

Interesting Case Reports of Neurenteric Cyst with Type 1 Spilt Cord Malformation at Conus Medullaris

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Abstract

Two rare cases of split cord malformation with neurenteric cyst are presented. The clinical, radiological and surgical findings are presented with brief review of literature. Co-existent neurenteric cysts along with SCM type 1 have been described very rarely in the literature.

Keywords: Lytic lesion of skull; Calvaria; Eosinophilic granuloma; Fibrous dysplasia

Introduction

Intraspinal neurenteric cysts are rare and account for only 0.3-0.5% of all spinal tumours. The first histologically confirmed endodermal cyst was reported by Puusepp in 1934 [1]. Spinal neurenteric cysts are infrequently reported congenital abnormalities that are believed to be derived from an abnormal connection between the primitive endoderm and ectoderm during the 3rd week of life. Neurenteric cysts are not confined to the spinal column and may be found within the brain, mediastinum, abdomen, pelvis, or even in a subcutaneous location. Split cord malformations (SCMs) are uncommon and usually diagnosed in children, although up to 20% may present in adulthood. Pain and progressive sensory-motor deficits are the most common complaints, and lower thoracic and lumbar regions are the most frequent locations. Diastematomyelia and neurenteric cyst are congenital anomalies and seem to arise from the same embryologic error. Their association is very rare [2]. We present two interesting cases of diastematomyelia associated with the neurenteric cyst at the same level at conus region.

Case Report 1

A twenty year old female presented to us with history of progressive non radicular low backache of six months. Pain was aggravated on bending forwards and walking and pain was relieved on taking rest. No h/o sphincter involvement or motor weakness.

On local examination, there was pigmented nevus at the L5-S1 level of 3 cm diameter.

Neurological examination was unremarkable.

MRI dorso lumbar spine showed intradural extramedullary lesion at D12 and L1 level which was well circumscribed, hyperintense on T1 and isointense in T2 w imaging, with no fat suppression s/o lipoma with split cord (Figure 1).

CT scan dorsolumbar spine confirmed diastematomyelia with bony spur at d12-L1 with hemicords.

Urodynamic study was normal (Figures 2 and 3).

Patient underwent surgery

Laminectomy of D11-L2 was performed bony spur at D12-L1 was excised, Oval shaped grayish cystic lesion was seen between the two spilt cords extending above and anterior to the nonspilt cord with arachnoid adhesions between the cyst and the cord Thick yellowish

fluid was aspirated from the cyst, and total excision of cyst was done (Figures 4 and 5).

Case Report 2

A 42 year old lady presented with h/o low backache with radiat-



Figure 1: MRI LS T2 w imaging spine sagittal and axial showing bony spur at level of L1 extending from posterior elements to anteriorly towards the body splitting the cord into two with intradural cystic lesion s/o dermoid cyst.

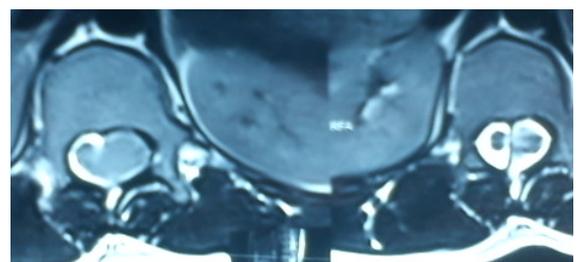


Figure 2: MRI DL spine axial section T2 W showing split cord with intra dural cyst which is hypo intense compressing the cord.

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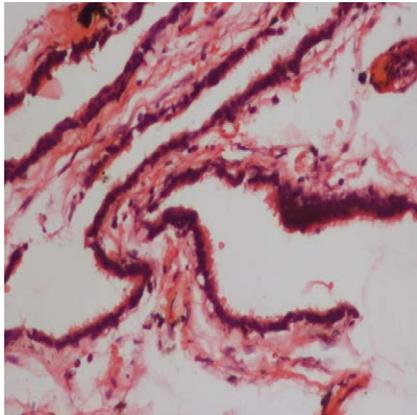


Figure 3: CT scan spine showing bony spur splitting the spinal canal into two.

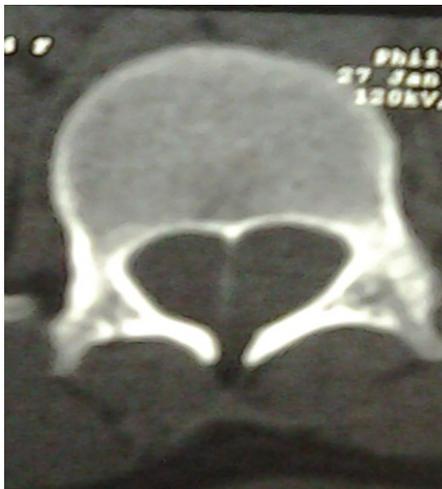


Figure 4: Pre-Operative photos showing cyst seen between the split cords.

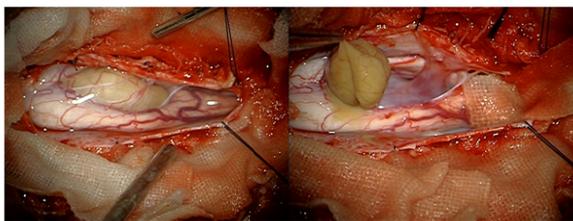


Figure 5: Patient improved symptomatically after surgery. Histopathology was suggestive of neuroenteric cyst with squamous metaplasia

ing pain to right groin and anterior aspect of thigh for period of three months prior to admission, patient also had gross spasticity with sensorimotor deficits and micturition involvement.

She was wheel chair bound

Clinical examination: tuft of hair was seen in the midline in lumbosacral region with deformities and trophic changes in both feet.

There was spastic paraparesis with spasms associated with loss of sensations from below L1

Investigations

- MRI dorso lumbar spine showed diastematomyelia at L1/2 with

bony spur and a well-defined intra medullary cystic lesion at D10-11 level thought to be a syrinx

- CT spine confirmed bony bar at L1/2 extending from body to posterior elements

• Patient underwent D12/L1/2 laminectomy and total excision of bony bar was performed, dural tubes were unified at level of L1, cord was lax and syrinx was not addressed

• There was significant improvement post operatively with patient becoming gradually ambulant but was complaining of persistence of spasticity at three months of follow up, MRI spine was repeated which showed persistence of Syrinx or cyst above the bony spur.

- CT spine showed complete excision of bony spur

• Re exploration was undertaken D10/11 laminectomy was performed.

• Dura was tense, on durotomy, cyst was seen splitting the cord into two. Capsule of cyst was thinned out and cyst was aspirated which showed yellowish fluid, piece meal excision of cyst was done.

- Post-surgery, spasticity came down significantly.

- HPE was suggestive of neurenteric cyst.

Discussion

Neurenteric cyst is classically reported as a solitary lesion in the cervical region, located anterior or anterolateral to the cord. The bony abnormalities that may be associated with neurenteric cyst are likely to involve the anterior column; there is a slight male predominance of 1.8:1 with the most common location of the cyst being intradural, extramedullary, and anterior or anterolateral to the cord [1].

In the cases we are reporting here neurenteric cysts are seen in the conus region and the cysts were in dorsal region, in both the cases the cyst was seen between the split cords and at the same level as that of the bony spur. Another notable feature is that our second case patient presented in fifth decade of life with severe myelopathy.

Possible theory of rare association of type 1 spilt cord with neurenteric cysts was explained by Pang et al. in unified theory of embryogenesis

According to this unified theory of pang the basic ontogenetic error is the formation of an accessory neurenteric canal that passes through the embryonic disc and maintains communication between the yolk sac and the amniotic cavity. This abnormal fistula causes localised splitting of the notochord and the overlying neural plate. As the abnormal fistula is forming the surrounding mesenchyme condenses around it. Together with the 'pinch' of endoderm protruding from the base of the fistula, the cuff of mesenchyme forms an endomesenchymal tract that invests the fistula and occupies the space between the split notochord and the split neural plate [3]. This fistulous communication may evolve in many ways.

(1) The mesenchyme that condenses around the fistula eventually may develop into a fibrous or osseocartilaginous septum

(2) Splitting of the neural plate into two halves results in the formation of two hemicords.

(3) The endoderm lining the fistula may differentiate into cysts. These cysts are made up of respiratory and gastrointestinal epithelium (neurenteric cysts), both being derivatives of the endoderm.

Although the fistula is lined by endoderm, the rarity of associa-

Cases	Author	Age/Sex	Diagnosis
1	Walsh et al. [4]	4 year male	L2-L3 Diastematomyelia with subcutaneous lipoma with cyst located at tapered end of lipoma
2	Mann et al. [5]	50 year male	L3-L5 Diastematomyelia with discharging dermal sinus, cyst at L5 level ventral to cord
3	Whitney et al. [6]	56 years male	D7 diastematomyelia with thoracic scoliosis(incidentally found)
4	Kaffenberger et al. [7]	38 years male	L3 diastematomyelia with hypertrichosis with cyst being dorsal to cord at L3 vertebral level
5	Pang et al. [8]	Aborted male foetus	C4-C6 Diastematomyelia with cyst, dorsal to cord at C6 vertebral level
6	Birsch et al. [9]	48 years female	High cervical diastematomyelia(C1) with cyst at Midline ventral to cord at C1 level
7	Prasad et al. [10]	11 years male	cervico thoracic meningocele with diastematomyelia with cyst posterior to cord at C6-T1 vertebral level patient was operated in neonatal period
8	Muthukumar et al. [3]	40 days female	D4-L1 diastematomyelia with dermal sinus with bony spur at D9-D12 level, cyst at D6-D7 level ventral to cord.
9	Soni et al. [2]	30 year, Female Postpartum	D6-7 diastematomyelia with cyst present dorsomedially

Table 1: Describes previously reported cases.

tion of split cord malformations with neurenteric cysts may be due to the absence of the 'inducer' molecules for intestinal epithelia near the notochord and neural tube. However, when the endoderm does undergo differentiation, a neurenteric cyst will form in close association with the split cord as in our cases.

According to Sensenig, the normal spinal cord dura matter develops from meninx primitiva, which appear at 21-30 days of gestational age. So, if endomesenchymal tract forms before 21 days then SCM-I (diplomelia) will occur, if after 30 days then SCM II (diastematomyelia), and between 21-30 days then composite variety will occur [1]. After review of literature, to our knowledge only 9 cases of neurenteric

cyst with type 1 spilt cord malformation have been reported so far.

Table 1 throws light on previously reported cases.

Conclusion

Although neurenteric cysts are rare, they should be considered in the differential diagnosis of intradural extramedullary mass lesions in presence of spilt cord malformations.

Association of such should be kept in mind when atypical imaging presents to clinician.

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