Spinal Neurenteric Cyst Presenting as Burning Feet Syndrome
RS Jain

Abstract
Spinal neurenteric cysts are rare congenital cysts of endodermal origin. A 34 years old man presented with burning feet syndrome of two years duration. Magnetic resonance imaging (MRI) revealed an intradural extramedullary cystic mass lesion extending from L2 to L5 vertebrae causing severe compression and displacement of terminal portion of conus medullaris and filum terminale to the right side. Additionally, tethered cord and filar lipoma were also present. Cystic mass and filar lipoma were resected alongwith release of tethered cord. Histopathology confirmed a neurenteric cyst. This case is reported in view of rare occurrence and peculiar presentation.

INTRODUCTION
Spinal neurenteric cysts are rare congenital endothelium-lined structures considered to be a form of occult spinal dysraphism (OSD). The presentation may vary from a simple isolated intraspinal cyst to that of the so-called split notochord syndrome, as proposed by Bentley and Smith in which there may be multiple visceral and vertebral anomalies. These cysts occur most frequently in the lower cervical and upper thoracic regions as intradural extramedullary mass. They manifest clinically as progressive compressive myelopathy in young adults but may also cause relapsing-remitting symptoms and recurrent meningitis.

This report describes occurrence of a rare congenital abnormality in an adult with peculiar clinical presentation of burning feet syndrome which has not been described earlier in the English literature.

CASE REPORT
A 34 years old man was referred to neurology services with two years history of burning sensation in both feet progressing to involve lower legs. He was not getting relief with multi-vitamins, tricyclic antidepressants and antiepileptics. He was a non-smoker but used to consume alcohol occasionally. There was nothing to suggest malnutrition or malabsorption. Neurological examination was perfectly normal and there were no neurocutaneous stigmata. Routine hematological and biochemical parameters were normal. Serum HIV and VDRL were non-reactive.

Electrophysiology revealed only slightly delayed distal latency of left common peroneal nerve (Table 1). Plain radiology of lumbo-sacral spine did not show any developmental bone abnormalities. As he was not improving with various drugs, an MRI scan of lumbosacral spine was advised which surprisingly revealed an intradural extramedullary cystic mass lesion in posterior subarachnoid space extending from L2 to L5 vertebrae (Figure 1). The signal intensity of mass lesion was hyperintense to CSF on both T1 and T2 weighted images. This mass was causing severe compression and displacement of thecal sac with conus medullaris, nerve roots, and filum terminale to the right side. Additionally, tethered cord, and filar lipoma opposite L3-4 and S1-2 vertebral bodies were seen.

The diagnostic possibilities considered included epidermoid and arachnoid cysts. He underwent L2 to L5 laminectomy. Cystic mass and filum terminale lipoma were completely resected along with release of tethered cord. Cyst was thin walled and contained milky white fluid. Histopathology revealed a cyst lined by cuboidal epithelioid pseudostratified columnar ciliated epithelium. At places, portions of smooth muscle, cartilage and mucous glands were identified in the wall. These features were compatible with type B variety as per the classification of Wilkins and Odum.1 The patient became completely symptom-free from burning feet within few days after surgery. A gadolinium enhanced MRI obtained three months after surgery showed no residual lesion.

DISCUSSION
Neurenteric cysts are rare congenital abnormalities believed to be derived from an abnormal connection between the primitive endoderm and ectoderm during third week of
Fig. 1: MRI scan (T₂ weighted) of lumbosacral spine showing an intradural extramedullary cystic mass lesion in posterior subarachnoid space extending from L₂ to L₅ vertebrae. The signal intensity of mass lesion was hyperintense to CSF on both T₁ and T₂ weighted images. This mass was causing severe compression and displacement of thecal sac with conus medullaris, nerve roots, and filum terminale to the right side.

life. Embryologically, the main theories for neurenteric cyst formation include, 1) a primary adhesion of endoderm anterior to the notochord, 2) incomplete excalation of the notochord, 3) persistence of the neurenteric canal with a split notochord, and 4) displacement of endodermal cells. They are not confined to the spinal column but may be found within the brain (cerebellopontine angle, medulla oblongata, lateral ventricles and fourth ventricle), mediastinum, abdomen, pelvis, or even in a subcutaneous location.

Associated vertebral abnormalities including hemivertebrae, fused vertebrae, spina bifida, diastematomyelia, or fibrous connecting remnant - Kovalesky canal and congenital skin anomalies (hyperpigmentation or hypertrichosis in the midline of the back) are often present. The case reported here serves to illustrate that intraspinal neurenteric cyst may occur in isolation, without vertebral, visceral or skin abnormalities.

These cysts occur most frequently in the lower cervical and upper thoracic region as intradural extramedullary mass and manifest clinically as progressive compressive myelopathy in young adults. It is interesting to note that the cyst was extending from L₂ to L₅ vertebral level in our case but did not produce any neurological signs. This may be related to generally slow cyst growth and space reserve in the lumbar region.

Burning feet syndrome is a clinically heterogenous entity associated with chronic malnutrition or malabsorption resulting in deficiencies of various vitamins. It is a manifestation of sensory neuropathy with predominant involvement of unmyelinated fibers. Burning feet were occasionally found to be associated with tarsal tunnel syndrome. Autosomal dominant burning feet syndrome was described by Stogbauer F and Kuhlabaumer G. However, there can be other mechanisms also as observed by Galer BS in a case where sympathetically maintained bilateral burning foot pain got promptly relieved following unilateral lumbar sympathetic nerve blocks suggesting a central mechanism operating in this disorder. Recently, there was a report of thoracic spinal arterio-venous malformation presenting as burning feet syndrome.

An unequivocal explanation for burning feet and neurenteric cyst in our patient is not available. Theoretically, the following mechanisms might play a part - (i) burning feet may be of radicular origin rather than neuropathy, (ii) large cyst extending from L₂ to L₅ with severe compression and displacement of terminal conus medullaris and filum terminale with tethered cord might have produced symptoms due to spinothalamic tract involvement, (iii) sympathetic mechanism might be operating as reported by Galer BS.

The reasons for the burning feet syndrome resulting from lumbar neurenteric cyst and relief of symptoms after surgical excision in our patient remain unknown, however. The present case demonstrates that the possibility of a structural lesion higher up should be kept in mind in cases of burning feet syndrome particularly when electrophysiology fails to document a definite neuropathy and patient does not respond to usual drug treatment. As far as I know, this is the first case report of spinal neurenteric cyst presenting as burning feet syndrome.

REFERENCES

Table 1: Electrophysiology

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<tr>
<th>Nerve</th>
<th>Stim. site</th>
<th>Rec. site</th>
<th>Latency (ms)</th>
<th>Amplitude (mV)</th>
<th>Distance (mm)</th>
<th>Velocity (m/s)</th>
<th>F-wave min. lat. (ms)</th>
<th>H-reflex lat. ampl. (mV)</th>
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<tbody>
<tr>
<td>Right common peroneal</td>
<td>Ankle</td>
<td>EDB</td>
<td>3.1</td>
<td>6.7mV</td>
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<td>5.9mV</td>
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<td>49.3</td>
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<td>AH</td>
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<td>7.3mV</td>
<td>140</td>
<td>43.7</td>
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<tr>
<td>Left sural</td>
<td>Mid calf</td>
<td>Ankle</td>
<td>3.5</td>
<td>6.9mV</td>
<td>140</td>
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<tr>
<td>Right sup. peroneal</td>
<td>Lat. Leg</td>
<td>Foot</td>
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<td>5.2mV</td>
<td>140</td>
<td>41.1</td>
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<tr>
<td>Left sup. peroneal</td>
<td>Lat. Leg</td>
<td>Foot</td>
<td>3.7</td>
<td>4.9mV</td>
<td>140</td>
<td>41.1</td>
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</tbody>
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Normal EMG study of extensor digitorum brevis (EDB), tibialis anterior, gastrocnemius, quadriceps, hamstrings and glutei muscles in both lower extremities.


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Announcement

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