

Clinical Study

Giant nondural-based cauda equina meningioma with multiple cysts

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Summary

A very rare case of a giant nondural-based cauda equina meningioma with multiple cysts was presented. Spinal meningioma most commonly occurs in the thoracic or cervical region and typically adheres to the dura. Only six cases of nondural-based meningioma have been reported in English literature. All occurred in the cauda equina region. These patients were predominantly female and younger than those with typical intraspinal meningioma.

A 46-year-old woman had a 4-year history of lower back pain and right leg pain. Progressive weakness of both lower extremities occurred. Magnetic resonance imaging revealed a giant cauda equina tumor with multiple cysts from T₁₂ to L₄. Following laminectomies from T₁₁ to L₅ and intradural exposure, the tumor was found to be draped loosely by the roots of the cauda equina and attached to a root without any firm connection with dura mater. Complete removal of the tumor was achieved after microdissection of arachnoid and sacrifice of an involved rootlet of the cauda equina. The appearance of tumor was that of a typical neurilemmoma. However, histological and immunohistochemical analyses were consistent with meningioma.

Nondural-based intraspinal meningiomas are very rare, particularly a giant tumor with multiple cysts as our presenting case. All of the cases previously reported, including our case, have been located in the cauda equina region. Most of the patients were female and were young, suggesting that the nondural-based cauda equina meningiomas are age- and sex-related. An accurate preoperative and operative diagnosis are difficult. Care must be taken in the management of cauda equina tumors resembling neurilemmoma which may in fact represent meningioma, particularly in the younger female.

Introduction

Cauda equina meningioma is especially rare. An overall incidence of 2.3% of meningiomas in the lumbosacral region was reported in an analysis of 705 spinal meningiomas [1–4]. Meningiomas typically adhere to the dura and, to date, only six cases of nondurally attached intraspinal meningioma have been reported in English literature [5–9]. Moreover, giant nondural-based cauda equina meningioma has been previously reported only once, in a 10-year-old girl. The authors report an unusual case of giant nondural-based meningioma with multiple cysts in the cauda equina region in a 46-year-old woman, which was completely excised under the microscope.

Case report

Presentation

A 46-year-old woman initially experienced lower back pain and right leg pain 4 years previously. Thereafter, she experienced intermittent pain episodes of short duration. About 2 years prior to presentation, she felt

weakness and numbness over both lower extremities. She was examined at a local hospital and conservative treatment with rehabilitation was initiated. Because of increasing lower back pain and weakness of both lower extremities, she came to our hospital in November 2003. She was unable to walk without the aid of a cane because of severe weakness of both lower extremities.

Neurological examination

Motor examination showed weakness of bilateral hip flexion, knee extension and knee flexion with muscle power of grade 4 and weakness of left ankle dorsiflexion, ankle plantar flexion, toes extension and toe flexion with muscle power of grade 3. The right foot and toes could not dorsiflex and plantar flex (muscle power grading 0). Bilateral knee and ankle jerks were absent. Hypoesthesia was noted below the L₁ level. However, there was no bladder or bowel dysfunction.

Imaging examination

Myelogram revealed a complete block at the L₄ level with a cup appearance (Figure 1). Magnetic resonance imaging



Figure 1. Myelogram showing a complete block at L₄ level with a cup appearance.

(MRI) disclosed a giant cauda equina tumor from the upper level of T₁₂ to lower level of L₄ (Figure 2). The tumor revealed low signal intensity on T₁-weighted image, increased signal intensity on T₂-weighted image, and increased signal intensity with multiple cysts on Gadolinium-enhanced T₁-weighted image.

Operation

Through a posterior midline incision, the T₁₁–L₅ laminectomy was done. A giant encapsulated tumor, 11 cm in length, was found intradurally. The tumor occupied the whole spinal canal and was draped loosely by the

roots of the cauda equina (Figure 3a). After internal debulking, the tumor was carefully dissected from the nerve root and arachnoid. No firm connection with dura mater or cauda equina was found at its lower part, but tight adhesions to nerve roots were found at its upper part, near the conus medullaris (Figure 3b). One rootlet of the cauda equina was involved deeply in the tumor. The appearance of the tumor was that of a typical neurilemmoma. Careful dissection was performed using microsurgical techniques. Complete resection of tumor, including the sacrifice of an involved rootlet of cauda equina, was achieved. Discrete investigation of the remaining distal part of the sacrificed rootlet confirmed that the involved part was the right L₅ root.



Figure 2. Sagittal MR images showing a giant cauda equina tumor with multiple cysts. (a) T₁-weighted image, (b) T₂-weighted image, (c) gadolinium-enhanced T₁-weighted image.

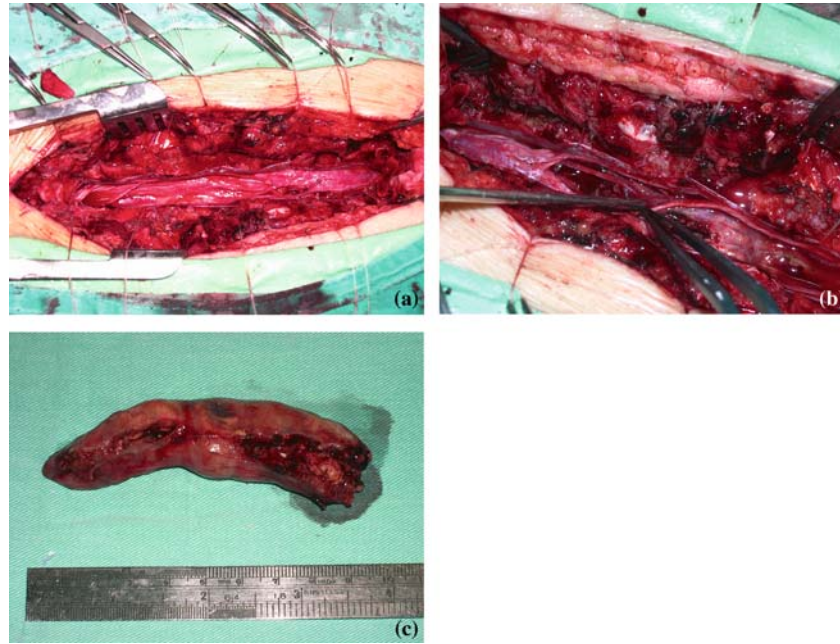


Figure 3. Intraoperative finding: (a) tumor was draped loosely by the nerve roots, (b) one root of the cauda equine was involved in the tumor, (c) gross pathology.

Pathological examination

Gross pathology demonstrated a 11 cm in length well-circumscribed mass (Figure 3C). Microscopically, the specimen demonstrated spindle and polyhedral cells forming occasional whorls typical of meningiomas (Figure 4). Immunohistochemical examination showed the tumor to be positive for epithelial membrane antigen (EMA) and vimentin. It was negative for glial fibrillary acidic protein (GFAP), neuron specific enolase (NSE), and neurofilament protein (NFP). It was also negative for both progesterone and estrogen receptors.

Postoperative course

The postoperative course was uneventful. The severe low back pain and numbness of legs were remarkably decreased. There was slow and gradual improvement of the muscle weakness of the lower extremities. No deterioration of bladder and sphincter function was noted after surgery. She were able to walk for at least hours alone without the aid of a walker 3 months after surgery. No tumor recurrence was found in the MRI 6 months postoperatively (Figure 5).

Discussion

An accurate preoperative and intraoperative diagnosis was difficult in this case. The preoperative diagnosis was cauda equina neurilemmoma or ependymoma. The diagnosis during operation suggested a neurilemmoma because the tumor was freely mobile and a nerve was seen entering and emerging from the tumor. However, histological study showed it to be a meningioma.

Intraspinal meningiomas are almost always adherent to the dura. These tumors are most prevalent in the

thoracic regions, followed in frequency by the cervical and lumbar regions [1,3,4,10]. Nondurally attached intraspinal meningiomas are very rare and only four

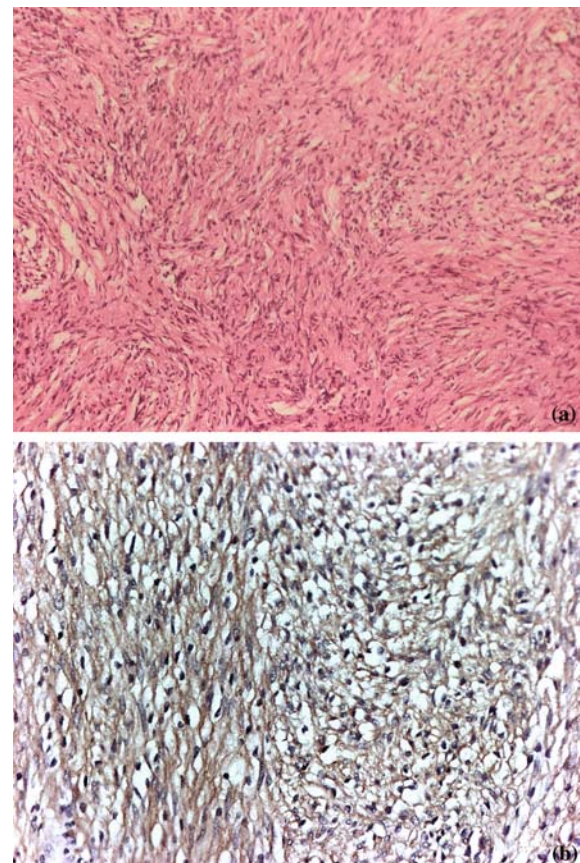


Figure 4. Photomicrograph showing a meningioma: (a) H&E, original magnification x100, (b) positive immunohistochemical staining for epithelial membrane antigen (x200).

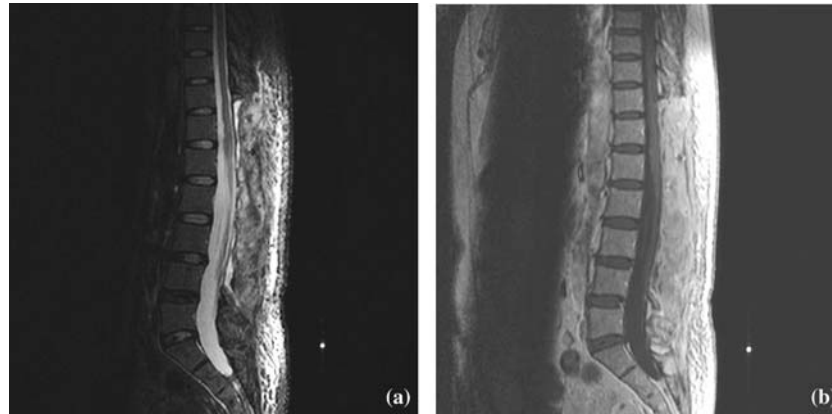


Figure 5. MRI 6 months after surgery. No tumor mass is seen: (a) T₂-weighted image, (b) Gadolinium-enhanced T₁-weighted image.

cases have been reported in detail [5–8]. Table 1 summarizes previously reported nondural-based meningiomas of the cauda equina. Complete excision is recommended because these tumors are easily dissected free from the dura and cauda equina. However, neurilemmomas in the cauda equina were usually excised incompletely because complete removal had the risk of sacrificing many nerve roots [11].

Meningiomas usually occur in patients older than 40 years of age. Most of the reported nondural-based meningioma cases are female and their ages are relatively younger. The mean age of the reported six patients with nondural-based cauda equina meningiomas was 21 years (range, 10–32 years). The mean age of the patients with intraspinal meningiomas was 56 years (range, 13–82 years) [4]. Mizutani et al. [7] reported a case of nondurally attached meningioma in the lumbosacral region in a 20-year-old woman, possibly induced by orally ingested sex steroid pills. They suggested that the high dose of orally ingested sex steroid pills had enlarged the tumor in their reported patient and caused it to become symptomatic, which might explain the unusually young onset age. Furthermore, there was a higher occurrence of spinal meningiomas in woman in the thoracic region (125 of 144 patients, 87%) and the cervical region (16 of 26 patients, 61%). Meningiomas of the lumbar region (4 patients) were found with equal frequency in men and women [4]. Interestingly, all the nondural-based cauda equina meningiomas were found

in women, except in one case. Therefore, the nondural-based cauda equina meningioma may be age- and sex-related.

Meningiomas arise from the cellular elements of the arachnoid. Typically, meningiomas are adherent to dura. Exactly why meningioma involving the cauda equina displays characteristics of easy separation is uncertain. The cauda equina does not contain cellular elements of the arachnoid, except in the nerve root sleeve. Thomas et al. [8] reported the possibility that such tumors arise from the nerve root sleeve. However, in their reported case, the tumor was located far from the nerve root sleeve. They also suggested the possibility of secondary attachment to the cauda equina after the loss of dural attachment as a possible explanation. Mizutani et al. [7] supported the secondary attachment hypothesis. They also speculated that one other possible explanation of migration of cellular elements was during prenatal development. Bain and Shnitka contended that Schwann cells may differentiate to meningocytes to form a meningioma under suitable conditions [12]. The current authors support the differentiation or migration hypothesis, first because nondural-based meningiomas occur in the young female, second because such tumors arise far from the root sleeve, and third because such tumors can grow and produce a giant tumor, as in our case and the case reported by Dubois et al. [5], but they have arachnoid-formed capsules allowing for an easy separation from dura mater.

Table 1. Reported cases of nondural-based intraspinal meningiomas

Authors (Year)	Age (year)	Sex	Location	Pathology	Follow-up (period)
Thomas et al. (1989)	23	F	T ₁₂ -L ₁	Angiomatous meningioma	–
Holtzman et al. (1996)	32	F	L ₄	Clear-cell meningioma	No tumor (1 m)
Dubois et al. (1998)	10	F	L ₁₋₄	Clear-cell meningioma	Small enhancing lesion (1 m) Recurrence (6 m) No tumor (unknown)
Mizutani et al. (2002)	20	F	S ₁	Transitional meningioma	No tumor (61 m)
Payano et al. (2004)	19	F	L ₃₋₄	Clear-cell meningioma	No tumor (52 m)
Authors (2004)	46	F	T ₁₂ -L ₄	Meningotheliomatous meningioma	No tumor (6 m)

Conclusion

The authors report a very unusual case of a giant nondural-based cauda equina meningioma with multiple cysts. An accurate preoperative and intraoperative diagnosis was difficult. A giant cauda equina tumor resembling a neurilemmoma should be aggressively managed, particularly in a younger woman, because the possible diagnosis of nondural-based meningioma raises the potential of complete tumor removal without damage of nerve. All nondural-based meningiomas, including the current case, have arisen from the lumbosacral region and are located far from the nerve root sleeve. Most of the cases occur in females and are relatively young. These interesting phenomena lead to the possibility of cellular migration or differentiation for the explanation of nondural attachment of cauda equina meningiomas.

References

1. Levy WJ, Bay J, Dohn D: Spinal cord meningioma. *J Neurosurg* 57: 804–812, 1982
2. Nittner K: Spinal meningiomas, neurinomas and neurofibromas and hourglass tumors. In: Vinken PJ, Bruyn BW (eds.) *Tumors of the Spine and Spinal Cord: Part II. Handbook of Clinical Neurology*, Vol. 20. North Holland, Amsterdam, 1976, pp. 177–322
3. Roux FX, Nataf F, Pinaudeau M et al.: Intraspinal meningiomas: review of 54 cases with discussion of poor prognosis factors and modern therapeutic management. *Surg Neurol* 46: 458–464, 1996
4. Solero CL, Fornari M, Giombini S et al.: Spinal meningiomas: review of 174 operated cases. *Neurosurgery* 125: 153–160, 1989
5. Dubois A, Sevely A, Boetto S et al.: Clear-cell meningioma of the cauda equina. *Neuroradiology* 40: 743–747, 1998
6. Holtzman RNN, Jourmark SC: Nondural-based lumbar clear cell meningioma case report. *J Neurosurg* 84: 264–266, 1996
7. Mizutani J, Fukuoka M, Tsubouchi S et al.: A rare case of lumbosacral meningioma. Nondural attachment and possible enlargement by orally administered sex steroid. *Spine* 27: E377–E381, 2002
8. Thomas HK, Chan KH, Kirpal SM et al.: Spine meningioma arising from a lumbar nerve root. *J Neurosurg* 70: 646–648, 1989
9. Payano M, Kondo Y, Kashima K, Daa T, Yatsuka T, Kida H, Nakayama I, Yokoyama S: Two cases of nondura-based clear cell meningioma of the cauda equina. *APMIS* 112: 141–147, 2004
10. Helseth A, Mork SJ: Primary intraspinal neoplasms in Norway, 1995 to 1986: a population-based survey of 467 patients. *J Neurosurg* 71: 842–845, 1989
11. Kagaya H, Abe E, Sato K et al.: Giant cauda equina schwannoma. A case report. *Spine* 25: 268–272, 2000
12. Bain GO, Shnitka TK: Cutaneous meningioma (psammoma): report of a case. *Arch Dermatol* 74: 590–594, 1956

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