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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 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,

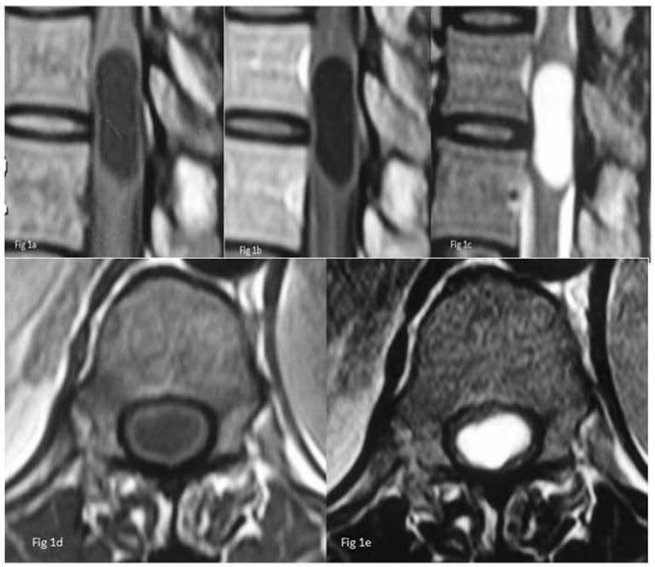

Figure 1: MRI of the dorsal spine.
Figure 1a, b: Sagittal T1W images pre and post contrast scan respectively depicting well defined, hypointense lesion at D11-12 with no contrast enhancement.
Figure 1c: Saggital T2W image shows a well defined hyperintese lesion at D11-12 vertebral level with no perilesional signal changes or syrinx.
Figure 1d & e: Axial T1W and T2W images shows well defined intramedullary which is hypointense and hyperintense respectively.

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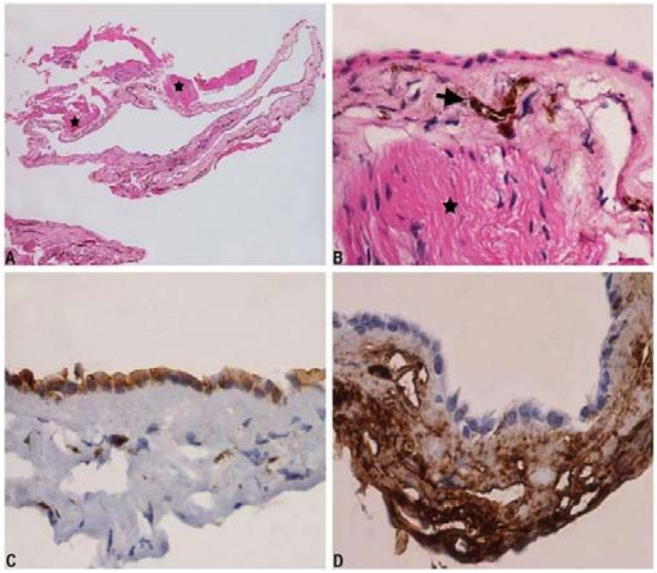

Figure 2: Histopathology and immunohistochemistry of intramedullary cyst wall.
A: Scanner view of the intramedullary cyst wall which is closely stuck to the cord tissue (star).
B: A higher magnification shows the characteristic flattened cuboidal arachnoid cells resting on a collagenous stroma which in turn is closely adherent to the neural parenchyma (star).
C: The lining epithelial cells express EMA.
D: The adjacent brain parenchyma shows reactive gliosis highlighted by GFAP immunostaing.
(A- HE stains objective 5 x; B- HE stain, objective 40 x; C-Immunoperoxidase against EMA, objective 40 x; D- Immunoperoxidase against GFAP, objective 40 x).

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 etio-pathogenesis 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 I 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.

Table 1: Literature review of spinal intramedullary arachnoid cyst.

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

8	Gezici et al[5]; 2008	35 yrs, F	paraparesis	D 5–T6	No h/o trauma,systemic illness, No other radiological abnormality mentioned	DREZ –M	Good recovery, ambulatory with little difficulty	3 years
9	Medved et al[15]; 2009	18 mos, M	progressive paraparesis, constipation	D 5–6	No H/o trauma, systemic illness,	DREZ myelotomy	complete recovery	1 month
10	Diyora et al[9]; 2010	45 yrs, M	Progressive neurological deficits	D 4-5	No H/o of trauma, systemic illness noted	DM-M	Gradual but Complete recovery	6 week
11	Kataria et. al[13]; 2012	9 yrs,F and 40 yrs,F	Progressive neurological deficits	Dorsolumbar	No H/o of trauma, systemic illness noted	DM-M	Complete recovery	1-6 months
12	Novegno et al [17; Ahead of print]; 2013	31 Yrs, F	Progressive paraparesis with bladder dysfunction	D11-12	No H/o of trauma, systemic illness noted	NA	Rapid and complete recovery	24 months
13	Present study	40 yrs, F	Paraplegia with bladder dysfunction	D11-12	No H/o trauma or systemic illness	DM-M	Rapid and Complete recovery (both motor and bladder)	50 months

DM-M: Dorsomedial Myelotomy; DREZ-M: Dorsal Root Entry Zone Myelotomy; NA: Not Available

However it remains a clinical speculation, as these are very rare and the natural history is still not clearly known. The available literature suggests a benign course and excellent recovery. There is no report of recurrence even with partial excision and/or fenestration, suggesting the importance of maintaining a free communication of the cyst with the subarachnoid space could be suffice, However long term follow up and MRI surveillance is warranted to follow the natural course of this disease and possible recurrence. With the increasing availability of the MRI and Cardiac-gated cine MRI (CMRI) for clinical screening, more cases of intramedullary cysts might be diagnosed and possibly studied in the future.

Our case highlights the imaging and histopathology of this lesion and the possibility of complete recovery from partial excision and fenestration. Similar experience from previous reported cases suggests that intramedullary arachnoid cyst really represent a distinct clinical variant and should be incorporated in the classification of spinal arachnoid cyst. It possibly does not represent a rare report of a case but rather represent a distinct though rare entity, which is further supported by the fact that increasing numbers of cases are being reported in last 10 years. More importantly, it is critical to realise that contrary to other intramedullary lesions, this intramedullary pathology is a treatable and has an excellent prognosis.

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