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A thoracic intradural intramedullary epidermoid in a 12-years old female without any evidence of spinal dysraphism. A rare case report and review of literature

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ABSTRACT

The spinal epidermoid lesion is an extremely rare benign condition, seen in < 1% of all spinal tumours and are most commonly associated with spinal dysraphism. They are more common in young children and have female preponderance. They can be either congenital or acquired with congenital being more common. They are most commonly located in the thoracic region. They grow slowly and present with back pain and progressive neurological deficit with or without bladder bowel involvement. We present a case of a 12-year-old female child with gradually progressive neurological deficit in the form of spastic paraparesis and decreased sensation with early bladder involvement. Her contrast MRI dorsolumbar spine showed a well-defined intradural intramedullary lesion hypointense on T1 image, hyperintense on T2 image with no contrast enhancement at D10-D11 level. The patient was managed by surgical intervention with D9-D10-D11 laminectomy with total excision of the mass. Postoperatively on follow up patient had gradually improved motor and sensory symptoms with no improvement in bladder symptoms. Her histopathological study was confirmative of an epidermoid cyst.

Keywords
ectodermal cells, spinal epidermoid, spinal dysraphism, intramedullary, intradural

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INTRODUCTION

Spinal epidermoid tumors are rare occurrence found in < 1% of all spinal tumors in adults and 3% in childrens .[1-3] They are benign lesion with slow and indolent growth. They are derived from epidermal elements and can be congenital with entrapment of ectodermal cells during development and acquired by iatrogenic injury after surgery, trauma (penetrating injuries), lumbar puncture, myelography .[4-5] They are most commonly associated with spinal dysraphism. It was first reported by chiari in 1833. [6-7] There most common location is thoracic followed by lumbosacral region. Most of these lesions are found extramedullary and intramedullary location is a rarity .[8-9]

CASE REPORT

A 12 year old female child presented with complaints of gradual progressive ascending weakness and numbness of both lower limbs for 6 months with difficulty in walking and bladder involvement in the form of intermittent retention of urine for last 3 months .There was no history of back pain ,fever, trauma , any previous spinal procedure , cutaneous manifestation of spinal dysraphism .Her neurological examination revealed spastic paraparesis with increased tone (grade 3 modified ashworth scale)in both lower limbs , power 3/5 in both lower limbs . Reflexes were +3 in both knee, ankle. DTR were brisk and planter was bilateral extensor. Sensory examination showed decreased pain, touch and temperature sensation of around 60% below L2 spinal segments with decreased perianal sensation. Her systemic, general, local, spine and upper limbs were normal in examination.

MRI dorsolumbar spine revealed a well-defined, homogenous, oval, cystic intramedullary lesion seen at D10-D11 level. It was hypointense on T1 image (figure 1a), hyperintense on T2 image (figure 1b,1c,) with no contrast enhancement (figure 2a,2b). D9-D10-D11 laminectomy was performed with bulging dura seen. After opening of dura, a 1.5 x .8 x .5 cm intramedullary lesion, bulging out from the cord, and attached to cord on right lateral aspect was seen, it was well encapsulated, pearly white in color, smooth, soft to firm, avascular in nature. The cord was thinned out and shifted to left side. Total excision of mass was done with watertight closure of dura was performed.

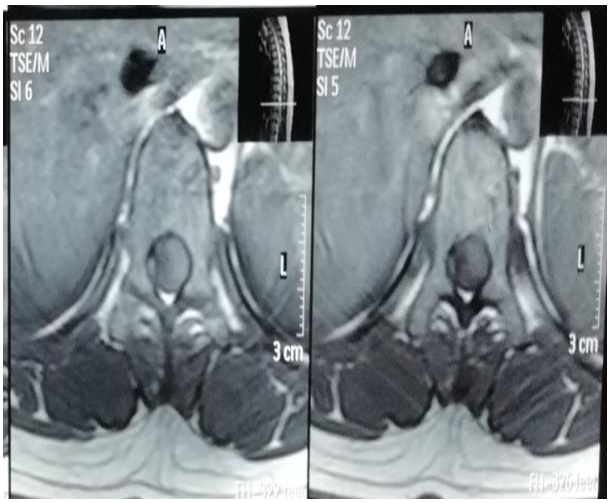
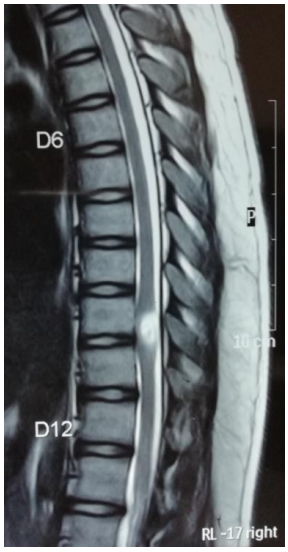


Figure 1a. T1 weighted MRI axial view showing hypointense lesion.



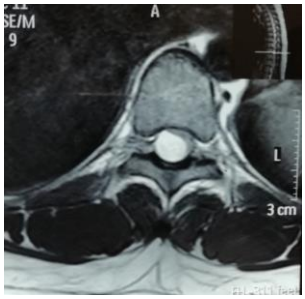
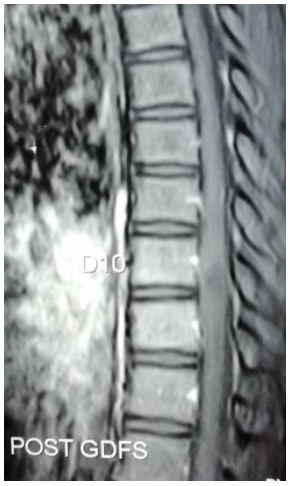


Figure 1. T2 weighted MRI b) sagittal view c) axial view showing single, cystic, hyperintense intradural lesion at D10 –D11 level, with cord shifted anteriorly and towards left side.



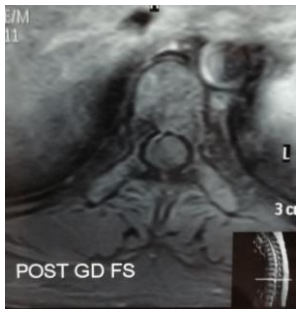


Figure 2. Post gadolinium contrast image a) sagittal, b) axial view with no contrast enhancement.

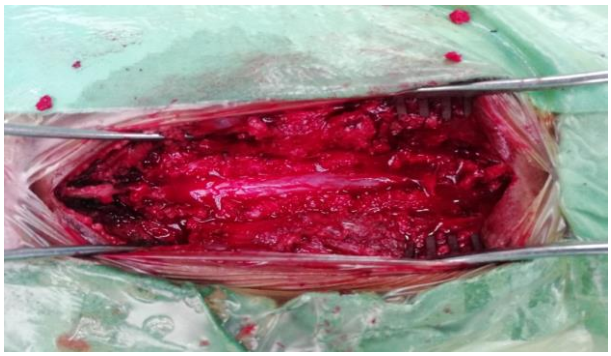


Image 1. Intraoperative image with dural bulge (arrow head).

Post-operative period was uneventful with mild improvement in power and tone. Post op MRI was done and revealed complete excision of mass (figure 3a, 3b, 3c). Pt was discharged and followed up at 3 and 6 months. Her power improved gradually to 4+/5 in both lower limbs and tone was decreased, sensory symptoms also improved but there was no

improvement in bladder symptom. Her histopathological study showed a fibrous capsule with stratified squamous epithelium with degenerated epithelial tissue (figure 4) confirming it to be epidermoid cyst.

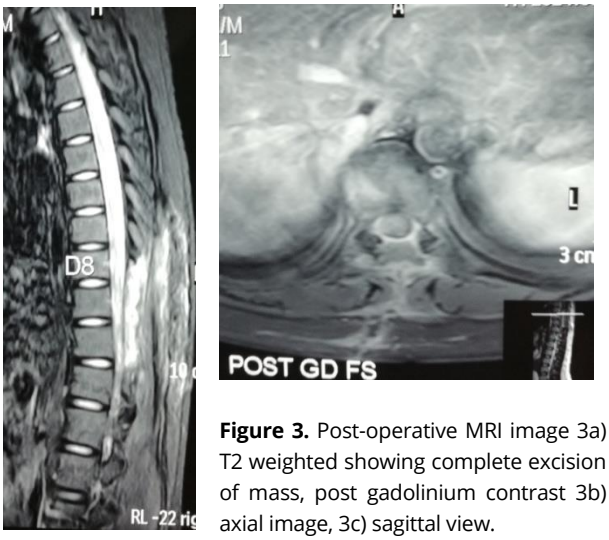


Figure 3. Post-operative MRI image 3a) T2 weighted showing complete excision of mass, post gadolinium contrast 3b) axial image, 3c) sagittal view.

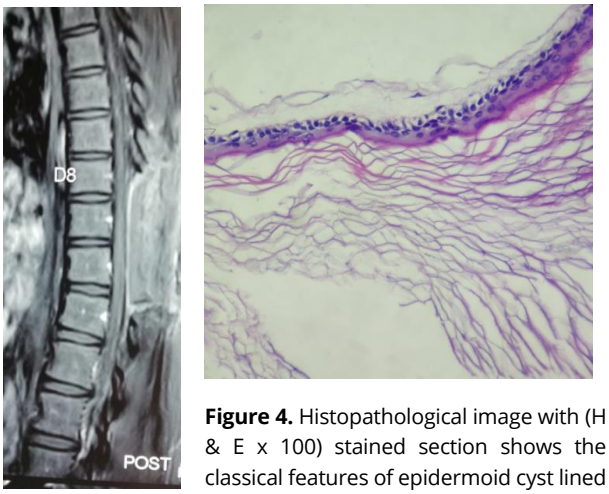


Figure 4. Histopathological image with (H & E x 100) stained section shows the classical features of epidermoid cyst lined with keratinizing stratified squamous epithelium with anucleate squamous debris.

Discussion

Spinal epidermoids are uncommon lesions and very rare without any evidence of spinal dysraphism. Its first description was given by cruveilhier in 1835 , who described them as tumors perlees (pearly tumors) .[10] Its first surgical removal was done by Gross in 1934 .[11] An incidence of .7% was reported by Guidetti and Gagliardi .[12] In 1956 Choremis *et al* described 6 case in childrens receiving subarachnoid injections of streptomycin for treatment of TBM . Roux *et al* in 1992 presented 47 cases of intramedullary epidermoid .[13] Manno et al reported a total of 90 cases, of which 39 were acquired and 51 congenital .[14] These are most commonly seen in thoracic spine in D4-D6 and D11-D12 region followed by lumbosacral region.

They are benign lesion which arise when epidermal cells are trapped into the thecal sac. This can occur congenitally or acquired. Congenital causes are spectrum of spinal dysraphism including spina bifida , myelomeningocele , split cord malformation- diastematomyelia ,dermal sinus ,syringomyelia , hemivertebrae .[15 -17-18] Love and kernohan in 1936 described them as congenital epithelial tumors .[16] They are believed to arise from displaced ectodermal inclusions in early development resulting from defective closure of neural tube between 3rd to 5th week of fetal life .[17] Dias and Walker corroborated a defect at the gastrulation stage .[18] On the other hand acquired lesions develop after iatrogenic displacement of epithelial cells after trauma , lumbar puncture ,previous surgery .[14,19] Gibson and Norris found epidermal tissue in needle bevels after skin puncture and needles with well fitted stylet did not carry any fragments with production of intramuscular dermoid in rats after injection of these skin fragments .[20] Oblu experimented development of intraspinal epidermoid cyst by introduction of dermoepidermal fragment into subrachnoid space of dogs by lumbar puncture .[21] A study by Van Gilder and Schwartz in rats showed , 89% of young rats develop intraspinal epidermoid and they failed to develop in adult rats .[22] Repeated punctures are thought to increase the risk of developing intraspinal epidermoid cyst . Since 1970 incidence has decreased because of use of styletted needles lumbar puncture.

The signs and symptoms are dependent on location of tumor and size of lesion. Slow and indolent growth causes the usual delay in diagnosis. They can range from back pain, painful radiculopathy to UMN signs below the level of lesion including weakness, spasticity, increased tone from involvement of corticospinal tract to sensory deficit, numbness and bladder -bowel dysfunction. Rarely they can also present with chemical meningitis .[23] Malignant transformation has not been documented so far in intraspinal epidermoid.

Radiologically, the X-ray of these patients are usually normal but may show the evidence of spinal

dysraphism, and scalloping of the vertebral bodies, and scoliosis. On CT spine they show widening of spinal canal, scalloping of the vertebral bodies, thinning of lamina. MRI of spine usually shows a well-defined homogenous lesion that is hypointense on T1 image , hyperintense on T2 image , shows intense diffusion restriction on DWI image and no significant enhancement on T1 gadolinium image [6,24,25,26,27,28,29]. Occasionally their margin may look "shaggy" due to leak of content causing chronic inflammatory response and gliosis along the margin .

Diagnosis is primarily done by proper history and clinical examination and established with preoperative radiological investigation and confirmed by postoperative histopathological findings. The differential diagnosis of these tumors includes dermoid, neurofibroma, meningioma, lipoma.

On HPE examination stratified squamous epithelium and an outer layer of collagenous tissue. With progressive desquamation, keratin breakage the inner part of tumor is filled with soft white material, rich in cholesterol .[13,28,30]

Treatment*:* In symptomatic patients. Total excision of mass without any neural damage is the primary goal. In asymptomatic cases found incidentally, it can be managed conservatively. Mostly their capsule is attached to the cord element, so some authors (including Rand and Rand [31]) believe in subtotal resection of tumor with preservation of neural structures. Subtotal resection frequently results in early relapse of symptoms and leak of contents causing more distressing meningitis. Total microsurgical excision with intact capsule is the best possible treatment option available [13,32,33], but may lead to some neurological injury. This can be prevented by using intraoperatively neuro-physiological monitoring guided by EMG and somatosensory evoked potential .[34] These lesions have high recurrence rate because of its dense adherence to neural tissue and the risk of recurrence is approximately equal to patients age plus 9 months .[35,36] Surgery remains the treatment of choice for recurrent lesions also. Radiotherapy was given to 1 patient with repeated relapses with good result .[37] Radiotherapy can be offered to multiple recurrence .[37] So, it can be considered an option for pts not fit to undergo surgery. Despite all this most pt treated surgically enjoy a normal life with good neurological function.

Conclusion

Spinal epidermoid tumors are extremely rare slow growing lesions with non-specific presentation. Most commonly these are congenital lesions seen in children. Mostly they present with gradually progressive neurological deficit. Diagnosis is predicted MRI with DWI features and are confirmed by histopathological evidences. Early diagnosis and prompt treatment limit the neurological deterioration. Surgical excision is the treatment of choice. The preoperative neurological status is the prime determinant of functional outcome. Maximal safe resection results in cessation of clinical progression and remission of symptoms. Subtotal resection results in tumor recurrence and surgery is the treatment option. Radiotherapy can be offered to multiple recurrence.

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