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Spinal intramedullary teratoma with exophytic components: report of two cases and review of the literature

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Abstract True intramedullary teratoma is an extremely rare tumor, with only seven cases reported in the literature. The authors present two personal cases of spinal intramedullary cystic teratoma of the conus medullaris with exophytic growth and some unusual aspects. Their cases are unusual not only because they were diagnosed by MRI but also because the monitored microsurgical removal of the tumor was apparently total, with excellent results, in clinical and neuroradiologic terms. We recommend early radical surgery whenever possible, when the patient's neurological status is still good. To prevent traumatizing adjacent spinal cord for cases in which the teratoma tightly adheres to the functional neurological tissue of the spinal cord with no cleavage plane, we do not recommend an aggressive surgical attempt. Because of the mostly benign nature of this disease, the symptomatic recurrence of such incompletely resected mature teratomas is slow and may eventually require a second surgical procedure. The relevant literature is also reviewed.

Keywords Intramedullary tumor · Spinal cord tumor · Teratoma

Introduction

The current and most adequate classification states that spinal teratoma is a mass composed of derivatives from all three primitive germ layers that distinguishes between mature and immature forms based on the degree of differentiation. However, the presence of only two germinal components does not necessarily rule out this diagnosis [1]. This classification does not include epidermoid and dermoid cysts or sacrococcygeal teratoid or enterogenous cysts [2]. The intramedullary variety of teratoma is extremely rare and has been reported only seven times during the last 30 years [3, 4, 5, 6, 7, 8, 9]. We present two new cases of intramedullary spinal teratoma (IST) of the conus medullaris with exophytic components diagnosed by magnetic resonance (MR) imaging and successfully treated with monitored microsurgery. The other seven cases in the literature are also discussed.

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Case reports

Case 1

This 45-year-old woman presented with a 2-year history of lumbago, a 1-year history of dysthesia, bilateral tingling in the lower extremities, and an 8-month history of mild paraparesis. During a 4-month period, she developed bladder incontinence and the inability to ambulate. An MR image of the spine revealed an expansive inhomogeneous lesion in the conus medullaris (Fig. 1).

Somatosensory potentials were recorded during the surgical procedure. A midline incision was made, centered at the level of the lesion. After laminectomy exposing one additional vertebra above and below the lesion (T11–L3), intraoperative ultrasonography was used to localize solid and cystic areas of the teratoma and select respective sites for the myelotomy. High-powered microscopic magnification allowed localization of the dorsal median sulcus. Once exposed, the conus medullaris and the cauda equina appeared normal on the surface, although swollen and enlarged pial arteries were visualized. The surgical field was gently opened progressively over the entire length of the lesion, finally exposing the poles of the lesion. A voluminous cystic mass was revealed within the conus extending from T12 to L2 and displacing the roots of the cauda equina.

The tumor was seen to be intramedullary in the conus medullaris region and presented a small caudal exophytic component that displaced but did not infiltrate the root of the cauda equina. The filum terminale was displaced laterally without any relation to the tumor, and thus its resection was not necessary. The cyst was encountered, and 7 ml of thick, yellow, sebaceous fluid was removed. The walls of the cyst appeared as a glistening membrane. The remainder of the cystic mass and an adjacent reddish-tan mass were then completely resected. The dissection and total removal of the tumor were achieved because it was possible under high microscopic magnification to identify a cleavage plane between the soft inhomogeneous mass and the nervous tissue.

Postoperatively, the patient did quite well, regaining bladder function within 3 weeks, and was able to walk independently within 3 days. She was discharged after 12 days, with mild super-

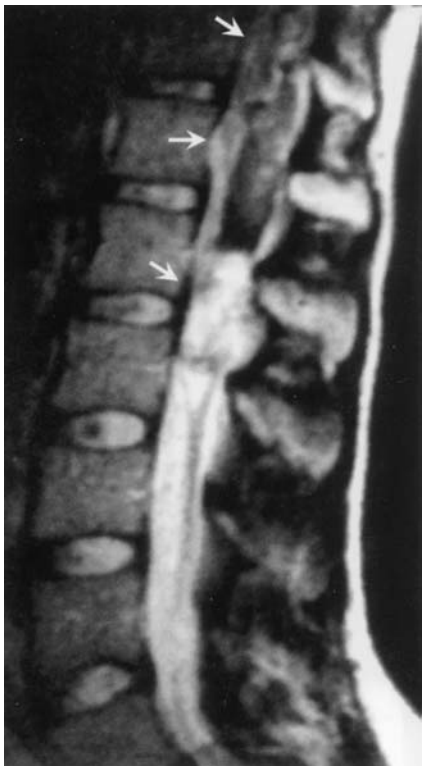

Fig. 1 Midline sagittal MR image of the spine reveals an expansive inhomogeneous lesion in the conus medullaris with high intensity fat, fluid component, and lower intensity tissue (*arrows*). Histological examination demonstrated a mature tridermal teratoma

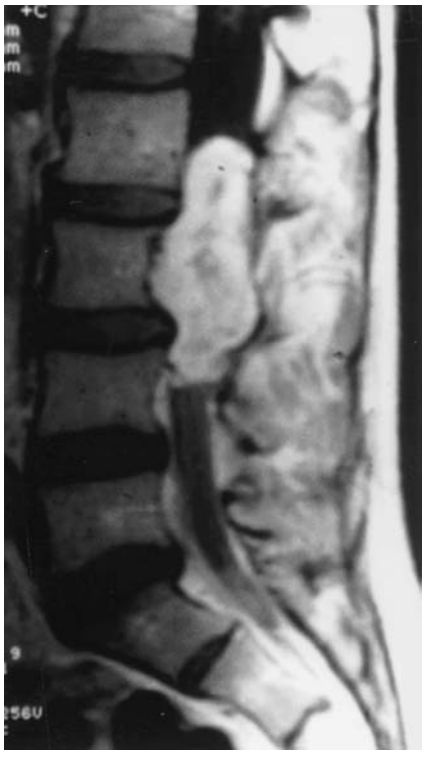

Fig. 3 Midline sagittal MR image (T1-weighted) showing an expansive intra- and extramedullary lesion between L2 and L4 with various signal characteristics of both solid and cystic components

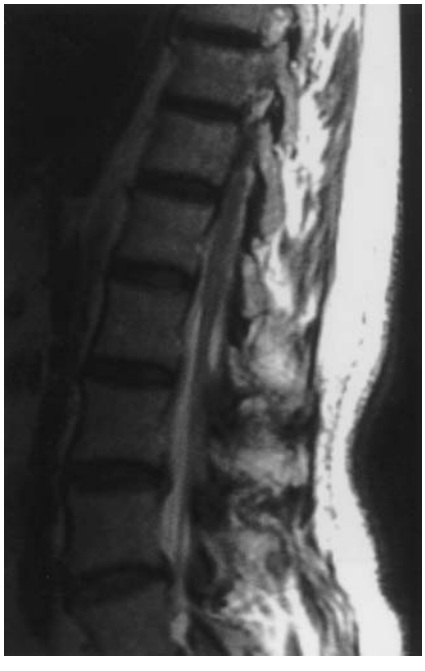

Fig. 2 Follow-up MR image 1 year after total removal of the teratoma shown in Fig. 1

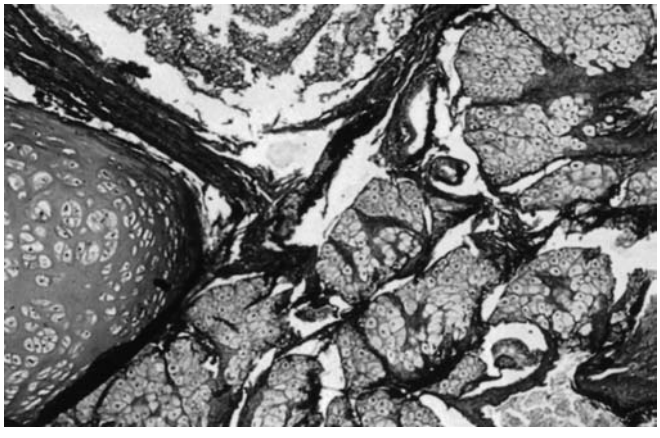

Fig. 4 Histological photomicrograph demonstrating multiplicity of tissue derived from three germ layers (H&E ×40) with heterogeneous, fully differentiated components including fibrofatty connective tissue, well-formed mucosa, striated muscle, and cutaneous epithelium (mature tridermal teratoma)

ficial hypesthesia in a saddle distribution. Follow-up MR imaging 1 year postoperatively showed no evidence of residual tumor or recurrence (Fig. 2).

Histological examination demonstrated a mature tridermal teratoma with heterogeneous, fully differentiated components including fibrofatty connective tissue, well-formed mucosa, striated muscle, and cutaneous epithelium.

Table 1 Intramedullary spinal teratomas previously reported in the literature (seven cases)

Case no.	Author	Age (years), sex	Duration of symptoms	Site of tumor	OP/laminectomy	Histological/intraoperative finding, associated lesions	Outcome
1	Lemmen 1951 [6]	11, M	3 years	T12–L1	T12–L2	Diplomyelia, malignant (bladder incontinence)	Incomplete recovery
2	Slooff 1964 [9]	?	?	L1–L3	T12–L3	Complete removal	Recovered
3	Eneström 1977 [3]	36, M	4 years	D11–L1	D11–L1	Homogeneous, no cystic components, incomplete removal	Incomplete recovery
4	Padovani 1982 [8]	21, F	5 months	cervical (C6–T1)	C6–T1	Incomplete removal	Incomplete recovery
5	Giacomini 1986 [4]	19, M	?	L1–L2	L1–L3	Complete removal	Recovered
6	Nicoletti 1994 [7]	47, M	8 years	T12–L2	T11–L4	Incomplete removal	Recovered
7	Hader 1999 [5]	16, F	2 months	T7–T9	T8–T11	Diastematomyelia, scoliosis, incomplete removal	Recovered

Case 2

This 20-year-old male gave a history of lumbar vertebral pain in the L4–S1 dermatomes radiating to the lower limbs. He developed progressive paraparesis and complete urinary retention during the ensuing 20 days, necessitating a suprapubic bladder catheter. Magnetic resonance imaging of the spine revealed a lesion between L2 and L4 with various signal characteristics of both solid and cystic components (Fig. 3). An L1–L4 laminoplastic laminotomy was performed, allowing complete removal of an inhomogeneous, soft mass with a cleavage plane between the lesion within the conus and the nervous tissue.

Histological examination showed that the tumor was a mature tridermal teratoma. The lesion was complex, consisting of fibromuscular tissue, mature fat, and cartilage surrounded by smooth and striated muscle and neural tissue. Mucous and serous glands were also seen. Collections of ependymal cells and irregular blood vessels were scattered throughout the mass (Fig. 4).

The postoperative course presented no problems, apart from slight urinary incontinence, which cleared up within 4 weeks. The patient was discharged after 14 days without a suprapubic bladder catheter and with mild hypesthesia in the right L5–S1 dermatomes, which resolved within 3 months after surgery.

Discussion

The origin of IST has been the subject of much speculation, and several theories have been offered in attempts at explanation [1, 10, 11]. The most accepted one is that IST arises from primordial germ cells misplaced from the primitive yolk sac into the dorsal midline during their normal migration from yolk sac to gonadal ridges. A dysembryogenic origin of IST is suggested by the association between dysraphic processes and teratomas and the pluripotential nature of the developing caudal spinal cord, which can potentially form all three germ layers, so that most central nervous system teratomas typically occur in midline structures and could derive from pluripotential cell rests at sites of early neural tube closure [1, 10]. Preoperative diagnosis of spinal teratoma is not easy. Magnetic resonance imaging cannot determine with certainty the differential diagnosis between teratomas and other intramedullary lesions. Signal characteristics compatible with heterogeneous solid and cystic components and the presence of fatty tissue associated with congenital abnormalities (myelomeningocele, spina bifida, dermal sinus, split cord malformation, or vertebral anomalies) enable the preoperative diagnosis of teratoma [12]. The finding of intramedullary teratoma in association with other vertebral abnormalities such as diastematomyelia represents an extremely rare constellation [5, 13].

The seven previous cases of IST in the literature are summarized in Table 1. In all of them, both cystic and solid components were found. Only one was found in the cervical region [8]. The other six cases were intramedullary in the cauda/conus medullaris region. Motor disturbances and pain in combination with bladder dysfunction were the most frequent symptoms. Based on these seven cases, the natural history of the disease appears to be prolonged. The patients experienced symptoms from 2 months to 8 years, although rapid decompensation with worsening of neurological status was frequently seen a few weeks or months preoperatively.

Technological advances in MR imaging contributed to the early detection of the teratoma and the definitive diagnosis in our patients. Complete removal of the tumor was achieved in two of seven other cases [4, 9]. In five, the tumors were adherent to the spinal cord and the roots of the cauda equina. Thus they were not completely resectable, and the remaining endodermal or ectodermal components sprouted again [3, 5, 7, 8, 11]. However, the growth of these components was very slow and associated with prolonged, symptom-free course.

In our cases, complete removal of the IST was possible. The role of surgical therapy appears to be limited when the tumors involve the cauda equina, especially if the involved roots have normal neurological function. Local decompressive procedures such as tumor debul-

king or cyst evacuation in conjunction with evoked potential monitoring and laminectomy may yield many pain-free years and stabilize neurological deterioration. The symptomatic recurrence of incompletely resected mature teratomas is slow and may eventually require a second surgical procedure. In extremely rare cases of immature teratomas with malignancy signs, adjuvant chemotherapy postoperatively may change the naturally aggressive behavior of the disease and achieve persistent, complete remission. However, the value of chemotherapy in such treatment has not been defined [14].

In conclusion, intramedullary teratoma of the conus medullaris is rare and should be considered in the differential diagnosis of masses involving the cauda equina. The teratomas in our two cases were mature, because well-differentiated derivatives of all three germinal layers were present. We recommend early radical surgery whenever possible, when the patient's neurological status is still good. To prevent traumatizing the adjacent spinal cord when the teratoma is tightly adherent to functional neurological tissue of the spinal cord without a cleavage plane, we do not recommend aggressive surgery. Because of the benign nature of the disease, the symptomatic recurrence of such incompletely resected mature teratomas is slow and may eventually require second surgical procedures.

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