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Case Report

Conus medullaris intramedullary arachnoid cyst- case report and review of the literature

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ABSTRACT

Background: Spinal intramedullary cysts present a radiological dilemma. We present a rare case of a conus intramedullary arachnoid cyst and report on its differentiating features and management.

Case Description: We report a case of a 30-month-old child who presented with decreased gluteal sensation and urinary dribbling for 6 months. Apart from some slowness in walking, the power was normal in all four limbs. Imaging showed a non-enhancing, T2-weighted hyperintense 12 × 8 mm conus intramedullary cyst without any edema. A T12-L1 laminotomy followed by marsupialization of the cyst was done. Histopathology was suggestive of an arachnoid cyst. The postoperative course was uneventful with improvement in muscle strength and achievement of regular milestones. We also present the pertinent review of the literature to date.

Conclusion: Intramedullary arachnoid cysts are a rare entity and should form the differential diagnosis for cysts presenting in the conus medullaris. Simple decompressive options may suffice for symptomatic cases and radical excision may be avoided. A high index of suspicion is essential considering the subtle nature of presenting symptoms.

Keywords: Conus medullaris cyst, Intramedullary arachnoid cyst, Spinal arachnoid cyst

INTRODUCTION

Intramedullary cystic lesions comprise a small subgroup of spinal lesions. Their radiological isolation is difficult due to common morphology. This makes choosing the correct surgical approach difficult. Usually, the diagnosis of the intramedullary cystic lesion is done on intraoperative or histopathological grounds.[14] Intramedullary arachnoid cysts (IMACs) represent an extremely rare entity among the intramedullary cystic lesions, with only 24 cases reported so far in the English literature. Although reported at different spinal levels, they are commonly found in the dorsal region in both pediatric and adult age groups.[19] Cysts in the conus medullaris represent a distinct entity with respect to the peculiar clinical presentation due to the particular anatomy of the conus.[15] We report a 30-month-old symptomatic male child with an IMAC in the region of conus medullaris who underwent marsupialization of the cyst. We also review pertinent literature on IMAC.

CASE REPORT

We present a 30-month-old male child with a normal perinatal history who was referred to our center with a history of decreased sensation over the gluteal region along with persistent dribbling of urine for 6 months. Neurological examination was normal, except for decreased perianal sensations. He was able to walk but at a slower pace than before; however, the exact power could not be graded due to the age factor limiting communication. Magnetic resonance imaging (MRI) of the spine showed a well-defined intramedullary cystic lesion of size 12 × 8 × 8 mm in the conus medullaris at the T12-L1 level, which was hypointense on T1-weighted images and hyperintense on T2-weighted images with no perilesional edema, solid component, or contrast enhancement [Figure 1].

The patient underwent T12-L1 laminotomy under intraoperative monitoring. After a midline durotomy, the cyst was seen surfacing near the midline covered by a thin layer of gliotic tissue. The cyst was opened at its most superficial point and clear fluid akin to cerebrospinal fluid was drained. Repeated Valsalva maneuver confirmed that the central canal was separate from the cyst. There was no definite plane of dissection between the conus and the cyst. The cyst was marsupialized by suturing its wall to the arachnoid of the cord to prevent reclosure [Figure 2]. This was followed by a watertight dural closure. The postoperative period was uneventful. On follow-up after 3 months, there was an increase in muscle strength with improved walking but dribbling continued to persist. Histopathological examination revealed arachnoid cells in the cyst wall along with glial tissue which was suggestive of an arachnoid cyst.

DISCUSSION

Spinal arachnoid cysts are usually seen in extradural or intradural extramedullary locations; however, their intramedullary occurrence is rare. Reports of IMAC are few and to date, only 24 cases have been reported.[1-3,5-7,9-14,16-21,23-26]

Spinal arachnoid cysts are benign developmental cystic lesions. Their origin remains unclear; however, congenital, traumatic, and inflammatory causes have been postulated.[12,24,25] Among the few reports available, the most commonly cited cause of the origin of IMAC is secondary cystic changes in the atypical intramedullary arachnoid granulations probably formed during infolding of the dura.[8,11] One report attributes their origin to the diverticula of surface arachnoid at areas of less resistance along the spinal cord.[17]

According to our literature review including our case report [Table 1], IMACs are reported commonly in both adult and pediatric age groups, with 14 and 11 cases, respectively. A slight female preponderance was noticed, with 14 female and 11 male cases.

IMAC can become symptomatic due to compression of the spinal cord or nerve roots, causing weakness, sensory loss, pain, or bladder and bowel complaints. Conus lesions can present with local low back pain, saddle anesthesia, prominent bladder or bowel incontinence, and motor weakness at late stages. Our patient had decreased gluteal region sensation and urinary incontinence. According to the literature reviewed,

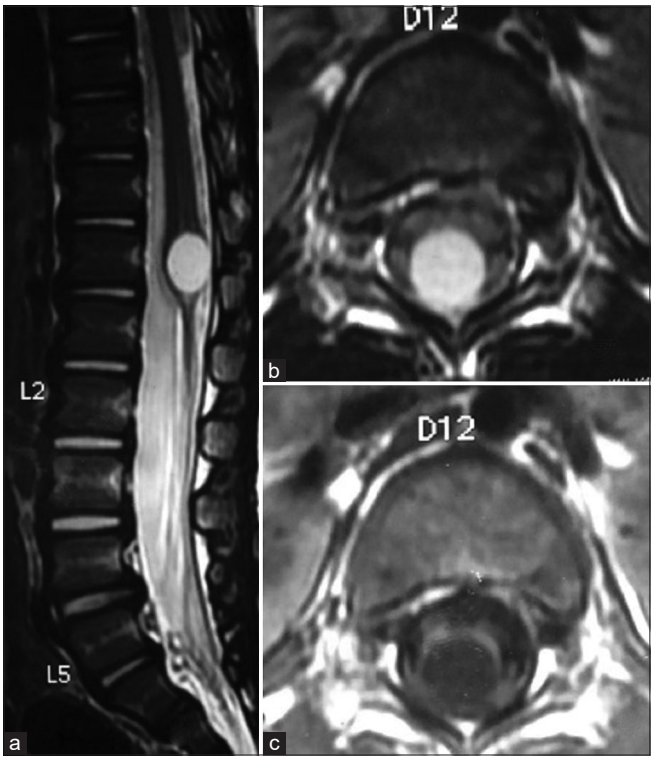

Figure 1: Preoperative (a) Sagittal T2-weighted, (b) Axial T2-weighted, (c) Axial T1-weighted dorsolumbar MRI showing an intramedullary arachnoid cyst in the conus medullaris.

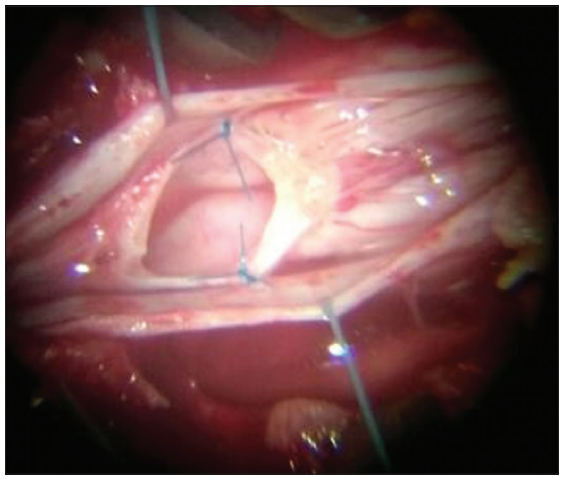

Figure 2: Intraoperative image showing cyst marsupialization by stitching cyst wall open edges to the arachnoid.

Table 1: Intramedullary arachnoid cysts-case reports.

Authors (Year of publication)	Age, Sex	Clinical presentation	Cyst location	Management	Follow-up	Outcome
Aithala *et al.*[1] (1999)	7 Y, M	Atypical abdominal pain, paraparesis, sensory loss	D-4	DM-myelotomy with fenestration	5 days	Rapid recovery
Goyal *et al.*[11] (2002)	63 Y, F	Back pain, paraparesis, bladder dysfunction	D9-L2	DM-myelotomy with partial cyst excision	3 m	Improved symptoms
Sharma *et al.*[24] (2004)	10 Y, F	Quadriparesis	C4-D2	DM-myelotomy and partial cyst excision	1 m	Improved symptoms
Sharma *et al.*[25] (2005)	4 Y, F	Quadriparesis	C4–6	DM-myelotomy with cyst excision	17 m	Complete recovery
Ghannane *et al.*[10] (2007)	8 Y, M	Paraparesis	D3-4	DM-myelotomy and partial cyst excision	8 m	Complete recovery
	4 Y, M	Paraparesis	D3-4	DM-myelotomy and partial cyst excision	6 m	Complete recovery
Guzel *et al.*[12] (2007)	7 Y, F	Quadriparesis	C2-4	DM-myelotomy with partial cyst excision	24 m	Complete recovery, except slight paresis of left UL
Lmejjati *et al.*[16] (2008)	12 Y, M	Paraparesis	D3-4	DM-myelotomy with cyst marsupialization	4 m	Complete recovery
Gezici and Ergün[9] (2008)	35 Y, F	Paraparesis, urinary incontinence	D5-6	DREZ-myelotomy with cyst excision and fenestration	3 Y	Remarkable recovery, walk with little difficulty
Medved *et al.*[17] (2009)	18 m, M	Paraparesis, constipation	D5–6	DREZ-myelotomy with partial cyst excision	1 m	Complete recovery
Diyora *et al.*[6] (2010)	45 Y, M	Back pain, paraparesis, bladder and bowel incontinence	D4-5	DM-myelotomy with partial excision	6 weeks	Complete recovery
Kataria *et al.*[14] (2012)	9 Y, F	Back pain, paraesthesia, urinary disturbance.	Conus medullaris	DM-myelotomy and partial excision	6 weeks	Complete recovery
	40 Y, F	Paraparesis, urinary disturbance	Conus medullaris	DM-myelotomy and partial excision	4 weeks	Complete recovery
Rahimizadeh and Soufiani[20] (2013)	58 Y, F	Quadriparesis	C6-D2 lesion +C3-C7 spondylosis	DREZ myelotomy and partial cyst excision+C3-D2 instrumentation	3 Y	Symptomatically well
Novegno *et al.*[18] (2014)	31 Y, F	Back pain, paraparesis, bladder dysfunction	D11-12	DM-myelotomy and cyst fenestration	26 m	Complete recovery
Ranjan *et al.*[21] (2014)	40 Y, F	Paraplegia, bladder dysfunction	D11-12	DM-myelotomy with partial excision	50 m	Complete recovery
Alugolu *et al.*[3] (2016)	54 Y, F	Backache, LL weakness, paraesthesia	D9-11	DM-myelotomy with cyst decompression	1 Y	Resolution of symptoms
Thakar and Hegde[26] (2016)	64 Y, M	Quadriparesis	C6-D1	DM-myelotomy with excision	NA	NA
Baeesa *et al.*[5] (2017)	47 Y, F	Neck and UL pain, UL weakness, bladder and bowel disturbance	C3-5	DREZ-myelotomy and CSA shunt	5 Y	Complete recovery, No radiologic recurrence
Fonseca and Ratilal[7] (2017	49 Y, F	Neck pain, UL paraesthesia	C 6-7	Conservative with imaging follow-up	5 Y	Neurologically and radiologically stable
Panwar *et al.*[19] (2019)	45 Y, F	Back pain and paraesthesia.	D9-11	DM myelotomy and cyst decompression.	11–12 m	Recovered.
	40 Y, M	Back pain, paraesthesia, weakness.	Conus medullaris	DM myelotomy and cyst decompression.	11–12 m	Recovered, Radiological recurrence

(Contd...)

Table 1: *(Continued)*.

Authors (Year of publication)	Age, Sex	Clinical presentation	Cyst location	Management	Follow-up	Outcome
Shaaban *et al*.[23] (2019)	31 Y, M	Heaviness and vague sensation in lower trunk and LL	D6-8	Cyst biopsy and DM-myelotomy with syrinx decompression	50 days	Improvement in symptoms
Ichinose *et al*.[13] (2020)	4 Y, M	Paraparesis, bladder and bowel disturbance	C3-4	DM-myelotomy with cyst fenestration-dorsal and ventral.	1 m	Cyst and symptoms recurred.
				Observation	1–27 m	Symptoms improved, lesion stable radiologically.
				Re-fenestration and excision of cyst wall and scar	4 m	Clinical and radiological improvement
Present study	30 m, M	Decreased perianal sensation, bladder disturbance,	Conus medullaris	DM-myelotomy and marsupialization	3 m	Motor and sensory improvement, urinary dribbling persisted

Y: Year/s, m: Months, M: Male, F: Female, UL: Upper limb, LL: Lower limb, DM: Dorsomedian, DREZ: Dorsal root entry zone, CSA: Cysto-subarachnoid

limb weakness was the most commonly reported symptom. Specifically in the pediatric age group weakness was the most common presentation, whereas, in the adults, the most common symptom was back or neck pain.

Like in the previous reports, the dorsal cord is the most common site reported for IMAC in both the pediatric and adult age groups in our literature review as well.[3,18,19] Three cases of conus IMAC have been reported previously.[14,19] Similar to our patient, other reported arachnoid cysts which were located in conus presented with weakness, saddle anesthesia, and decreased anal tone. They usually produce a radiological dilemma, with the diagnosis being made intraoperatively or on histopathology.

MRI with contrast is the investigation of choice for diagnosing IMAC. They are characterized by non-enhancing homogenous well-circumscribed cysts, hypointense on T1-weighted images, and hyperintense on T2-weighted images, without any surrounding edema. The radiological differential diagnosis for IMAC includes primary or secondary syrinx, epidermoid cyst, cystic astrocytoma, teratoma, ganglioglioma, dermoid, and meningomyelocele, cystic lesions such as neuroenteric, neuroglial, ependymal, and parasitic cysts.[4] However, it is usually not possible to differentiate on imaging between an arachnoid cyst, focal syrinx, and terminal ventricle.[14] In some of the reports, IMAC was seen to be eccentrically located, which can be a differentiating factor from focal syrinx or ventriculus terminalis. The location of ependymal cysts may be anterior in the cord, while neurenteric cysts may be associated with other congenital anomalies.[18] The absence of any solid component and any enhancement rules out tumorigenic or infective cysts. Surrounding edema is absent in IMAC but may be seen in inflammatory causes.

Treatment is reserved for symptomatic lesions. Management options range from percutaneous aspiration to surgical fenestration, myelotomy with partial or complete removal of cyst wall, or placement of cysto-subarachnoid shunt.[5,24,27] Most authors recommend wide fenestration of the cyst with excision of as much of its wall as possible.[1,11] Myelotomy can be made with relation to the location of the lesion, usually over the surfacing component along with judicious use of intraoperative monitoring.[3] Most authors use dorsomedian myelotomy approach. However, Gezici and Ergün reported a thoracic IMAC treated by bilateral dorsal root entry zone (DREZ) myelotomy.[9] They propose DREZ myelotomy to be advantageous over dorsomedian myelotomy, as it leads to a reduced incidence of dorsal column sensory loss, surgery-related granulation and fibrosis, and a longer duration of patency of the fenestration.[9] In our case, we chose the most surfacing part of the lesion for myelotomy, which was essentially a dorsomedian myelotomy. DREZ myelotomy may be a good alternative for eccentric lesions as it causes less neural injury mainly to the undisturbed dorsal tracts near the midline.[9]

In IMAC excising the cyst wall is usually difficult due to its adherence to the cord parenchyma. Radical excision can be circumvented with simple fenestration or decompression with marsupialization.[22,25] We also advocate arachnoidal suturing of the cyst wall to prevent reclosure and recurrence; however, more research is required to substantiate this hypothesis. The majority of the case reports show improvement of symptoms with such simple procedures rather than radical excision.

On histopathology, the presence of arachnoid cells is the key for differentiating radiologically similar lesions such as ependymal and neurenteric cysts, which have ependymal and endodermal lining respectively, whereas neuroglial cysts have an absence of any arachnoid cells and tend to have a poor capsule.[8,18]

Although most reports emphasize surgery for symptomatic cases, Fonseca and Ratilal demonstrated that clinical observation can be done in mild or asymptomatic cases.[7] The late presentation of symptoms in some adult cases can be used as a reference in support of observation of mild or asymptomatic patients. However, the literature is limited and we can only speculate about the natural history of IMAC at this point.

Most of the operated cases show complete recovery on short-term follow-up along with radiologic resolution of the cyst on follow-up. However, radiologic recurrence of the cyst has been reported in two cases so far, with one of them becoming symptomatic. The reasons speculated for recurrence were incomplete excision of the cyst, an unnoticed ventral subarachnoid channel, postoperative adhesion, and fibrosis.[13,19] Zekaj *et al*. reported a case of development of an IMAC on follow-up after near-total removal of an intradural extramedullary cyst, with a small part of cyst wall left behind.[28]

CONCLUSION

IMACs are a rare entity and should form the differential diagnosis for cysts presenting in the conus medullaris. Simple decompressive surgical options for symptomatic lesions may suffice and radical excision may be avoided.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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