Abstract: Spinal epidermoid cysts, whether congenital or iatrogenic, are relatively uncommon in the spinal cord. When they occur, the typical location is in the subdural, extramedullary space of the lumbo-sacral region. We describe an unusual presentation in a 6-year-old female child with extra-medullary intradural epidermoid tumour at D9 level. The child developed sudden onset of inability in walking and weakness of both lower limbs and urinary incontinence. There was a dramatic reversal of symptoms after surgery. Histopathology revealed an epidermoid cyst of the spine. On the follow-up visit at 2 months, the child was asymptomatic.

Keyword: Spinal epidermoid tumour,

CASE REPORT:
A 6 year female child presented with complaints of pain over upper back region which was aggravated by sneezing and coughing, inability to walk without support, and urinary incontinence. On examination tenderness over D8 to D10 region, weakness of both lower limb (power both hip and knee 4, ankle 5), sensation intact. Urinary incontinence present. Base line blood investigations and X-ray of Dorso-lumbar spine within normal limit. MRI of the spine shows intradural extra-medullary mass at the level of D9 vertebra hypointense on T1 and hyperintense T2 images also no enhancement with contrast.

SURIGAL PROCEDURE:
We done a D7,D8,D9, laminectomy, and dura opened in the midline, mass was excised. Some of the capsule was adherent to the cord so left alone to avoid cord damage. Post-operative period was uneventful. After 2 months patient neurological status dramatically improved to the normal.
Discussion:

Epidermoid cysts are mainly congenital as they take origin from anomalous inclusion of the ectoderm tissue during the closure of the neural tube in early fetal life and possibly may be associated with defective closure of the dural tube. This may have manifestations of other forms of dysraphism, such as syringomyelia, dorsal dermal sinus, spina bifida and hemivertebrae.[6,7] Iatrogenic penetration of the skin fragments after single or multiple spinal lumbar punctures or after meningocele repair may result in an acquired form of epidermoid cyst. This has been even reported after the spine procedure. Epidermoid tumors rarely occur in the central nervous system, and are even rarer in the spinal canal. Even the largest series of neural tumor found an incidence of spinal epidermoid cyst at 0.7%. Intramedullary localization is extremely rare. In 1989, Roux et al. found 47 cases of spinal intramedullary epidermoid cysts in the literature and in their clinic. There are other entities that may pose a challenge to preoperative diagnosis, like dermoid cyst, teratoma, ependymomas, astrocytomas and hemangiomas, owing to their intramedullary localization. However, typical signal intensity, absence of peritumoural edema, sharp boundaries and minimal peripheral enhancement with gadolinium confine the diagnosis to an epidermoid cyst.[4,10] Our case showed similar features to conclude a diagnosis. Histologically, epidermoid cysts are lined by the stratified squamous epithelium supported by an outer layer of collagenous tissue. Desquamation of keratin from the epithelial lining toward the interior of the cyst produces a soft white material. Absence of skin adnexa will rule out the diagnosis of dermoid cyst.[8] The treatment of epidermoid cyst is essentially surgical. Literature shows radiotherapy as a modality in only one case.[11] Emptying of the cyst material and removal of the capsule is the treatment of choice.[5,8,12] In our case, the capsule was not so adherent to the neural tissue and it was removed near completely without damaging the neural tissue. Attempts to completely dissect out remnants of the capsule may result in avoidable sequelae of neurological deficit.

Conclusion:

Epidermoid cyst of the spinal cord very rare entity, and in spite of its vast involvement of the cord, benign nature of the epidermoid cyst offers an opportunity for a better neurological outcome if detected and operated early.

References