

# Primary intradural mesenchymal chondrosarcoma of the spine in a child

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**Abstract** We report a primary intradural mesenchymal chondrosarcoma of the spine in a 3-year-old girl. MRI revealed a markedly enhancing oval mass associated with focal areas of low signal intensity extending from T11 to L1. The lesion was located posterolateral to the right side of the spinal cord, pushing the conus medullaris and cauda equina anteriorly and to the left. The adjacent spinal cord also showed serpiginous areas of flow void. The mass was completely removed. Microscopic examination and immunohistochemical studies confirmed the diagnosis of mesenchymal chondrosarcoma. The patient was free of symptoms after surgery.

**Keywords** Mesenchymal chondrosarcoma · Intradural · MRI · Child

## Introduction

Mesenchymal chondrosarcoma is a primary malignant neoplasm arising from bone and soft tissue with both cartilaginous and undifferentiated round-cell components. Mesenchymal chondrosarcomas in the primary central nervous system are rare. Most of these sarcomas are reported to be intracranial in location [1]. There are only a few series in the literature

concerning intraspinal chondrosarcomas [2, 3]. Radical surgery is the treatment of choice. We report a case of intradural mesenchymal chondrosarcoma of the spine arising from the pia matter at the T11–L1 level in a 3-year-old girl; we also review the literature on this tumor.

## Case report

A 3-year-old girl was admitted to our hospital with a 10-month history of intermittent pain in the right lower limb and 1 month of progressive weakness in the same leg. This was associated with dribbling and soiling. Physical and neurological examinations on admission showed hypoaesthesia of the right lower limb. The muscle strength in this leg was 2/5 and muscular atrophy was observed. Laboratory tests including determination of complete blood count, clotting profiles, electrolytes, and blood chemistry values were all within the normal range.

MRI of the thoracic and lumbar region revealed an intradural mass measuring 3×2×2 cm that extended from T11 to L1. The mass was located posterolateral to the right side of the spinal cord, pushing the conus medullaris and cauda equina anteriorly and to the left. The adjacent spinal cord also showed serpiginous areas of flow void. The mass appeared isointense on T1-weighted (T1-W) images and mildly hyperintense on T2-weighted (T2-W) images with multiple punctate regions of low-signal intensity (Figs. 1 and 2). After intravenous gadolinium administration, a very prominent enhancement of the tumor was observed along with curvilinear enhancement of the adjacent spinal cord (Fig. 3).

Dorsal laminectomy of T11–L2 was performed with the patient in a prone position. A firm, purplish mass with some calcification was found in the intradural space. It was

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**Fig. 1** Sagittal T1-W MR image shows an intradural mass extending from T11 to L1. The tumor appears isointense



observed that the mass arose from the pia mater and adhered to the cord. The mass was located posterolateral to the right side of the cord, pushing the conus medullaris and cauda equina anteriorly and to the left. The mass was rich in vessels and was associated with adjacent meningeal varicosities. The mass was totally resected.

Histopathological examination revealed islands of well-differentiated cartilage surrounded by a diffuse proliferation of small, primitive, undifferentiated mesenchymal cells. There were numerous dilated vascular channels resembling the histological pattern of a hemangiopericytoma (Fig. 4). Foci of calcification were also seen among the islands of cartilage (Fig. 5). Immunohistochemical analysis was positive for S-100 protein in the chondroid areas and for CD-99 and

**Fig. 2** Sagittal T2-W MR image. The tumor appears mildly hyperintense with multiple punctate regions of low signal intensity



**Fig. 3** Sagittal MR image after intravenous gadolinium administration. The tumor shows marked enhancement and the adjacent spinal cord shows curvilinear enhancement. The adjacent spinal cord also shows serpiginous areas of flow void

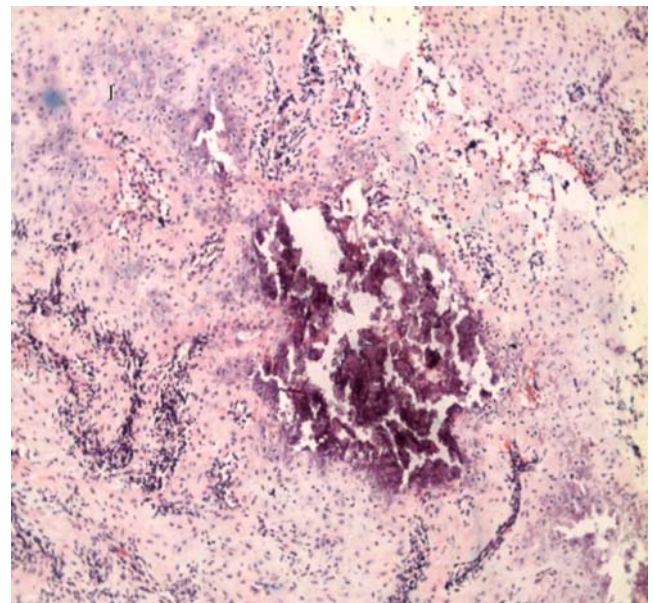


vimentin in the surrounding mesenchymal cells. A diagnosis of mesenchymal chondrosarcoma was made.

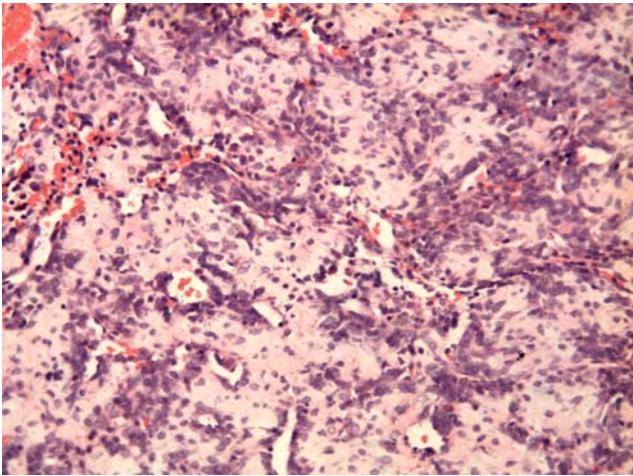
Following surgery, the child was treated with radiotherapy to the primary tumor site. She improved gradually. Two months after surgery and radiotherapy, MRI revealed no intraspinal mass (Fig. 6).

## Discussion

In 1959, Lichtenstein and Bernstein [4] originally described mesenchymal chondrosarcoma as a tumor arising from bone. In 1964, Dowling, [5] described the first case of mesenchymal chondrosarcoma of nonosseous origin. The



**Fig. 4** Histological section shows small round cells surrounding hemangiopericytoma-like vessels (H&E)



**Fig. 5** Histological section shows focal calcification in the cartilage region (H&E,  $\times 20$ )

tumors were characterized by proliferating undifferentiated mesenchymal cells in which scattered islands of well-differentiated cartilage were present.

Extraskeletal mesenchymal chondrosarcoma accounts for 33–50% of total mesenchymal chondrosarcomas, with 50% involving the brain and meninges [6]. Most of these tumors were reported to be intracranial in location. Harsh et al. [1] reviewed 16 mesenchymal chondrosarcomas of the primary central nervous system; only five occurred in intraspinal regions. These tumors have been found more frequently in an extradural location, with the majority having a dural attachment. Intraspinal mesenchymal chondrosarcomas are rare. To our knowledge, more than 20 cases have been described. Only six cases have been reported in children, four in boys and two in girls [1, 2, 7]. The youngest patient was a 3-year-old boy. Involvement of the lower thoracic and upper lumbar vertebrae is more common. However, Ranjan et al. [3] reported a case of an intraspinal tumor in the cervical region without dural attachment. The case we present is unique with respect to both the location of the tumor—intraspinal without dural attachment—and the patient's sex. This is the first case reported in a female toddler.

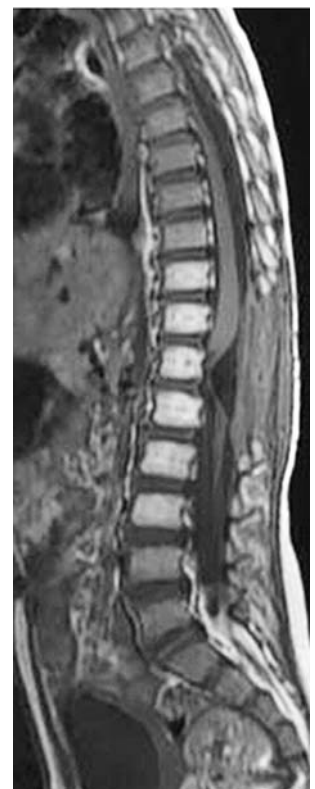
MRI is the imaging modality of choice for intraspinal tumors. Only a few authors have described the imaging features of these intraspinal mesenchymal chondrosarcomas in detail. The tumor generally appears isointense on T1-W images and T2-W images, with marked homogeneous enhancement [2, 8]. This tumor also exhibited heterogeneous features on both T1-W and T2-W images and enhancement similar to that reported by Prevedello et al. [9]. In our patient, the tumor appeared isointense both on T1-W and T2-W images, with marked homogeneous enhancement. However, we also observed clearly visible varices in the surrounding cord. This feature has not previously been described. In this tumor, multiple punctate

regions of low-signal intensity were observed, suggesting calcification that was confirmed by histopathological study. We originally considered the tumor to be a hemangioblastoma in an unusual location because of the presence of marked enhancement and the adjacent spinal cord varices. Although it is more difficult to make a diagnosis of intraspinal mesenchymal chondrosarcoma from preoperative imaging studies, radiologists should remember that it is one of the intraspinal tumors occurring in both children and adults.

The differential diagnosis should include hemangioblastoma, neurofibroma, and schwannoma. In spinal hemangioblastomas, cervical and thoracic cords are often involved. Most hemangioblastomas are intramedullary; however, they can also be intradural or purely extradural. Fortunately, hemangioblastomas are generally present in adults, and approximately 30% of patients with spinal cord hemangioblastomas have von Hippel-Lindau syndrome. Neurofibromas of the spine are rarely observed as isolated lesions and are most often seen in patients with neurofibromatosis type 1. Schwannomas are very uncommon in children and are sometimes associated with neurofibromatosis type 2.

In conclusion, the prognosis of this malignant tumor is poor regardless of the site of occurrence. Local or distant metastases may appear 20 or more years after the appearance of the primary tumor. Radical surgical removal is the treatment of choice for the primary tumor. The use of postoperative radiotherapy and chemotherapy for the

**Fig. 6** Postoperative sagittal T1-W gadolinium-enhanced MR image shows the complete removal of the tumor and no tumor recurrence



treatment of this tumor remains controversial. Most authors agree that radiotherapy is recommended in cases in which the tumor is resectable; chemotherapy is the treatment of choice in recurrent cases.

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