



Case report

Long segment spinal intramedullary dermoid cyst: A case report

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ABSTRACT

Introduction: Spinal intramedullary dermoid is very rare, accounting for <1% of intraspinal tumors. It can be congenital or acquired. They usually present in 2nd or 3rd decade of life in adults. It may or may not associated with spinal dysraphism. It is asymptomatic in most cases, manifest acutely if it ruptures.

Long segment involvement of spinal intramedullary tumor in adult without the history of trauma makes this case unique. Fat suppression imaging helps to distinguish adipose tissue from lesions causing hematomyelia in patients presented with intramedullary hyperintensity in both T1 & T2 sequences of MRI spine.

Case presentation: We report here a rare case of 30 years old male who presented to us with sudden urinary retention followed by rapidly progressive quadriparesis and paresthesia in his right arm. In MRI, spinal intramedullary tumor was noted from medulla till D5 vertebra. We performed subtotal excision of tumor and sample sent for histopathology which proved it to be intramedullary spinal dermoid cyst.

Conclusion: Long segment involvement of spinal intramedullary dermoid cyst in adult without history of trauma makes this case different. Fat suppression imaging must be done in patients with intramedullary hyperintensity in both T1 and T2 sequences of MRI spine. Early diagnosis and appropriate management will be helpful in reducing morbidity.

1. Introduction

Long segment spinal intramedullary dermoid cyst is very rare, accounting for <1% of intraspinal tumors [1]. It can be congenital or acquired. Congenital form occurs due to intrauterine defective closure of neural tube. Acquired variety occurs after trauma or surgery. Spinal dermoid grows slowly. They usually present in 2nd and 3rd decade of life in adults [2]. It mostly occurs in lumbosacral area (60%), cauda equina (20%), thoracic spine (10%) or cervical spine (5%) [1]. It is associated with spinal dysraphism [3]. They are asymptomatic in most cases, presents with progressive sensorimotor deficits or defect in bowel or bladder control, manifest acutely if it ruptures. Mortality and morbidity increase in case of rupture.

Hyperintense spinal lesion on both T1 & T2 MRI can either be due to fat or blood (hematomyelia). Hematomyelia may be due to trauma, vascular malformations (arteriovenous malformation (AVM), cavernous hemangioma, angiomas), intramedullary tumors (ependymoma, epidermoid), spinal cord metastases, radiation therapy, anticoagulant therapy, previous spinal instrumentation due to spinal surgery or lumbar puncture, coagulopathy, dural arteriovenous fistula (dAVF) [4].

Lesions containing fat could be intramedullary dermoid cyst or intradural lipofibromatosis.

Fat suppression imaging is used to suppress adipose tissue. This helps us to distinguish whether the hyperintensity noticed in both T1 & T2 sequences is due to lesion containing fat.

One case of long segment intramedullary dermoid was reported in India in 2009 with one segment extending from the base of the skull to the D1 level, the second from D2 to D11, and the third segment from D11-D12 to L2, in the conus medullaris [2].

Shukla and others reported in 2019 with multifocal intramedullary dermoid extending from D4 to L1–2 which was partially debulked by surgery [5].

Another case was reported in 2020 of multifocal heterogeneous intramedullary masses extending from C2 to T4 and T12–L1 which was resected sub totally [6].

We are reporting here congenital spinal intramedullary dermoid cyst extending through medulla, cervical and thoracic spinal cord presented in our academic practice setting. We performed gross total excision and had done histopathology examination with result dermoid cyst. Written consent has been received from the subject. The authors declare no

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conflicts of interest. This work has been reported in line with the SCARE criteria [7].

The aim of this case report is to highlight the importance of fat suppression imaging in diagnostic assessment and pre-operative planning and management of patients presenting with intramedullary hyperintensity in both MRI (T1 & T2) sequences.

2. Case history

A 30-year-old man had history of urinary retention 2 weeks ago for which he was treated at Basic health unit with foley's catheter and discharged home. After two days, he started to develop weakness of all four limbs and numbness in right arm. He was then admitted in Neurology department at tertiary care hospital with these complaints. Initial MRI was done there, and they made provisional diagnosis of intramedullary cervical spine hemorrhage. He was treated with steroid and his weakness improved considerably. He was then shifted to neurosurgical ward for emergency decompression and evacuation of intramedullary hemorrhage. He had history of pulmonary tuberculosis 9 years back for which he took anti-tuberculous therapy. He did not take anti-coagulants. There was no history of spinal trauma, lumbar puncture, previous spinal surgery or radiation therapy. There was no history of bleeding disorders. There was no history of tumor in family. He was laborer by occupation.

On examination he was vitally stable, conscious, and oriented. He showed signs of central cord syndrome with mixed upper and lower motor neuron (LMN) signs in upper limbs and upper motor neuron (UMN) signs in lower limbs.

Right upper limb weakness (MRC 4 on right, 5 on left) [8], absent upper limb reflexes on Right & 1+ on left, variable sensory findings (absent temperature sensation on right, hypoesthesia on right side to

crude and fine touch, lost two-point discrimination on both sides).

Reduced Power (MRC grade 4 on right & 5 on left), hypertonia in both lower limbs (Modified Ashworth Grade 1 on right, 2 on left) [9], hyperreflexia (Knee & Ankle reflexes- 3+ on both sides), upgoing Plantar, lost two-point discrimination, intact pain, crude touch, temperature & joint proprioception in both lower limbs.

There was no tuft of hairs at lower back or dermal sinus.

3. Diagnostic assessment

Hematologic reports were normal. Coagulation profile was normal. Initially MRI spine was done for intramedullary hemorrhage, thinking it to be due to hematomyelia [especially arteriovenous malformation (AVM), Dural arteriovenous fistula (dAVF)]. CT angiography spine was done for AVM/dAVF, but it came out to be normal. So, we decided to go for fat suppression MRI, keeping in mind the possibility of intramedullary dermoid or intradural lipofibromatosis which showed suppression of adipose tissue (Fig. 1). On radiography, spinal dysraphism was ruled out.

4. Surgery

After informed consent from patient, definitive surgery was planned. This procedure was performed by neurosurgical consultant. Under general anesthesia, patient was placed in prone position. Skin incision was marked from C2 to D4. Skin, subcutaneous fat, fascia and muscle separated. Laminectomy from C3 to D3 was done. Meningeal layers (dura & arachnoid) were not showing any signs of inflammation (thickening, opacification, or sugar coating) which ruled out spinal infections (Fig. 2). Dura opened. Paramedian Myelotomy done. Cheesy



Fig. 1. (A) T1C (Contrast Enhanced T1 sequence showing hyperintense intramedullary signal extending from medulla up till visualized cord D5); (B) T2 showing same hyperintensity with occasional hypointense flakes (calcifications?); (C) T2 Fat suppression (STIR) sequence showing change of hyperintensity to hypo intensity suggesting intramedullary dermoid.

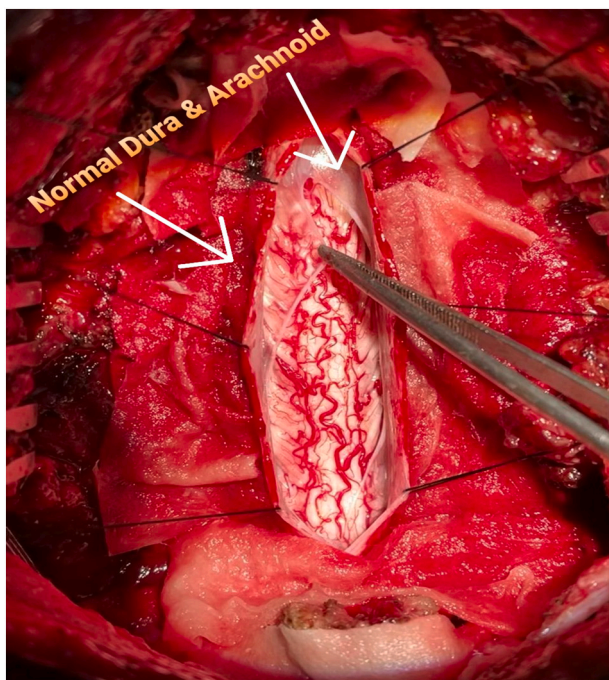


Fig. 2. Normal dura and arachnoid.
(Note: No thickening, opacification or sugar coating).

caseous thick yellowish material evacuated. Remaining lesion was gently washed with saline using 7Fr NG tube. Subtotal resection was performed. Dura closed in watertight manner. Wound was sutured layer by layer. Sample sent for histopathology.

5. Histopathology

Histopathological examination shows fragments of glial tissue and fibroconnective tissue with fibrosis and mild lymphohistiocytic inflammatory infiltrates. No granuloma or malignant cells seen (Fig. 3).

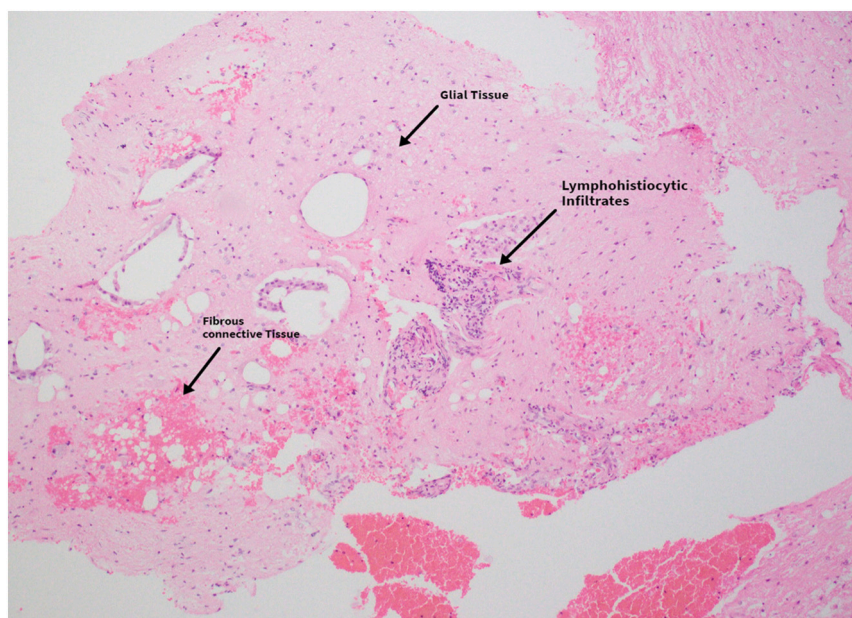


Fig. 3. Histopathologic H&E stained 10× view.

6. Post-operative status

Postoperatively patient stayed for 1 week in hospital. Patient was referred to medical rehabilitation center for physiotherapy for improving his quality of life. We also advised the patient to follow in neurosurgical clinic and medical rehabilitation center. Patient was assessed after 2 weeks according to Modified McCormick's score which was IV [10]. After 6 weeks, patient's score was 2. Patient told that his paresthesia settled and weakness and spasticity improved after surgery.

7. Discussion

Intraspinal dermoid is a benign, slow growing tumor, can be congenital or acquired, accounts for <1% of intraspinal tumors [1].

Congenital form is associated with spinal dysraphism [3], occurs due to defective closure of neural tube during embryogenesis (3rd to 5th week of fetal life) [5]. Acquired form occurs after trauma or surgery. In this patient however, the tumor was reported to occur in adult phase of life and there was no previous history of trauma or surgery. This indicates the atypical dermoid cyst case.

Both genders are equally affected. They predominantly occur in 2nd or 3rd decades of life in adults and are most common in children aged less than 20 yrs. (mostly in less than 1 year of age) [2]. Common locations of dermoid cysts are scalp, skull, intracranial, and intraspinal, especially intradural extramedullary; rarely intramedullary and associated with other spinal defects [11]. In spine, most common location is lumbosacral region (60%), followed by cauda equina (20%), then cervical or thoracic spinal cord (5%). Holocord lesion is very rare. In our report, patient was 30 yrs. old male with dermoid cyst extending from medulla, whole cervical spine, and dorsal spine till D5.

Most cases are asymptomatic, symptoms vary according to location, ranging from progressive motor or sensory deficits, loss of bowel or bladder control. It may present acutely after rupture. Our patient presented with classical symptoms (urinary retention, rapidly progressive quadriparesis and paresthesia).

Other case reports have multifocal unruptured long segment intramedullary dermoid lesion either involving cervicothoracic or cervicothoracic and conal lesion. Some had signs of spinal dysraphism. In our case report, long segment involvement of spinal intramedullary dermoid cyst (from medulla till D5) presenting in 3rd decade with central cord

symptoms, makes it unique.

Treatment modalities for intramedullary dermoid depends upon symptoms, extent of lesion and anticipated post-operative neurologic deficit. Conservative management is advised for patients who are asymptomatic [12]. Biopsy, partial or subtotal resection is advised for symptomatic patients in which gross post-operative neurologic deficit is likely to occur [12,13]. Gross total resection must be done for smaller lesions. Treatment of choice is maximal tumor resection at early stage. Dermoid cysts can rupture during surgery, after trauma, or spontaneously. This can cause the spread of content along the subarachnoid and ventricular spaces which causes aseptic chemical meningitis [14]. In this case there were no postoperative chemical meningitis, after removal of intramedullary dermoid cyst.

Kane et al. published a series of 54 patients with intramedullary spinal cord tumors treated with surgery reported that adjuvant post-operative radiotherapy was not used routinely if complete tumor removal was achieved, it can be concluded that the effectiveness of radiotherapy is still unclear and has not been demonstrated [15]. In this patient no postoperative radiotherapy was performed.

In the early postoperative course, we observed CSF leaks and meningitis [16]. Cases of recurrence of literature are rare. In this patient we did not get any signs of CSF leak and no signs of meningitis were found.

8. Conclusion

Long segment involvement of spinal intramedullary dermoid cyst in adult without history of trauma makes this case different. Fat suppression imaging must be done in patients with intramedullary hyperintensity in both T1 and T2 sequences of MRI spine. Early diagnosis and appropriate management will be helpful in reducing morbidity.

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Ethical approval

Ethical approval was not required in the treatment of the patient in this report.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Hina Anwer contributes to the study concept or design, data

collection and writing the paper.

Faiq Sheikh contributes to the study concept or design, writing, analysis and interpretation, oversight and leadership responsibility for the research activity planning and execution, including mentorship external to the core team.

Muhammad Anwar Chaudary was the neurosurgical consultant who operated this case.

Registration of research studies

Nil.

Guarantor

Faiq Sheikh.

Declaration of competing interest

There was no conflict of interest.

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