# INTRAMEDULLARY SPINAL TERATOMA

## A rare condition with a good outcome

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Approximately one-third of spinal tumors in childhood[1] are intramedullary neoplasms[1] and up to 80% are gliomas[2]. The occurrence of teratomas in the spine is extremely rare[3]. Except for in the sacrococcygeal region, teratomas constitute less than 0.5% of all intraspinal tumors. It is believed that they arise from multipotential germinal cells misplaced in the early embryonic development. It was also proposed that teratomas represent abortive "twin" formation cystic component ependymal diverticula of the central canal of the spinal cord[4,5]. Teratomas may be intra- or extramedullary and usually fill the spinal canal. Some teratomas have been reported in association with split cord malformations and other developmental spinal abnormalities, like myelomeningoceles[3,6,7]. The tumor may be partially solid or wholly cystic. Intramedullary teratomas usually are mature and regarded as benign. Most patients often present myelopathic symptoms with insidious onset[1,2]. Surgical resection provides good outcome and tumor recurrence is uncommon[2].

We present a case of a child with progressive neurological and painful symptoms whose diagnostic investigation revealed a spinal teratoma and whose outcome was favorable after surgical treatment.

### CASE

A 5-years-old girl, born from healthy and nonconsanguineous parents, presents with normal development, weakness in lower limbs associated with abdominal and back pain lasting two months. She was admitted at Clinics Hospital of São Paulo University Medical School in 2008. The symptoms were slowly progressive. The general physical examination was unremarkable. At the admission the patient was able to walk without aids, but always with her legs inflected. Neurological examination showed bilateral paraparesis of the lower limbs (power grade 4), bilateral lower extremities hyperreflexia and flexor plantar reflexes bilaterally. Sensory examination and sphincters control were normal. No signs of dysraphic congenital malformations were present.

MRI showed a well-delineated enlargement of the spinal cord with high intramedullary signal on T2-weighted images (Fig 1A, 1B and 1C) and heterogeneous enhancement post-contrast T1-weighted image (Fig 1D) extending from T5–T7 vertebral level. The patient underwent surgery for tumor removal. After a dorsal median incision at the desired level and a T5–T8 laminotomy were performed, and dura-mater was opened, a T5–T7 intramedullary solid-cystic tumor containing irregular surface, purple and brown colors was totally removed microsurgically. Hair was found inside the cystic component of the tumor. The histopathological examination revealed a mature teratoma. There was no evidence of malignant components. The postoperative period was uneventful and the patient remained stable.

### DISCUSSION

Spinal tumors comprise 5–10% of the tumors of central nervous system in children. They have annual incidence at around 1 per million[8,9]. Although uncommon, spinal cord neoplasm should be considered the differential diagnosis in patients presenting back or radicular pain associated with neurological deficits[2].

Extradural tumors account for 60% of the spinal cord neoplasms, and 30% of the tumors are intradural. The 10% remaining have intradural and extradural concomitant components.

Within the intradural space, tumors can be present adjacent to (extramedullary) or within (intramedullary) the spinal cord parenchyma[2]. Astrocytomas represent more than 70% of the intramedullary spinal cord neoplasms in childhood and ependymomas 15–35%[2]. Hemangioblastomas, gangliogliomas, lipomas, developmental tumors, and metastatic intramedullary tumors are rare. Teratomas are specially unusual, their incidence range from 0.2–0.5% of

**TERATOMA INTRAMEDULAR: BOM RESULTADO EM CASO INCOMUM**

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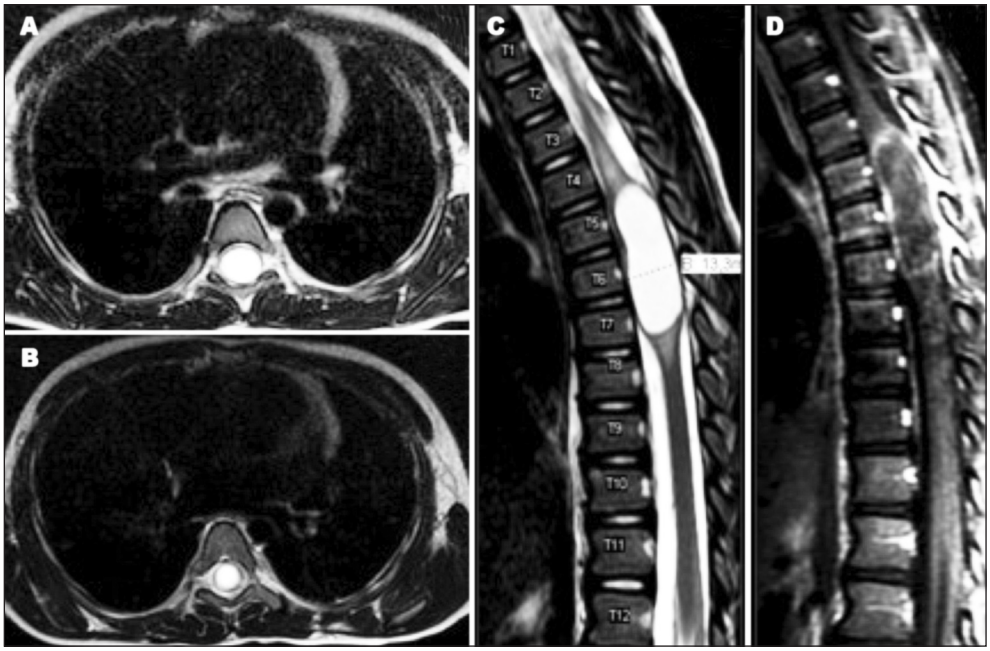


*Fig 1. MRI showing high signal lesion on T2, extending from T5–T7 [A, B and C]. T1 after gadolinium shows heterogeneous enhancement [D].*

all spinal cord tumors[1,9]. In a retrospective study of 22 children with intramedullary mass lesions excluding the developmental tumors associated with spinal dysraphism the authors found only one teratoma, nine astrocytomas and ependymomas[10]. Sloof et al. reported only two teratomas out of a total of 301 intramedullary spinal cord lesions[11].

Tapper and Lack published 254 teratomas in 21 year-old patients or younger diagnosed and treated during a period of 54 years. The tumor arose in the central nervous system in nine patients (3.5%), and just four (1.5%) were placed into the spinal canal, excluding those localized in the sacrococcygeal region which were 40% of the total teratomas[12]. In another review including 18 cases of spinal intramedullary teratoma, the most frequent place of the tumor was the conus medullaris (70.6%). There was a male predominance (3:1), and adults were more commonly affected than children. Seven patients (41.1%) had dysraphic congenital malformations associated with the tumor[13].

Habibi et al. reported that the association between myelomeningocele to teratoma is uncommon (4.5%). They supposed that there is a unique origin to both conditions through the incomplete fusion of dorsal structures and that the neural tube defect has been subsequent to the incorporation of primitive cells in the protruded sac[7]. Suri et al. presented 14 cases with slip cord malformations and spinal teratomas in close proximity to diastematomyelia and displayed intramedullary location in only three[6].

In the present patient there was not association with spinal dysraphism, the patient was a young female child and the tumor was located in the thoracic spinal cord, finding much rarer than those generally observed in relation to intramedullary teratomas.

MRI allows the subdivision of tumors in extradural, intradural-extramedullary and intramedullary. The contrast is useful for the differentiation between neoplastic and non-neoplastic lesions. The enlargement of the spinal cord, the high signal on T2-weighted images and the heterogeneous enhancement post-contrast T1-weighted images are common characteristics of the main intramedullary tumors, as observed in the presented case. Preoperative diagnosis of a spinal teratoma is not easy, since the MRI features cannot determine with certainty the differential diagnosis between teratoma and other intramedullary lesions[14].

The diagnosis of teratoma depends on the histopathological identification of the tissues representing the three germinal layers (ectoderm, mesoderm and endoderm)[12] . However, the presence of just two layers does not rule out the diagnosis[4].

Total surgical resection, as in our case, is the best treatment and it provides good outcome. The surgical methods to expose tumors are laminectomy or laminotomy. The latter is preferred for patients below three years of age or for those who require large intraspinal exposure for tumor removal[9]. However, tumor recurrence may occur, probably because of incomplete removal[2,13]. The symptomatic recurrence of incompletely removed teratomas is slow and may eventually require a second surgical procedure[14].The adjuvant therapy is based on the histopatho-

logical features. Radiotherapy is indicated when there is malignant component[15].

In summary, intramedullary teratoma is an extremely rare tumor. The diagnosis is based on the intra-operative and the histopathological examination. Total excision is the primary treatment modality. Radiotherapy is indicated when malignant features are present.

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