Spinal Canal Involvement in Solitary Infantile Myofibromatosis: Case Report

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Abstract
Infantile myofibromatosis involving the spinal canal is very rare; only 11 cases have been reported so far in the literature. The authors present a case of an 18-month-old male child who presented with the history of dribbling of urine and weakness in bilateral lower limbs since 2 months. MRI of spine revealed single intramedullary intradural space-occupying lesion (SOL) at D1 to D2.

Keywords
► solitary infantile myofibromatosis
► spinal canal
► intramedullary

Introduction
Infantile myofibromatosis is the most common fibrous disorder of infancy and early childhood. It was first described by Stout1 in 1954 who named this disease congenital generalized fibromatosis. In 1981, Chung and Enzinger2 renamed the disease as infantile myofibromatosis to emphasize the microscopic resemblance to smooth muscle tissue. It may occur in two distinct forms: multicentric and solitary. In both cases, involvement of the central nervous system is unusual. Spine myofibromas are exceptional, and most of the cases reported in the literature represent secondary locations of visceral lesions. Here, we are reporting good outcome in the case of intramedullary solitary type of infantile myofibromatosis with no visceral involvement.

Case Report

History and Examination
An 18-month-old male child presented with history of dribbling of urine and weakness in bilateral lower limbs noticed by his parents since about 2 months for which the patient underwent MRI scan of spine.

Operative Course
C-7 to D-2 laminectomy was done under general anesthesia after localizing D1 in prone position. Dura was identified and midline cordotomy was done with complete excision of encapsulated intramedullary SOL of ~2 × 1.4 cm in size.

Postoperative Course
On the first postoperative day, the patient was taking oral feeds and passed stools. On the second postoperative day, a catheter free trial was given and a clear stream of urine with improved motor power of bilateral lower limbs

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(MRC grade ⅖) was observed. The patient gradually recovered and was discharged on 14th postoperative day. The patient was under regular follow-up and within a span of 2 months was able to walk with support.

Histopathology
Encapsulated SOL was sent for histopathological examination which revealed well-circumscribed spindle cell tumor composed of oval to elongated to spindle cells arranged in sheets. Neoplastic cells were desmin negative (see Fig. 2), smooth muscle actin positive (see Fig. 3), S-100p focally positive, KI-67 positive, 2 to 3%, PAN cytokeratin negative suggestive of infantile myofibromatosis (see Fig. 4).

Discussion
Infantile myofibromatosis involving the spinal canal is very rare; 11 cases have been reported so far in the literature including 4 cases of the solitary form and a single case of intramedullary solitary form (See Table 1).3–6 To the best of our knowledge, we are reporting second case of

Fig. 1 Preoperative MRI images.

Fig. 2 Desmin—negative.

Fig. 3 Smooth muscle actin—positive.
intramedullary solitary type IM localized within the spinal cord without any visceral involvement.

The etiology and pathogenesis of IM are still obscure. The solitary form of infantile myofibromatosis mostly occurs in the soft tissues of the head and neck, followed by the upper extremities and trunk. Patients usually have a poor clinical symptomatology, except for occasional pain caused by compression of adjacent nerves.\(^7\)–\(^9\)

Our patient’s symptoms of dribbling of urine and weakness in bilateral lower limbs pointed toward compressive symptoms of tumor in spinal canal.

The usual clinical course of the solitary form is initial rapid growth followed by spontaneous regression within the first 2 years.

Conservative management is usually adopted for those without visceral involvement and complications. We did surgical decompression that was probably the correct decision in our case as literature says that younger patient age at diagnosis is associated with a more rapid expansion rate for infantile myofibromatosis.\(^10\)

In aggressive cases, there is limited experience of success with radiation therapy, different combination of chemotherapy,\(^11,12\) steroid injection,\(^11,12\) and α-interferon.

### Conclusion

Given the rarity of this condition, correct preoperative diagnosis could not be made initially until after histopathological examination. Though a rare disorder, IM must be suspected when evaluating children who present with either multiple lytic bone lesion or solitary/multiple tumors in the soft tissues, particularly during the neonatal or infancy period. Early surgical decompression of spinal cord led to improved neurological outcome in the patient with infantile myofibromatosis.
Table 1  Reported patients with infantile myofibromatosis involving the spinal canal

<table>
<thead>
<tr>
<th>Authors and year</th>
<th>Age, sex</th>
<th>Symptoms</th>
<th>Disease type</th>
<th>Lesion location</th>
<th>Spinal cord invasion</th>
<th>Management</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Davies et al, 1994</td>
<td>Birth, F</td>
<td>Brachial palsy</td>
<td>Multicentric</td>
<td>Cervical spinal cord, left side neck</td>
<td>No</td>
<td>Chemotherapy</td>
<td>Good</td>
</tr>
<tr>
<td>Giannini et al, 1995</td>
<td>33 years, F</td>
<td>Paralysis of left leg</td>
<td>Multicentric</td>
<td>Iliac bone, posterior vertebral element, left side vertebral column</td>
<td>No</td>
<td>Chemotherapy</td>
<td>Good</td>
</tr>
<tr>
<td>Wada et al, 1998</td>
<td>Birth, F</td>
<td>Paraplegia, bladder and bowel dysfunction</td>
<td>Multicentric</td>
<td>Bone, left side of vertebra, intrapelvic, spinal cord (L3–5)</td>
<td>No</td>
<td>Partial removal</td>
<td>Good</td>
</tr>
<tr>
<td>Dimmick and Wood, 1983</td>
<td>Birth, M</td>
<td>Quadriplegia</td>
<td>Multicentric</td>
<td>Skin, tongue, spinal cord (C3-T3)</td>
<td>Yes</td>
<td>Supportive care</td>
<td>Good</td>
</tr>
<tr>
<td>Christensen et al, 1961</td>
<td>Birth, M</td>
<td>No spontaneous movement, respiratory failure</td>
<td>Multicentric</td>
<td>Bone, skin, heart, liver, brain, thymus, gut wall, kidney, spinal cord (T2–4, T8–10)</td>
<td>Yes</td>
<td>Supportive care</td>
<td>Died (10 days)</td>
</tr>
<tr>
<td>Altemani et al, 1985</td>
<td>Birth, F</td>
<td>Quadriplegia, dyspnea</td>
<td>Multicentric</td>
<td>Bone, right side of neck</td>
<td>Yes</td>
<td>Supportive care</td>
<td>Died (75 days)</td>
</tr>
<tr>
<td>Stewart et al, 1989</td>
<td>Birth, M</td>
<td>Paraplegia, respiratory failure</td>
<td>Multicentric</td>
<td>Skin, muscle, lung, spinal cord (T6–8), myocardium, GI tract</td>
<td>Yes</td>
<td>Supportive care</td>
<td>Died (45 days)</td>
</tr>
<tr>
<td>Beyer et al, 1990</td>
<td>5 years, M</td>
<td>Local pain</td>
<td>Solitary</td>
<td>Sacrum</td>
<td>No</td>
<td>Surgical removal</td>
<td>Good</td>
</tr>
<tr>
<td>Asirvatham et al, 1994</td>
<td>18 years, M</td>
<td>Local pain</td>
<td>Solitary</td>
<td>Spinal cord (C2)</td>
<td>No</td>
<td>Surgical removal</td>
<td>Good</td>
</tr>
<tr>
<td>Tamburini et al, 2003</td>
<td>4 months, M</td>
<td>Spastic paraparesis</td>
<td>Solitary (intramedullary)</td>
<td>Spinal cord (C7-conus)</td>
<td>Yes</td>
<td>Partial removal</td>
<td>Good</td>
</tr>
<tr>
<td>Eun Ji Kim, et al, 2013</td>
<td>8 months, F</td>
<td>Paraplegia</td>
<td>Solitary</td>
<td>Spinal cord (T6-conus)</td>
<td>Yes</td>
<td>Partial removal</td>
<td>Good</td>
</tr>
<tr>
<td>Present case</td>
<td>18 months, M</td>
<td>Paraparesis, bladder dysfunction</td>
<td>Solitary (intramedullary)</td>
<td>Spinal cord (T1–2)</td>
<td>Yes</td>
<td>Complete removal</td>
<td>Good</td>
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References