM
6	Ahn et al. 1992[1]	NM/F	NM	NM	Yes	NM	Total	NM
7	Bruni et al. 1994 cited by[10]	28/M	SAH Sd/SO	L2	Yes	Root	Total	7d/Excellent
8	Cervoni et al. 1995 cited by[10]	26/F	SAH Sd/SO	L1-2	Yes	Root	Total	DH/Excellent
9	Cervoni et al. 1995 cited by[10]	32/M	Cs Sd (Back pain, SM deficit)/3 yrs	L5	Yes	Root	Total	6 mo/Excellent
10	Makino et al. 1995 cited by[17]	67/M	ICHT Sd + SAH Sd + Cs Sd (Gait dis)/3 yrs	L2	Yes	Root	Total (+RR)	6 mo/Excellent
11	Choi et al. 1996[5]	46/F	Cs Sd (Back pain)/NM	L1	Yes	NM	Total	DH/ Excellent
12	Rao et al. 1997 cited by[10]	60/M	Cs Sd (SM deficit)/NM	L1-3	NM	Root	Total	NM/Excellent
13	Duke et al. 1998 cited by[15]	49/F	Cs Sd (Sciatic and back pain, S deficit)/SO	L4	Yes	Root	Total	3 mo/Excellent
14	Kim et al. 2001[14]	65/M	NM/NM	L4	Yes	Root	NM	NM
15	Park et al. 2003[18]	33/M	Cs Sd (Back pain)/1 yr	L2-L3	Yes	Root	Total	DH/Excellent
16	Falavigna et al. 2005[11]	44/F	Cs Sd (Back pain, SM deficit, Sph-dys)/4 mo	L3-4	Yes	Root (intra)	Total (+RR)	6 mo/Excellent
17	Chung DY et al. 2005[7]	52/M	CS Sd (Back pain, S deficit)/2 yrs	L2	Yes	Root (intra)	Total (+RR)	DH/Excellent
18	Caroli et al. 2007[3]	71/M	Cs Sd (Sciatic and back pain, S deficit)/NM	L4	Yes	Root (intra)	Total	1 yr/Excellent
19	Miyake et al. 2007[17]	18/M	CS Sd (Pain)/8d – SO	L1 multiple	Yes	Root	Total (+RR)	4 mo/Excellent
20	Cecchi et al. 2007[4]		Cs Sd (Paresthesia in both legs)/2 mo	L3-L4	Yes	Root (intra)	Total	DH/Excellent
21	Chung JY et al. 2008[8]	58/F	NM/NM	L2	Yes	NM	Total	NM
22	Chung JY et al. 2008[8]	59/F	NM/NM	L4	Yes	NM	Total	NM
23	Chun et al. 2010[6]	74/F	Cs Sd (Sciatic pain)/3–4 yrs	Below L4	Yes	Root (intra)	Total	DH/Excellent
24	Khalatbari et al. 2011[13]	NM/NM	Cs Sd (Sciatica and back pain)/SO	NM	NM	NM	NM	NM
25	Khalatbari et al. 2011[13]	NM/NM	Cs Sd (Sciatica and back pain)/SO	NM	NM	NM	NM	NM
26	Popescu et al. (presented case)	60/F	Cs Sd (Back pain)/4d – SO	L4	Yes	Root	Total	2 yrs/Excellent

Cs: compression, d: day/s, DH: discharge from hospital, FT: filum terminale, Gait dis: gait disturbance, ICHT: intracranial hypertension, IR: incomplete recovery, mo: month/s, MRI: magnetic resonance imaging, NM: not mentioned, RR: nerve root resection, S: sensory, SAH: subarchnoid hemorrhage, Sd: syndrome, SM: sensorimotor, SO: sudden onset, Sph-dys: sphincter dysfunction, wk/s: week/s, yr/s: year/s.

as a well-defined lesion with mixed signal intensity on both T_1- and T_2-weighted images (presumably indicating the presence of mixed subacute-chronic intratumoral hemorrhage), often circumscribed by a hypointense ring on T_2-weighted images (macrophage uptake of hemosiderin).[4,8,10] All reviewed cases after 1990, including ours, had an MRI suggestive picture.

IV. Pathological diagnosis

Cavernomas in the cauda equina are benign vascular neoplasias, encapsulated, up to 3 cm, most likely originating from the abnormal development of periradicular vessels, which could explain their close adherence to the nerve roots.[9–11] Some of them (5 of the reviewed cases) arise within the sheets of one nerve root, which become encapsulated within the lesion.

Intratumoral bleedings, either small and repeated or isolate and abrupt, confirmed by histopathological examination, as in our case, may result in progressive or sudden neurologic deterioration or SAH, with subsequent subacute-chronic or acute onset, respectively.[8]

V. Therapeutical management and patient outcome

Surgery is the treatment of choice.[3] Total excision was possible in all cases, including ours, with careful microsurgical technique for sparing the nerve root(s), where the tumors were closely attached to them. In the above mentioned 5 cases with intraroot origin of lesions, the tumoral mass was excised along with the attached thin nerve root. All reviewed patients had excellent postoperative recoveries, excepting the two cases from the 60's (Table 1, cases 1 and 2), which both had previous sphincter dysfunction and the 7 cases with no mention about the outcome in the abstract/article.

Conclusion

Even so rare, cavernoma of the cauda equina should be suspected when local compression syndrome but also SAH or ICHT syndromes are present because, once diagnosed, with the precious help of MRI, the tumor is perfectly curable. Further data are expected to complete the profile of this uncommon vascular lesion.

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Conflicts of Interest Disclosure

The authors declare that they have no competing interests and no financial support.
The patient was informed, approved and signed an informed consent form.

References

1) Ahn CH, Ji C, Cho KK, Lee KJ, Lee GS, Yoon SH, Choi CR: Two cases of cavernous hemangioma of the cauda equina: case report. *J Korean Neurosurg Soc* 21: 739–745, 1992
2) Calonje E, Flecher CMD: Vascular tumors, *in* Flecher CMD (*ed*): *Diagnostic Histopathology of Tumors.* Philadelphia, Churchill Livingstone, 2007, pp 41–81
3) Caroli E, Acqui M, Trasimeni G, Di Stefano D, Ferrante L: A case of intraroot cauda equina cavernous angioma: clinical considerations. *Spinal Cord* 45: 318–321, 2007
4) Cecchi PC, Rizzo P, Faccioli F, Bontempini L, Schwarz A, Bricolo A: Intraneural cavernous malformation of the cauda equina. *J Clin Neurosci* 14: 984–986, 2007
5) Choi CY, Min BK, Suk JS: A case of cavernous hemangioma of the cauda equina. *J Korean Neurosurg Soc* 25: 2103–2106, 1996
6) Chun SW, Kim SJ, Lee TH, Koo HS: Intra-root cavernous angioma of the cauda equina: a case report and review of the literature. *J Korean Neurosurg Soc* 47: 291–294, 2010
7) Chung DY, Shin YH, Sung JK: A case of spinal intradural extramedullary cavernous angioma. *J Korean Neurosurg Soc* 38: 74–76, 2005
8) Chung JY, Lee JJ, Kim HJ, Seo HY: Characterization of magnetic resonance images for spinal cord tumors. *Asian Spine J* 2: 15–21, 2008
9) Crispino M, Vecchioni S, Galli G, Olivetti L: Spinal intradural extramedullary haemangioma: MRI and neurosurgical findings. *Acta Neurochir (Wien)* 147: 1195–1198; discussion 1198, 2005
10) Er U, Yigitkanli K, Simsek S, Adabag A, Bavbek M: Spinal intradural extramedullary cavernous angioma: case report and review of the literature. *Spinal Cord* 45: 632–636, 2007
11) Falavigna A, Righesso Neto O, dos Santos JA, Ferraz FA: Cavernous angioma of the cauda equina: case report. *Arq Neuropsiquiatr* 62: 531–534, 2004

12) Jin YJ, Chung SB, Kim KJ, Kim HJ: Spinal intradural extramedullary cavernoma presenting with intracranial superficial hemosiderosis. *J Korean Neurosurg Soc* 49: 377–380, 2011
13) Khalatbari MR, Hamidi M, Moharamzad Y, Taheri B: Cauda equina cavernous angioma presenting as acute low back pain and sciatica—a report of two cases and literature review. *Neuroradiol J* 24: 636–642, 2011
14) Kim KS, Ahn BJ, Han DH: Cavernous hemangioma of the cauda equina. *J Korean Neurosurg Soc* 30: 662–665, 2001
15) Lin DDM, Abdalla W: Neuroimaging of cavernous malformations, *in* Rigamonti D (*ed*): *Cavernous Malformations of the Nervous System*. Cambridge, Cambridge University Press, 2011, pp 49–64
16) Mitchell RN, Schoen FJ: Blood vessels, *in* Kumar V, Abbas AK, Fausto N, Aster JC (*eds*): *Robbins and Cotran Pathologic Basis of Disease*. Philadelphia, Elsevier Saunders, 2010, pp 487–528
17) Miyake S, Uchihashi Y, Takaishi Y, Sakagami Y, Kohmura E: Multiple cavernous angiomas of the cauda equina. Case report. *Neurol Med Chir (Tokyo)* 47: 178–181, 2007
18) Park JC, Chung CK, Kim HJ: Intradural-extramedullary spinal cavernous angioma as a cause of subarachnoid hemorrhage. *J Korean Neurosurg Soc* 34: 174–176, 2003
19) Rosai J: *Rosai and Ackerman's Surgical Pathology*. Philadelphia, Mosby-Elsevier, 2011
20) Sheth S, Lai CK, Dry S, Binder S, Fishbein MC: Benign vascular tumors and tumor-like proliferations. *Semin Diagn Pathol* 25: 1–16, 2008

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