Case Report

Spinal Intramedullary Arachnoid Cyst – a Rare Case or a Distinct Rare Entity?

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Abstract

Background: Spinal arachnoid cyst is a common cause of non-neoplastic spinal compressive myelopathy. Intramedullary arachnoid cyst is very unusual and rarely addressed in the literature.

Methods: We report the Clinico-pathological account of a thoracic intramedullary arachnoid cyst in an adult female with an unusual course of neurological recovery and present a review on spinal intramedullary arachnoid cyst.

Results: Fourteen cases of primary intramedullary cyst have been reported in the literature, mostly affecting the paediatric population. Only 5 cases have been reported in adults. Though rare, similar clinical experience from previous reported cases suggests that intramedullary arachnoid cyst really represent a distinct clinical entity. It should be considered in the differential diagnosis of cystic intramedullary lesions and merit incorporation into the existing classification of spinal arachnoid cyst.

Introduction

Arachnoid cysts are the commonest non-neoplastic causes of spinal cord compression [1-4]. They are more often encountered as incidental findings in examinations performed for other reasons [2]. Most of the arachnoid cysts are extradural in location, though it can be intradural [5,6]. We report clinicopathological detail of a rare case of thoracic intramedullary arachnoid cyst in an adult and review the literature of the spinal intramedullary arachnoid cyst.

Case Presentation

A 40-year-old lady presented with progressive weakness of both lower limbs since the last two months and was bed ridden for the last 15 days. She also developed urinary hesitancy and urgency in last 15 days.

On examination, the tone in the lower limbs was grossly increased. She was paraplegic and had graded sensory loss below L1. Posterior column sensations were impaired in lower limbs. There was extensor plantar response, and the reflexes in lower limbs were exaggerated. She also had bladder dysfunction. MRI of the spine revealed a well circumscribed, cystic non-enhancing intramedullary lesion at D11-12 (Figure 1a,b,c). The lesion was entirely intramedullary, hypointense on T1 and hyperintense on T2 without perilesional signal changes or syrinx formation (Figure 1c,d,e). She underwent laminotomy, midline myelotomy, midline myelotomy and partial excision of the cyst. At surgery there was focal bulge of the spinal cord. After performing the mid line myelotomy, cyst was identified one mm beneath the medullary tissue, containing clear fluid. The cyst wall was thin and translucent. The cyst was not under tension with no communication with the subarachnoid space nor was there an extramedullary component. The cyst was fenestrated and partially excised, as the cyst wall was adherent to the cord.

She regained significant power in lower limb immediately after surgery. She was ambulatory and had regained bladder control following day. At the time of discharge, she had complete recovery of motor weakness and control of bladder function. At the follow up of 50 months post surgery, she is functionally independent and has no neurological deficits.

Pathology

The cyst wall was variably thickened and lined by flattened,

Figure 1: MRI of the dorsal spine.
Figure 1a, b: Sagittal T1W images pre and post contrast scan respectively depicting well defined, hypointense lesion at D11-12 with no contrast enhancement.
Figure 1c: Sagittal T2W image shows a well defined hyperintense lesion at D11-12 vertebral level with no perilesional signal changes or syrinx.
Figure 1d & e: Axial T1W and T2W images shows well defined intramedullary which is hypointense and hyperintense respectively.
cuboidal epithelium - arachnoidal cells, resting on a collagenous subepithelial stroma. Ciliated cells or goblet cells were not present, thus excluding a neurenteric cyst. The hyalinised cyst wall contained several melanocytes and was closely adhere to the surrounding gliotic cord parenchyma (Figure 2). The epithelial cells expressed EMA confirming their meningothelial nature while the GFAP immunohistochemistry emphasized the reactive gliosis in the adjacent neural tissue.

**Discussion**

Spinal arachnoid cyst are common cause of non neoplastic spinal cord compression, however their etiopathogenesis and classification are still debated. Based on the findings of surgical examinations, radiological features and histopathological review of 22 cases, Nabors et al. [7], proposed a classification of spinal meningeal cysts into three categories: spinal extradural meningeal cysts without spinal nerve root fibers (Type I), spinal extradural meningeal cysts with spinal nerve root fibers (Type II), and spinal intradural meningeal cysts (Type III). Type 1 meningeal cysts were further classified into Type IA, the extradural arachnoid cyst and Type IB, the sacral meningocele.

Primary spinal intramedullary arachnoid cyst is extremely uncommon and has been sparsely reported in the English literature (Table 1) [5,6,8-17] and mostly in the paediatric population. To best of our knowledge only 5 such reports are available in adult population [5,9,11,13,17]. The existing classification lacks this entity which is usually described as an unusual location of arachnoid cyst with unusual clinical course. The etiopathogenesis and progression of spinal intramedullary arachnoid cyst is still elusive. Goyal et al. [11] suggested that the misplaced cellular elements during embryogenesis as the possible etiology. Fortuna and Mercuri [18] suggested trapping of arachnoid granulation at various locations including intramedullary as a pathogenic factor in cyst formation and CSF production and accumulation. We believe that CSF hemodynamic variation associated with normal activity produces a state of continued stress to an intrinsic arachnoid defect or an intraparenchymal rest with tenuous continuity with the subarachnoid space which progressively enlarges because of a ball valve mechanism causing entrapping of CSF within the cyst. The intrinsic arachnoid defect could be congenital. Whether there is genetic predisposition for such arachnoid defect is currently not known. However report of familial spinal intradural arachnoid cysts [19] raises a concern and may merit further study.

Interestingly the clinical course of all the reported intramedullary arachnoid cysts is comparable, suggesting a distinct pathomorphology.

![Figure 2: Histopathology and immunohistochemistry of intramedullary cyst wall.](image)

**Table 1:** Literature review of spinal intramedullary arachnoid cyst.

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Patient Age, Sex</th>
<th>Clinical Findings</th>
<th>Cyst Location</th>
<th>Association</th>
<th>operation</th>
<th>Outcome</th>
<th>FUP</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 Aithala et al[8]; 1999</td>
<td>7 yrs, M</td>
<td>progressive paraparesis,</td>
<td>D-4</td>
<td>No H/o systemic illness, H/o of trivial fall but no injury</td>
<td>DM-M</td>
<td>Rapid and complete recovery</td>
<td>5 days</td>
</tr>
<tr>
<td>2 Goyal et al[11]; 2002</td>
<td>63 yrs, F</td>
<td>progressive paraparesis, and bladder dysfunction</td>
<td>D 9–L2</td>
<td>No H/o trauma, systemic illness noted</td>
<td>DM-M</td>
<td>Rapid and Good recovery</td>
<td>3 months</td>
</tr>
<tr>
<td>3 Sharma et al[16]; 2004</td>
<td>10 yrs, F</td>
<td>progressive quadriaparesis</td>
<td>C4– D 2</td>
<td>No H/o trauma, systemic illness or any radiological abnormality noted</td>
<td>DM-M</td>
<td>Rapid and good recovery</td>
<td>1 month</td>
</tr>
<tr>
<td>4 Sharma et al[6]; 2005</td>
<td>4 yrs,F</td>
<td>quadriaparesis</td>
<td>C4–6</td>
<td>No trauma, systemic illness, No clinico-radiological abnormality</td>
<td>DM-M</td>
<td>Rapid and complete recovery</td>
<td>17 months</td>
</tr>
<tr>
<td>5 Ghannane et al[10], 2007</td>
<td>4 &amp; 8 yrs,</td>
<td>progressive paraparesis</td>
<td>D 3–4 (both patients)</td>
<td>No association described</td>
<td>DM-M</td>
<td>Complete recovery</td>
<td>NA</td>
</tr>
<tr>
<td>6 Guzel et al[12]; 2007</td>
<td>7 yrs, F</td>
<td>quadriaparesis</td>
<td>C2–4</td>
<td>H/o respiratory infection, H/o Trauma. No clinico-radiological abnormality mentioned.</td>
<td>DM-M</td>
<td>Rapid and near complete recovery</td>
<td>24 months</td>
</tr>
<tr>
<td>7 Lmejai et al[14]; 2008</td>
<td>12 yrs, M</td>
<td>progressive paraparesis</td>
<td>D 3–4</td>
<td>No documentation of trauma, systemic illness. No other radiological abnormality described.</td>
<td>DM-M</td>
<td>Rapid and complete recovery</td>
<td>4 months</td>
</tr>
</tbody>
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