

Isolated thoracic intramedullary epidermoid cyst - a rare entity

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Abstract: Intramedullary epidermoid cysts of the spinal cord are rare tumours, especially those not associated with spinal dysraphism. Around 60 cases have been reported in the literature. Of these, only 10 cases have had magnetic resonance imaging (MRI) studies. Here, we report a case of isolated intramedullary epidermoid tumour at D2-D3 level. The etiology, pathology, clinical features, MRI characteristics and surgical treatment of such rare intramedullary tumours are discussed.

Key words: Intramedullary, Epidermoid cyst, thoracic

Introduction

Spinal epidermoid tumours are rare and comprises < 1% of all spinal tumours. These are mostly located in intradural and extramedullary position and commonly associated with congenital malformations like syringomyelia, dermal sinus, spina bifida or may be iatrogenic following procedures such as surgery for bullet wounds, myelography or lumbar puncture. (1) Isolated intramedullary epidermoid tumours without any congenital malformations are even rarer. Thoracic region is the commonest site of intramedullary cysts followed by lumbar and rarely cervical region. We encountered a patient, where epidermoid

was not only intramedullary, it was not associated with any congenital stigmata or malformation. Only 60 cases of intramedullary epidermoid have been reported till now. (2) Rarity of such lesion prompted us to report this case.

Case report

A 27 year old man presented with gradually progressive and descending weakness of bilateral lower limbs for the past one year. It was associated with spasticity and paraesthesia in both lower limbs. There was no history of fever, back pain, bowel/bladder involvement, trauma, any surgical procedure involving spine or any congenital malformation of spine.

Clinical examination revealed spastic paraparesis with power of grade 4/5 MRC in both lower limbs, with maximum weakness at right ankle joint (2/5). Reflexes were 3+ bilaterally at knee and ankle and plantars were bilateral extensors. Sensory examination revealed decrease in pain and touch sensation of up to 20-30% from D4 to D9 spinal segments along with decreased proprioception below D4 with intact perianal sensations. Examination of upper limbs revealed no abnormality.

Magnetic resonance imaging (MRI) dorsal spine revealed a well-defined intramedullary cystic lesion at D2-D3 level. The lesion was hypointense on T1 and hyperintense on T2 weighted image with no enhancement of lesion after administration of gadolinium (Figure 1, Figure 2). Patient underwent D2-D3 laminectomy and cord was found to be widened at the level of lesion. A midline dorsal myelotomy was performed. A 2cmx1.5cm sized intramedullary pearly white lesion was found. Lesion was encapsulated, soft, nonvascular and gross total excision was performed using microsurgical technique and electrophysiological monitoring. The part of capsule which was attached densely to the cord was left behind. Immediate post-operative period was uneventful without any deterioration of neurological symptoms.

At 6 months of follow up, spasticity reduced in both lower limbs and power too improved to 3/5 MRC at right ankle joint. There was no improvement in sensory loss. On histopathological examination, the lesion had a thin fibrous capsule encircled by gliotic tissue. Cyst was lined by compressed stratified

squamous epithelium and contained degenerated squamous tissue, thereby confirming the diagnosis of epidermoid cyst (Figure 3).



Figure 1 - Saggital T1 and T2 weighted image showing lesion at D2-D3 level



Figure 3 - Axial T1 and T2 MRI showing intramedullary lesion

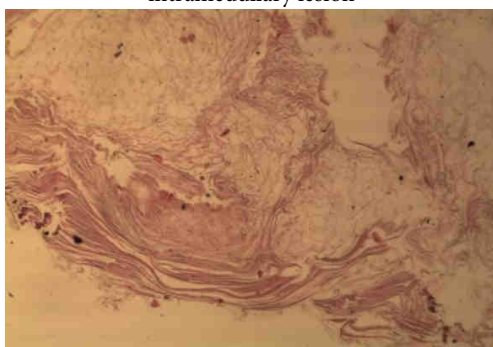


Figure 3 - Microphotograph showing Epidermoid cyst filled with lamellated keratin. H&E 40x

Discussion

Epidermoid tumour was first described by Cruveilhier in 1835, who called them "tumors perlees" (pearly) tumour. (3) The first description of an intramedullary epidermoid cyst belongs to Hans Chiari in 1883. Incidence of epidermoid cysts among intracranial tumours in most large series is estimated to be 0.2%-1%. It is approximately 0.7% of all spinal epidermoid cysts. (4) Symptoms of epidermoid cyst vary with the level of involvement but same as other compressive lesions in spinal column. Presentation is usually in the form of progressive neurological deterioration like paraparesis sensory loss urological manifestation and back pain. Epidermoids grow very slowly in path of the least resistance and may lead to delay in diagnosis. Duration of symptoms before diagnosis may extend for years.

It is generally believed that congenital epidermoid cysts originate from displaced ectodermal inclusion arising in early fetal life and possibly may be associated with defective closure of the dural tube. (5) Acquired epidermoid cysts are thought to result from iatrogenic proliferation of skin fragments especially after lumbar puncture or meningocele repair. (6, 7) Epidermoid cysts are generally characterized on MRI by variable signal intensity, they are usually slightly hyperintense as opposed to cerebrospinal fluid, and their lobulated aspect helps differentiating these lesions from other cysts. This disparity in signal intensity most likely reflects variable lipid and protein composition in these lesions. Other features

include well defined palne of cleavage, calcification and absence of surrounding edema. Additionally, it has been noticed that the margins of these lesions are shaggy, possibly because of chronic inflammatory response to squamous tissue leak through the capsule and variable gliosis along the margin, extending into the cord. Diagnosis of epidermoid cyst is usually based on inspection due to typical aspect of their content. Histologically, epidermoid cyst is lined by stratified squamous epithelium supported by another layer of collagenous tissue, and progressive desquamation of keratin from epithelial lining toward the interior of the cyst produces a soft whitish material. To the best of our knowledge, there have been only seven cases of intramedullary epidermoid with MRI features reported till date in the literature. (8, 9, 10, 11, 12, 13, 14).

Management of intramedullary epidermoid cyst is surgical. Decompression of the cyst material is easily performed, but tumour capsule is usually adherent to the cord and complete removal may cause permanent neurological deficit, so it should be left behind. The risk of recurrence exists, however, in most cases even partial removal of the intramedullary epidermoid resulted in total remission of symptoms. If recurrence occurs again, surgery is the treatment of choice.

Prognosis of these tumours is good because of their histologically benign nature and surgical intervention should be done as soon they are diagnosed.

Conclusion

Epidermoid tumours are infrequent, without any specific clinical features. Due to atypical features and delay in the onset of symptoms, high index of suspicion should be kept in mind and detailed neurological examination along with MRI of the spinal cord is warranted to diagnose these cases at the earliest. Generous reporting of such cases is to be done to better understand this rare entity.

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