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Case Report

Spinal intradural extramedullary mature cystic teratoma in young adult: a rare tumor with review of literature

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ABSTRACT

Spinal intradural extramedullary teratoma is a rare condition that develops more commonly in children than in adults. We report a rare case of adult onset intradural extramedullary teratoma in the thoracolumbar spinal cord with no evidence of spinal dysraphism and without the history of prior spinal surgery. The patient was a 20 year young adult male whose chief complaint was back pain and urinary incontinence. X-ray images of the thoraco-lumbar spine showed no abnormalities. Magnetic resonance imagings of spine revealed a well marginated cystic intradural extramedullary mass at the D11 to D12 vertebral levels. The patient was operated and the mass was totally excised. It was histopathologically diagnosed as a mature cystic teratoma. The patient's symptom of back pain and urinary incontinence was improved following the surgery.

Keywords: Mature cystic teratoma, Intradural- extramedullary, Young adult

INTRODUCTION

Teratomas are tumors that typically, but not always contain all three germ cell layers ectoderm, mesoderm, and endoderm. Spinal teratoma is an uncommon diagnosis. It constitute 0.2-0.5% of all intraspinal tumors. These tumors usually develop in the sacrococcygeal region in infancy period.[1,2] It occur more commonly in children than in adults and may be associated with spinal dysraphism. In adults, these lesions can occur throughout the spinal canal, but there is a thoracic and lumbar predominance among reported cases with the conus medullaris region being the most frequent. We describe a rare case of Thoraco-lumber spinal intradural extramedullary mature cystic teratoma in a 20 year old male.[3]

CASE REPORT

A 20 year old male was referred to M.D.M hospital Jodhpur with a 1 year history of back pain, numbness in bilateral lower limb, urinary incontinence and constipation.

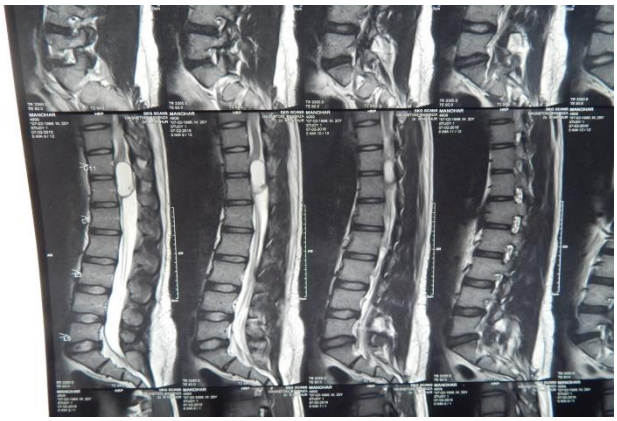

Figure 1: Dorso-lumber spine MRI scan.

On physical examination, no gross motor deficit but numbness in lower limb. Bilateral lower- extremity

hyperreflexia was found. He had no cutaneous abnormalities and there was no evidence of spinal dysraphism, trauma or previous surgery. A plain x-ray film of spine showed no abnormalities. Thoraco-lumber spine magnetic resonance imaging revealed a well marginated cystic intradural extramedullary 14.3x16.3x38.6 mm mass at the D11 to D12 vertebral levels. Conus medullaris shifted upward up to D11 level. The mass was appearing hypointense on T1W and hyperintense on T2W sequences.

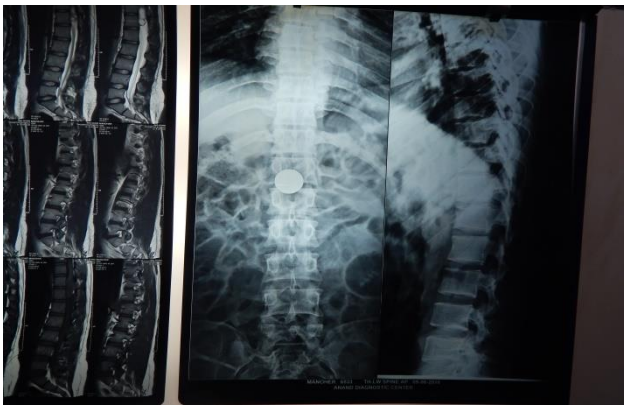

Figure 2: Dorso-lumber spine xray A/P and Lateral view.

There was an internal T1W hyperintense signal focus along posterior wall and post contrast study shows relatively poor enhancement in lesion. The patient underwent surgery, the procedure involved total laminectomy at the D11to L2 vertebral levels. Duramater was opened to expose the tumour.

An oval yellowish uniloculated cystic mass was found attached with cauda equine roots, loosly adherent to conus medullaris. Gross total resection was done. Duramater was reconstructed.

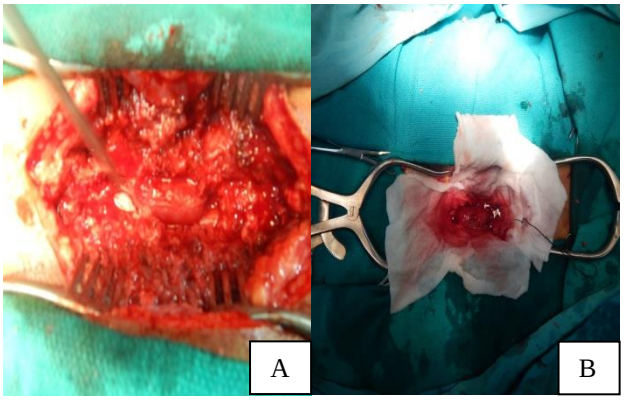

Figure 3: Intraoperative image of spinal tumor.

Histopathology of mass revealed mature cystic teratoma, no evidence of immature and malignant components. The patient had an uneventful postoperative course and his back pain and lower limb numbness improved after surgery. Urinary incontinence and constipation complaints were improved in 2 weeks after surgery.

DISCUSSION

Spinal tumors can be extradural or intradural. Intradural tumors can further be classified as intramedullary or extramedullary. Among the wide variety of intradural extramedullary tumors in adults, the most common are nerve sheath tumors (neurofibromas and schwannomas) and meningiomas with spinal teratoma being a rare one. Intradural extramedullary tumors are the most common primary neoplasm in the spine, accounting for approximately 55% of primary spinal tumors. The first reported case of spinal teratoma was described in 1863 by Virchow and Gowers and Horsley described another case 25 years later.[1,5] Spinal teratomas account for only 0.2-0.5% of all spinal cord tumors.[2]

They can be intramedullary or extramedullary. In according to Poeze et the 31 cases of 83 cases were intramedullary type, and the most of 52 cases were intradural extramedullary type.[5] Spinal intradural extramedullary teratoma is rare entity in adults. It is more common in children than adults. The association of teratomas and spinal malformations such as spina bifida, partial sacral agenesis, hemivertebrae, myelomeningocele, tethered cord syndrome, and diastematomyelia has been described in the literature.[7]

Besides congenital abnormality, cases have been reported where a history of trauma or surgical interventions (example lumbar puncture) often preceded the clinical presentation in adults.[6] However, present patient did not show any evidence of spinal dysraphism during physical and radiologic examination. Also, he had not previously undergone any spinal surgery or procedure. Although sacrococcygeal teratoma in newborn babies has frequently been reported. In adult cases of spinal teratoma, the thoracolumbar region is most commonly affected, particularly in the area of conus medullaris.[7] The clinical features, including weakness of the leg, sensory changes, and reflex abnormalities, are related to the location of the tumors. In the literature, most of cases have presented with these symptoms. The current classification states that a teratoma is a tumor that typically but not always composed of derivatives from all 3 primitive germ layers.[5] Histologically, these lesions are divided into three categories; mature, immature, and malignant. Mature and immature teratomas may be considered benign, especially if treated early. Mature teratomas contain components that are fully differentiated. Immature lesions typically contain fetal tissue. Malignant teratomas are named for the presence of the malignant germ cell layer component.

In the classification of spinal teratomas, several authors have used numerous terms interchangeably ; these term include"epidermoid," "dermoid," "enterogenous," "teratoid," and "teratomatous" cysts.[8]

There are two dominant theories3 regarding the origin of intraspinal teratomas the dysembryogenic theory and the misplaced germ cell theory. According to the dysembryogenic theory, spinal teratomas arise from the pluripotent cell and that in a locally disturbed developmental environment, these pluripotent cells differentiate chaotically. When such disordered development occurs in a primitive streak or a caudal cell mass, a spinal teratoma forms. Another is the misplaced germ cell theory according to which certain pluripotent primordial germ cells of the neural tube that get misplaced during migration from the yolk sac to the gonad, lead to spinal teratoma formation. In adult intraspinal teratomas, which rarely present with significant dysraphism, the misplaced germ cell theory is likely to be more feasible.[3]

The mature teratoma in our case may support the idea of a tumor actually arising from misplaced pluripotent primordial germ cells. The differential diagnosis of lumbar radiculopathy in this case includes degenerative conditions like disc herniation, inflammatory conditions like multiple sclerosis, transverse myelitis, infective conditions like an abscess, tuberculosis, vascular conditions like cord infarction and neoplasms. Although uncommon, spinal cord neoplasm should be considered the differential diagnosis in patients presenting back or radicular pain associated with neurological deficits. In our case, a young adult patient, clinical history, physical examination, and laboratory results increased our suspicion of degenerative condition or neoplasms, which was resolved with the help of imaging studies.

In the diagnosis of spinal teratomas, the role of plain X-ray is limited to detecting changes in the vertebral bodies, such as the erosion and widening of the interpedicular space due to the presence of a mass in the spinal canal, with or without vertebral abnormalities, including spinal bifida, vertebral body fusion or asymmetry, and diastematomyelia, at the level of the lesion. A CT scan may show variable tumor density or calcification. MRI is regarded as the gold standard diagnostic technique that can reveal the location of teratomas and, consequently, the degree of spinal cord involvement. The finding of mixed high and low intensity signals reflects the cystic and solid compositions of the tumor. Definitive diagnosis is only possible with histopathological examination, representing the three germinal layers (ectoderm, mesoderm, and endoderm). However, the presence of just two layers does not rule out the diagnosis.

Total resection is the treatment of choice .Removal of the tumor should be stopped when the neurological function is at risk due to the tenacious adhesions of the tumor to the adjacent parenchyma. Intimal adhesion of the teratomas to the surrounding neural parenchyma is observed about 50% cases.[7] Recurrence rates depend on the histopathological characteristics of the tumors. Recurrences were found to be more commonly encountered with immature and malignant teratomas. symptomatic recurrence rates of mature teratomas were very low even in subtotal resections. Radiotherapy is an adjuvant treatment modality that should be indicated when the tumors contain malignant components. However, adjuvant radiotherapy is not recommended for benign teratomas, and there is not any role of adjuvant chemotheraphy in the treatment of spinal teratomas.4

CONCLUSION

Intradural extramedullary teratoma is a rare tumor. The diagnosis is based on the intraoperative and the histopathological examination. Total surgical excision is the primary treatment modality. Long term follow-up is required to rule out recurrence of the tumor.

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