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CASE REPORT

A rare case of solid calcified intramedullary neurenteric cyst: Case report and technical note

Krishna Chaitanya Joshi, Daljit Singh, Sudhir Suggala, Tushit Mewada
Department of Neurosurgery, G B Pant Hospital, New Delhi, India

ABSTRACT

Neurenteric cysts are rare lesions of the spinal cord, constituting less than 1.3% of all spinal cord tumors. Approximately 90% of neurenteric cysts are located in the intradural extramedullary compartment, while the remaining 10% are divided between an intradural intramedullary or extradural location. They are usually extramedullary and found in the lower cervical and thoracic spine. We report a case of 8 year old child with totally solid calcified intramedullary cyst which has hitherto not been reported in literature. We discuss the challenges faced in the diagnosis and surgical management of such lesions and a brief review of pertinent literature.

Key words: Enterogenous cyst, intramedullary lesions, myelotomy, ziploc technique, neurenteric cyst

Introduction

Neurentric cysts are thought to embryologically originate from an abnormal communication between the primitive neurenteric canal, notocord, and neural tube to the adjacent endoderm and mesenchyme during the 3rd week of embryogenesis and account for 0.7-1.3% of all spinal cord tumors.[1] They are lined by nonciliated simple, pseudostratified and cuboidal or columnar epithelium. The male: female ratio is 2:1, most frequently seen in the second and third decades of life. In pediatric age group, the mean age of presentation is 6.4 years.[2] Approximately, 90% of neurenteric cysts are located in the intradural extramedullary compartment while the remaining 10% are divided between an intradural intramedullary or extradural location. They are usually found in the lower cervical and thoracic spine.[3]

These lesions are usually isointense on T1-weighted images and hyperintense on T2-weighted imaging and display minimal or no enhancement on T1-weighted postcontrast imaging. In this paper, we report a rare case of a solid calcified intramedullary neurenteric cyst hitherto not reported in the literature and discuss the challenges faced in the diagnosis and treatment of this entity.

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Address for correspondence:
Dr. Krishna Chaitanya Joshi, Department of Neurosurgery, G B Pant Hospital, New Delhi - 110 002, India.
E-mail: krishnacjoshi@yahoo.com

Case Report

An 8-year-old male presented with progressive weakness in both legs since 3 years. He had numbness and paresthesia extending from his knees to his feet on both sides and had no symptoms suggestive of bladder and bowel involvement. On neurological examination, he had no cranial nerve deficits, motor power and sensation were normal in both upper extremities. He had bilateral lower extremity weakness; power was Medical Research Council grade 3/5 in both dorsiflexors of the ankle and 4/5 in all other muscle groups in both the legs. His reflexes were brisk at the knees and ankles with wide-based gait. Babinski sign was present on both sides and no sensory loss.

Contrast magnetic resonance imaging of the spine was performed which showed intramedullary lesion occupying the central cord from D3 to D6 that was isointense to the cord on T1-weighted and mildly hyperintense on T2-weighted with multiple areas of central hypointensity, probably representing

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calcifications, with minimal contrast enhancement in the periphery [Figure 1]. There was no associated vertebral anomaly.

A laminotomy was performed from D3 to D6. The dura was opened from D3 to D6, and the spinal cord was visualized with an intraoperative microscope. The cord appeared to bulge in the midline with absent cord pulsations. The pia was incised in the midline and retracted using 6-0 prolene sutures. A midline myelotomy using a fresh number 11 knife was performed; hard capsule was encountered within the cord. The lesion resembled a white pebble, it was not adhered to the cord, and it was carefully dissected out and was delivered *en bloc* [Figure 2]. After extraction of the lesion we closed the pia using a novel technique by approximating the free edges of the pia by crushing them between the interdigitating teeth of a dural forceps, in a manner resembling a Ziploc bag (which is a type of resealable storage bag). The dura was closed primarily and wound closed in layers. Intraoperative neurophysiological monitoring was done during the procedure.

In the postoperative period, the patient showed remarkable improvement in power in lower limbs in 3 months. Histopathological examination revealed cyst lined by pseudostratified ciliated columnar epithelium. Few goblet cells were also seen. Fibrocollagenous tissue was present along with areas of dense calcification [Figure 3].

Discussion

Intramedullary neurenteric cysts are thought to represent 12.2% of all neurenteric cysts. In pediatric age group, differential diagnosis of intramedullary lesions are astrocytomas, dermoid, epidermoid cyst, hemangioblastoma, ependymomas, and syringohydromyelia. The astrocytomas are eccentric and show infiltration into the surrounding cord on imaging. Dermoid and epidermoid cysts are hypointense on T1-weighted and hyper or iso intense on T2-weighted. Hemangioblastomas usually have cyst with enhancing mural nodule and flow voids whereas syringohydromyelia have CSF like intensity on T2-weighted. Ependymomas are characterized by cystic lesions with or without a necrotic component with not infrequent eccentric growth pattern.

Various approaches have been described for intradural extra medullary cysts that include anterior, posterior, lateral, and percutaneous aspiration. But till date, no consensus is present for the approach and completeness of removal in intramedullary cysts.[4] Complete resection of the cyst is recommended whenever safely possible.

Since the patient is in the developing age group and required three level exposure a laminotomy in place of laminectomy was done, which is highly recommended.[5] Intraoperative use of an operating microscope helped to confirm the location of the tumor by noticing pointers like flattening of the cord and decreased vascular markings at the level of the lesion. A clean

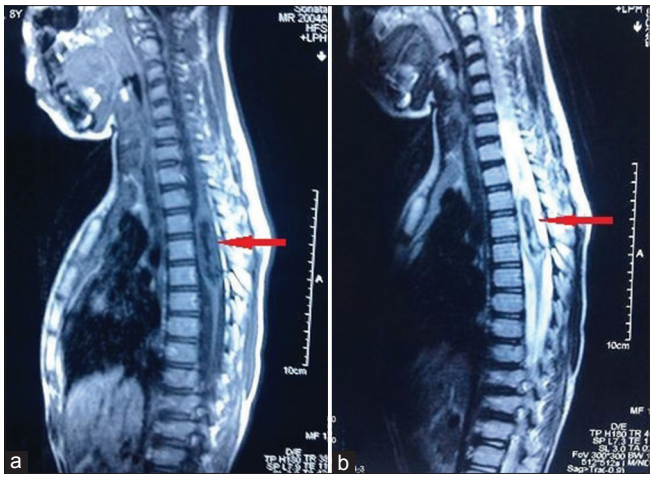

Figure 1: (a) T1-weighted image showing isointense lesion (b) T2-weighted image showing hyperintense lesion with areas of hypointensity

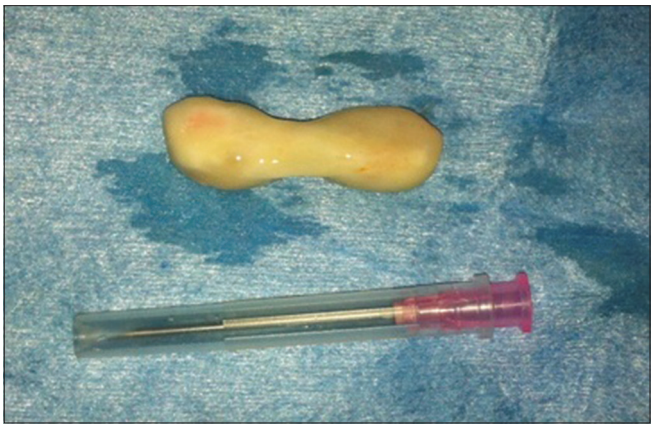

Figure 2: Image showing the excised specimen

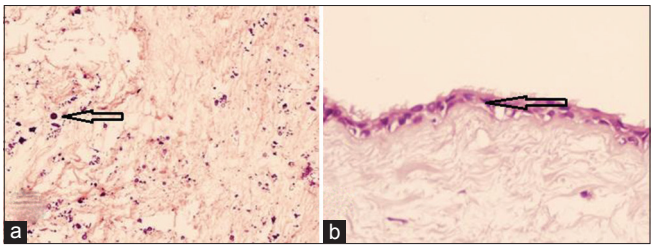

Figure 3: Microphotograph showing (a) calcification and (b) goblet cell lining

cut myelotomy using a fresh number 11 knife reduces the chances of contusion of the adjacent cord. Since the lesion was large, and consistency was hard we gently freed the margins and the tumor delivered itself without the use of any retraction or coagulation, which if used could have seriously compromised the chances of a smooth postoperative recovery.

Of note in our surgical technique was the innovative and novel method closing the pia after the surgery to restore normal anatomy. After the extraction of the tumor, the cord became very lax, and there was redundant pia. The interdigitating teeth of the dural forceps were used to approximate the pia

which snugly held on to each other and beautifully restored the normal contour of the cord. This reduces the cumbersome suturing of the pia and reduces the cord handling.

Conclusions

Neurenteric cysts represent a small percentage of spinal tumors, solid calcified intramedullary neurenteric cysts can masquerade other more common developmental lesions like dermoids. This is the first reported case of totally solid, intramedullary neurenteric cyst in the thoracic cord. These lesions can be a diagnostic challenge to the neurosurgeon and the pathologist. Intramedullary neurenteric cysts can be completely excised with a good clinical outcome. Intraoperative electrophysiological monitoring and minimal use of retraction and coagulation, with meticulous closure of pia and restoration of normal anatomy portend a good outcome.

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Conflicts of interest

There are no conflicts of interest.

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