Rapid-onset paraparesis and quadriplegia in patients with intramedullary spinal dermoid cysts: report of 10 cases

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OBJECTIVE Intramedullary dermoid cysts are rare tumors of the spinal cord. Presentation with rapid onset of paraparesis or quadriplegia (onset within 2 weeks) is rarer still. The authors present their experience in the management and outcome of patients with such a presentation.

METHODS Patient records between 2000 and 2014 were retrospectively reviewed to identify those with intraspinal dermoid cysts who presented with rapid-onset paraparesis or quadriplegia. Their clinical, radiological, operative, and follow-up data were analyzed.

RESULTS Of a total of 50 patients with intraspinal dermoid cysts managed during the study period, 10 (20%) presented with rapid-onset paraparesis or quadriplegia; 9 patients ranged in age from 8 months to 2 years, and 1 patient was 25 years old. A dermal sinus was seen in the lumbar region of 4 patients, the sacral region of 3, and the thoracic region of 1, and in 1 patient no sinus was found. All except 1 patient presented with rapid-onset paraparesis secondary to infection of the intramedullary dermoid cyst. One patient presented with rupture of a dermoid cyst with extension into the central canal up to the medulla. Early surgery was done soon after presentation in all except 2 patients. Among the 9 patients who underwent surgery (1 patient did not undergo surgery), total excision of the intramedullary dermoid cyst was done in 3 patients, near-total excision in 4 patients, and partial excision in 2 patients. Of the 9 patients who underwent surgery, 8 showed significant improvement in their neurological status, and 1 patient remained stable. The 1 patient who did not undergo surgery died as a result of an uncontrolled infection after being discharged to a local facility for management of wound infection.

CONCLUSIONS Early recognition of a dermal sinus and the associated intraspinal dermoid cyst and timely surgical intervention can eliminate the chances of acute deterioration of neurological function. Even after an acute onset of paraparesis or quadriplegia, appropriate antibiotic therapy and prompt surgery can provide reasonably good outcomes in these patients.

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KEY WORDS children; dermal sinus; spinal cord; tumor; paraparesis; quadriplegia; spine
Methods

Patient Data

In a retrospective chart review performed at our institution, we identified 10 patients with intraspinal dermoid cysts who presented between 2000 and 2014 with rapid-onset (within 2 weeks) paraparesis or quadriparesis. In the same period, we performed surgery on 40 other patients; 9 had no neurological compromise. These 9 patients were treated for a dermal sinus with intermittent discharge, subcutaneous swelling, or meningitis. The other 31 patients presented with weakness of gradual onset of more than 1 month’s duration. The study patient population comprised 9 pediatric patients (5 boys and 4 girls) with an age range of 8 months to 2 years (median 19 months); the age of the adult patient was 25 years.

Clinical Presentation

The clinical presentations are summarized in the Table 1. Five patients presented with paraparesis, 2 patients with quadriparesis, 2 with paraplegia, and 1 with quadriplegia. The onset of weakness was sudden in 3 patients, and the other patients presented with progressive weakness that they noticed over a period of 2 weeks. Eight patients were unable to stand or walk and were bed bound at presentation. In 1 infant (aged 8 months) with paraparesis, the parents noticed paucity in limb movements, and another child older than 1 year could not walk, even with support. All 9 pediatric patients had a dermal sinus (4 in the lumbar region, 3 in the sacral region, and 2 in the thoracic region). Seven patients had serous or purulent discharge from the dermal sinus. Two patients (Patients 2 and 4) had recurrent episodes of meningitis before presenting to us, and 1 of these patients had meningitis at presentation to us. One patient (Patient 4) who presented with quadriparesis had an episode of meningitis 4 months before the onset of weakness. As a result of the meningitis, he developed generalized seizures and left middle cerebral artery territory infarct, which caused right hemiparesis. After the second episode of meningitis, he developed progressive worsening of weakness in all 4 limbs over a period of 10 days. Another patient who presented with paraplegia (Patient 10) had undergone incomplete excision of a sacral dermal sinus without excision of the dermoid cyst at another institution 1 year before presenting to us.

Preoperative Bacteriology

Of the 7 patients with an active discharge from the dermal sinus, culture results were positive in 6. The commonest organisms were Gram-negative bacilli (Escherichia coli in 2 patients, Klebsiella in 1, and Pseudomonas in 1) sensitive to amikacin and gentamicin. The other 2 organisms found were enterococcus and coagulase-negative Staphylococcus.

Imaging Findings

The commonest location of the dermoid cyst was the lumbosacral region, seen in 4 (40%) patients. The other sites were thoracolumbar (30%), thoracic (20%), and sacral (10%). All 9 patients with an intramedullary abscess had radiological evidence of a dermal sinus seen as a T1-weighted hypointense tract that extended intradurally from the skin through a defect in the posterior elements at the affected level (Fig. 1). The intramedullary abscess was seen as a mass with T2-weighted heterogeneous intensity that caused expansion of the cord with mass effect and widespread perilesional cord edema (Fig. 2). In contrast images, there was an enhancing peripheral rim and a solid component in all the cases with septations (Fig. 3). The diagnosis of infected dermoid cyst was made in the presence of the above-mentioned features of an abscess, an intramedullary location, and a dermal sinus. For Patient 4, the images showed extension of the abscess into the central canal, which caused enhancement up to the cervicomedullary junction (Fig. 4). MR images of the brain of the same patient showed features of an old left middle cerebral artery infarct, which he developed 4 months before the onset of quadriparesis (Fig. 4). One patient (Patient 3) lacked a dermal sinus both clinically and on imaging. This patient presented with quadriplegia secondary to rupture of cyst contents into the cord and central canal with evidence of fat droplets seen at the level of the medulla in plain T1-weighted images (Fig. 5).

TABLE 1. Demographic and clinical profile of the study patients

<table>
<thead>
<tr>
<th>Patient No.</th>
<th>Age &amp; Sex</th>
<th>Site of Dermoid Cyst</th>
<th>Duration of Symptoms (days)</th>
<th>Type of Weakness</th>
<th>Dermal Sinus</th>
<th>Functional Status</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>10 mos, M</td>
<td>T6–8</td>
<td>1</td>
<td>Paraparesis</td>
<td>+</td>
<td>Bedridden</td>
</tr>
<tr>
<td>2</td>
<td>2 yrs, M</td>
<td>L3–S2</td>
<td>4</td>
<td>Quadriplegia</td>
<td>+</td>
<td>Bedridden</td>
</tr>
<tr>
<td>3</td>
<td>25 yrs, F</td>
<td>T11–L1</td>
<td>7</td>
<td>Quadriplegia</td>
<td>–</td>
<td>Bedridden</td>
</tr>
<tr>
<td>4</td>
<td>1 yr, M</td>
<td>L3–S1</td>
<td>10</td>
<td>Quadriplegia</td>
<td>+</td>
<td>Bedridden</td>
</tr>
<tr>
<td>5</td>
<td>8 mos, F</td>
<td>T6–10</td>
<td>15</td>
<td>Paraparesis</td>
<td>+</td>
<td>Bedridden</td>
</tr>
<tr>
<td>6</td>
<td>2 yrs, M</td>
<td>S3–4</td>
<td>5</td>
<td>Paraplegia</td>
<td>+</td>
<td>Bedridden</td>
</tr>
<tr>
<td>7</td>
<td>14 mos, M</td>
<td>L5–S1</td>
<td>1</td>
<td>Paraparesis</td>
<td>+</td>
<td>Sit with support</td>
</tr>
<tr>
<td>8</td>
<td>2 yrs, M</td>
<td>T10–L3</td>
<td>10</td>
<td>Paraparesis</td>
<td>+</td>
<td>Sit with support</td>
</tr>
<tr>
<td>9</td>
<td>1 yrs, F</td>
<td>T11–L5</td>
<td>15</td>
<td>Paraparesis</td>
<td>+</td>
<td>Bedridden</td>
</tr>
<tr>
<td>10</td>
<td>2 yrs, F</td>
<td>L5–S2</td>
<td>1</td>
<td>Paraplegia</td>
<td>+</td>
<td>Bedridden</td>
</tr>
</tbody>
</table>

* = present; – = absent.
Preoperative Management

In the 7 patients with a dermal sinus and active discharge, antibiotic therapy was initiated based on the sensitivity report. After starting on appropriate antibiotics, surgery was performed, and antibiotics were continued postoperatively. The total duration of antibiotic therapy from the time of presentation (preoperatively and postoperatively) was 6 weeks for intravenous antibiotics and 4 weeks for oral antibiotics. Antibiotics such as amikacin and gentamicin were used in most of our cases. One patient (Patient 2) had improvement in limb power after treatment with preoperative antibiotics. Another patient (Patient 7) had a paraspinal abscess that was aspirated under ultrasound guidance, and the pus was sent for culture and sensitivity testing. He was treated with appropriate antibiotics. This patient, too, had good improvement in limb power preoperatively. The patient with a ruptured dermoid cyst (Patient 3) was treated with steroids at presentation and was followed up with definitive surgery after 1 month. In the interval, there was improvement in her function to Nurick Grade 3 (from Nurick Grade 5 at presentation). The patient with an infected dermoid cyst (Patient 6) underwent incision and drainage of the subcutaneous abscess, which had extended into the spinal canal. After the drainage, he was managed with appropriate antibiotics. This patient was discharged with advice to return for surgery after the wound had healed.

Surgical Management

Timing of Surgery

Of the 10 patients, 7 were managed with surgical intervention soon after presentation. These 7 patients underwent surgery at a median interval of 8 days (range 7–40 days) from the day of admission; the median time included the time for evaluation and time to control meningitis and/or local infection with appropriate antibiotic therapy for at least 5 to 7 days. Of the 3 patients who were managed conservatively at presentation, 1 had a ruptured dermoid cyst (Patient 3). This patient underwent surgery 1 month after presentation. The remaining 2 patients had an infected dermoid cyst with associated lower respiratory tract infection (Patient 2) and dermal sinus infection with skin involvement (Patient 6). Patient 2 underwent surgery 2 months after presentation. Patient 6 did not undergo surgery, because he was lost to follow-up.

Surgery

In 7 patients, we performed laminoplasty, and in 2 pa-
tients, laminectomy was done. In all 9 patients a midline incision was used to isolate the dermal sinus, and the sinus tract was followed into the intradural compartment. The dura was opened in the midline around the entry of the dermal sinus. The arachnoid was thick and opaque. The cord was entered in every case. A myelotomy had to be performed to excise dermoid cysts located in the thoracic region. In most of the cases, pus present in the cord or the central canal was drained along with the excision of the cyst. The cyst was seen as a pale, grayish mass filling the spinal canal. The mass consisted of granulation tissue and hair with pus-like material from within the cord. The wall of the cyst was separated from the spinal cord and excised totally in 3 patients, near totally in 4 patients, and partially in 2 patients. In cases of lumbosacral dermoid cyst, the cauda equina nerve roots were densely adherent to the walls of the cyst laterally and ventrally. Superiorly, these cysts were seen to be entering into the spinal cord. Intraoperative monitoring (IOM) using motor evoked potentials (MEPs) was used for 1 patient who presented with a ruptured dermoid cyst (Patient 3) at the conus. In this patient, there was improvement in the intraoperative MEPs after excision of the cyst. IOM was not used in other cases because the patients’ functional grade and limb power were poor.15 Postoperative drains were used routinely in all the cases. All patients were kept on bed rest in a prone position with elevation of the foot end of the bed for 5 days. None of the patients had any wound complications such as wound infection or CSF leak.

Follow-Up and Outcome

All patients were followed up in the outpatient clinic except for 2 patients who were contacted over the telephone. Nine patients had a follow-up time of 1–64 months (median 39 months) (Fig. 6). Of these 9 patients, 8 had significant improvement in their lower-limb function, and the remaining patient did not show any improvement. Of 8 patients who improved, 4 patients (Patients 1, 3, 7, and 8) were able to walk without support, 3 (Patients 4, 5, and 9) could walk with support, and 1 patient (Patient 10) was able to stand with support. One patient (Patient 2) who presented with weakness of all 4 limbs showed improvement in his upper-limb power after initiation of preoperative antibiotics. However, no additional improvement in neurological status was noticed after the surgery. At the last follow-up, 3 years after surgery, his neurological status remained stable and he was able to sit with support.

Discussion

Epidemiology

Spinal dermoid cysts are rare benign developmental tumors that constitute 5%–7% of all primary intradural tumors in the pediatric population and 1.1% of all intraspinal tumors.6 Chan and Gold3 found that infected dermoid cysts are a rare cause of intramedullary abscess. In their study, only 1 of 25 intramedullary abscesses was caused by a dermoid cyst infected by contiguous spread from a dermal sinus. In children, a dermal sinus is the most common source of an intramedullary abscess; it is the cause of an intramedullary abscess in 45% of pediatric patients. In contrast, a dermal sinus is the cause of only 24% of intramedullary abscesses in adults.3,17 There have been only isolated case reports of infected dermoid cysts located in the thoracic region. In most of the cases, pus present in the cord or the central canal was drained along with the excision of the cyst. The cyst was seen as a pale, grayish mass filling the spinal canal. The mass consisted of granulation tissue and hair with pus-like material from within the cord. The wall of the cyst was separated from the spinal cord and excised totally in 3 patients, near totally in 4 patients, and partially in 2 patients. In cases of lumbosacral dermoid cyst, the cauda equina nerve roots were densely adherent to the walls of the cyst laterally and ventrally. Superiorly, these cysts were seen to be entering into the spinal cord. Intraoperative monitoring (IOM) using motor evoked potentials (MEPs) was used for 1 patient who presented with a ruptured dermoid cyst (Patient 3) at the conus. In this patient, there was improvement in the intraoperative MEPs after excision of the cyst. IOM was not used in other cases because the patients’ functional grade and limb power were poor.15 Postoperative drains were used routinely in all the cases. All patients were kept on bed rest in a prone position with elevation of the foot end of the bed for 5 days. None of the patients had any wound complications such as wound infection or CSF leak.
patients, all but 1 (9 of 10) had a dermal sinus, but every patient (9 of 9) who had an intramedullary abscess had an associated dermal sinus.

**Embryology of Dermoid Cysts**

There are various theories on the origin of dermoid cysts. In 1897, Von Bostroem postulated that dermoid cysts are formed as a result of a defect in neural tube closure, which occurs between the 3rd and 5th gestational weeks. He suggested that these cysts originate from the totipotent ectoderm inclusions, which could be either congenital or acquired. These ectodermal inclusions give rise to all the epidermal and dermal elements found within these cysts. He proposed that the formation of these dysembryogenic tumors depends on the timing of the defective events. Predilection of the lumbosacral region for dermoid cysts is a result of the caudal neuropore being the last to close in the formation of the neural tube. Holmdahl discovered that the conus developed from the caudal mass of cells by secondary neurulation and reported that dermoid cysts are formed by these pluripotential cells. The dermal sinus with or without the dermoid cyst is formed by the failure of separation of the surface ectoderm from the neural tube.

**Clinical Presentation**

The median age at presentation in our series of patients with dermoid cysts was 19 months. The age distribution in our series is similar to that reported by Simon et al., who reported that the median age at presentation of an intramedullary abscess in children with a preexisting anatomical defect (dermal sinus) was 15.5 months with the intramedullary abscesses developing before 5 years of age in 68% of the patients. The age of the patient who presented with a ruptured dermoid cyst (Patient 3) was 25 years. Most patients with a dermoid cyst present because of mass effect on neural tissues, which causes low back pain or radiculopathy. If there is an associated dermal sinus, then the presentation is likely to be early (within the first decade of life) because of infection contracted through this sinus causing recurrent meningitis, local abscess, or an in-
tradural abscess. Among the various manifestations, the least common is an intramedullary abscess.\textsuperscript{10} It was previously documented that dermoid cysts without a dermal sinus might present acutely later in life because of a rupture.\textsuperscript{7,12} However, patients with an intramedullary abscess associated with a dermal sinus present at a younger age because of the presence of a pathway for the spread of infection; this was noted in our series and has been reported by others.\textsuperscript{1,3,16} Patients with a dermal sinus and a dermoid cyst can also present with only discharge of purulent materials or cyst debris from the sinus without any neurological impairment. The most common site for a dermoid cyst and the dermal sinus in our study was the lumbar region, similar to what has been reported in previous studies.\textsuperscript{5,8} The acute onset of significant weakness described in this series is an unusual presentation of these rare benign lesions and might be the result of a possible ischemic process secondary to infective vasculitis.

Dermal sinuses need not always be associated with an intraspinal dermoid cyst. During the period of the study, we performed surgery on 8 patients with a dermal sinus without an intraspinal dermoid cyst. Of 50 patients with dermoid cysts who underwent surgery in the study period, 9 presented with only a dermal sinus with intermittent discharge. These patients (with a dermal sinus with or without an intraspinal dermoid cyst) presented with either intermittent discharge or episodes of meningitis without any neurological deficits. Although it is possible, we did not note any patient with a dermal sinus with an intraspinal abscess in the absence of an intraspinal dermoid cyst.

Radiology

A dermoid cyst without infection can be recognized by the signal characteristics of fat, lack of enhancement, and heterogeneous intensity seen in T1- and T2-weighted images. These features are not usually seen in MR images.
of other intramedullary tumors.6 The presence in contrast MR images of an intramedullary mass with peripheral rim enhancement and cord edema appreciated in T2-weighted images is suggestive of an intramedullary abscess.6 The presence of a dermal sinus in these cases, seen as a T2-weighted hypointense tract extending intradurally from the skin, favors the preoperative diagnosis of an infected dermoid cyst. In the majority of our cases, there was extensive ependymal and arachnoid enhancement associated with cord edema, which extended up to the cervical cord. One of our patients had features of a ruptured dermoid cyst, with T1-weighted hyperintense fat signals seen within the central canal extending up to the level of medulla. These MRI features are similar to those reported by Garg and Kumar7 from their series of 3 ruptured spinal dermoid cysts. However, their patients had only mild limb weakness, which is in contrast to our patient who had significant quadripareisis.

Surgery and Outcome

Most authors recommend early surgical management for an intramedullary abscess caused by a dermal sinus.1,5,10,18 The aims of surgery are to obtain a diagnosis, ascertain the causative organism if not already determined through culture of sinus wound discharge, drain the abscess, excise the wall of the dermoid cyst as much as safely possible, and remove the source of infection (dermal sinus). We prefer to perform surgery early in children with a diagnosis of dermal sinus with or without an intraspinal dermoid cyst. We preferred laminectomy to laminectomy for most of our pediatric patients to avoid progressive deformity. We decided to intervene surgically at first presentation for most of our patients (77%). In the cases reviewed by Chan and Gold,7 all the patients were managed with surgical intervention at presentation, which is also the preferred timing of surgery reported by others.1,10,14,19 However, there is no mention in the literature regarding the duration of antibiotic therapy before or after surgical drainage.

Intraoperative monitoring with MEPs was used for 1 patient who presented with a ruptured dermoid cyst (Patient 5) at the conus. In this patient, there was improvement in intraoperative MEPs after excision of the cyst. In the other cases, IOM was not used because the patients’ functional grade and lower-limb power were poor. In our previous publication, we noted that in patients with lower-limb motor power Grade 0/5 to 2/5, baseline intraoperative MEPs were never obtained.15 It is also well known that in children younger than 2 years (before the myelination of the corticospinal tract is complete), lower-limb MEPs are notably difficult to obtain. Hence, we do not attempt MEP monitoring in very young children or in those with a lower-limb power of Grade 2/5 or worse. The major factor that prevents the total excision of an intramedullary dermoid cyst is dense adhesion of the wall of the cyst to the surrounding neural structures. This adhesion is worse in patients with infection, and total excision in this situation can result in permanent neurological damage. In our study, improvement in neurofunctional function was seen in 70% of the patients, although good recovery (able to walk without support) was seen in only 30% of the cases. These results are similar to those reported in most studies, in which recovery was noted in approximately 20% of patients with an infected intramedullary dermoid cyst.3,4,11,17

Conclusions

The presence of a dermal sinus and spinal dermoid cyst is associated with a significant risk of developing an intramedullary abscess with catastrophic neurological consequences. Early recognition of these lesions and resection of the dermal sinus and dermoid cyst before the onset of neurological dysfunction is the ideal management plan. Our series of cases emphasizes the need for a high index of suspicion for a dermal sinus in children with acute-onset limb weakness. Even in patients who present with acute-onset weakness, prompt and appropriate antibiotic therapy and surgery can lead to a reasonably good outcome in most patients.

References


**Disclosure**
The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

**Author Contributions**
Conception and design: Rajshekhar. Acquisition of data: Girishan. Analysis and interpretation of data: Girishan. Drafting the article: Girishan. Critically revising the article: Rajshekhar. Approved the final version of the manuscript on behalf of both authors: Rajshekhar. Statistical analysis: Girishan. Study supervision: Rajshekhar

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