

Intramedullary spinal epidermoid cyst: The Value of MRI in early diagnosis and management

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Abstract

We report a case of a very rare intraspinal tumor. It is intramedullary epidermoid cyst in a child 3 years-old, which is also rarely an age of presentation. This tumor was diagnosed through spine MRI examination. The child presented with spastic paraparesis of three months duration. Prior to the MRI study, the child was submitted to spinal CT examination which failed to fully detect the lesion, leading to delay of correct management. MRI should be carried out directly as the investigation of first choice whenever spinal lesion is suspected. In this case report, the MR imaging characteristics of such a rare benign tumor are discussed with a review of the relevant literature. Key words: Epidural analgesia, Ischemic heart disease, Haemodynamics.

Introduction:

Most of spinal canal epidermoid cysts are intradural extramedullary in location. Epidermoid cyst within the spinal cord (intramedullary epidermoid cyst) is a very rare condition. Less than 55 cases have been reported in the literature, of these, only 12 cases have been subjected to magnetic resonance imaging (MRI) studies{1}. The first objective of this report is to emphasize the role of MRI in the early diagnosis of intramedullary tumors, analyzing the MRI features of this rare lesion. The second objective is to create awareness of the possibility of early presentation of such benign tumor.

Case history:

A 3 year- old male child was referred to our radiology department for spine MRI because of progressive lower limbs weakness and arching of the back for 3 months. There was no history of trauma, fever or convulsion. The child was vaccinated against polio. The physical examination revealed bilateral decrease in motor power of lower limbs (grade 2-3).

The ankle and knee reflexes were slightly exaggerated. There was bilateral decreased proprioception below T12. Upper limbs were normal. Spine CT scan which had been done before (Figure 1) showed ill defined hypodense expansion of the spinal canal at the level of lower thoracic region, but no well demarcated lesion could be outlined. During this period antibiotics and anti-inflammatory drugs were prescribed but without benefit.

Under general anaesthesia the MRI for the whole spine was performed in sagittal and axial planes using closed type MRI machine of 1.5 Tesla. The used sequences were T1 and T2, as well as, T1 after 2.5ml of Gd-DTPA contrast administration. STIR sequence was not used because no hyperintensity was found on the pre-contrast T1 WI. In the axial cuts, 4mm. slices thickness were applied.

The MRI revealed an intramedullary well defined, smoothly outlined space occupying lesion that expanded the conus medullaris. The vertical diameter of the lesion was about 12mm. The lesion was hypointense on T1 WI and homogenous hyperintense on T2 WI (Figure2a&2b).

There was no contrast enhancement at all (Figure 3). The surrounded area of the cord didn't show edema or satellite focal lesions. No hydromyelia noticed. The MRI didn't reveal occult spinal dysraphism. The case was reported as intramedullary cystic lesion of benign character with reference to the possibility of epidermoid cyst.

The case was operated and near total excision of the tumor, including the capsule was performed and sent for histopathological evaluation. The histopathological report stated the wall of the lesion formed of a thin fibrous capsule encircled by gliotic tissue and the luminal aspect was lined by stratified squamous epithelium, so the diagnosis of an epidermoid cyst was confirmed. The patient is recovering well and his motor power is improving.

Discussion:

The spinal canal tumors are traditionally classified as follows:

Extradural: including tumors arising from the vertebrae.

Intradural extramedullary: Meningiomas and neurofibromas are the main tumors encountered of this group. Epidermoid cyst and arachnoid cysts are less commonly found.

Intramedullary: The commonest of these are astrocytomas and ependymomas.

Intramedullary epidermoid cysts represents about 0.6-1.1% of all intraspinal tumors {2}.

The cause of these benign intramedullary tumors could be either congenital or acquired. The congenital variety is more common and may occur as a result of displaced ectodermal tissue in association with closure of neural tube, or rarely due to intraembryonal inclusion of this ectodermal tissue in early fetal life {3} The acquired epidermoid cysts could be found years after single or multiple lumbar spinal punctures, and are thought to result from iatrogenic penetration of skin fragments {4,5}.

The patient with this benign tumor usually presents with neurological deficits due to compressive myelopathy. The sites affected depend on the segment of the cord involved. Sphincter troubles may occur later {2}.

The radiological literature on spinal cord epidermoids is scarce. Chiari reported the first case in 1833 {6,7}. The majority of reported cases occurring in young adults and were associated with spinal congenital anomalies as meningocele or spina bifida. The cause of this association is displacement of ectodermal tissue due to defect of closure of neural tube. Similar

mechanism can explain the pathology of acquired cysts that may appear if the child exposed to lumbar puncture in neonatal period as iatrogenic penetration of ectodermal tissue may accidentally happen {5}. In such conditions, the cysts grow slowly and become symptomatic later in life, usually after ten-year old. Our presented case was not associated with spinal anomalies and there was no past history of any lumbar punctures. The cause in such case is mostly intraembryonal inclusion of epidermoid tissue in early fetal life which may explain the early presentation of the disease in our presented case.

Intramedullary epidermoid cyst is common in lumbosacral and thoracic regions while only three cases have been reported with cervical cord involvement {8}.

Most of reported cases that evaluated MRI features of spinal cord epidermoids are equal with our case in possessing well circumscribed margins and in absence of perilesional edema. Regarding the MRI signals, the tumors in reported cases showed hyperintensity in T2 WI and hypointensity in T1 WI, except for one case reported by Lt Col MN Swamy {9}, in which the tumor appeared iso-intense in T1 WI. Our case showed characteristic MRI features that support, to a major extent, the diagnosis of an intramedullary epidermoid cyst. Firstly, the high sensitivity of MRI for medullary tissue detection allowed good visualization of the very thin compressed cord tissue surrounding the cystic tumor, hence confirming its intramedullary location (Figure 2a&2b). Secondly, the homogeneity in both T1 WI and T2 WI sequences indicates the absence of haemorrhage, necrosis or calcification within this lesion and confirms its pure cystic nature. Thirdly, the absence of wall enhancement of the lesion is an important criterion on which cystic astrocytoma, ependymoma or abscess can be excluded, however, few reported cases of inflamed epidermoid cysts showed mild peripheral contrast enhancement {10} Considerable confusion still exists in differentiating between dermoid and epidermoid cysts. Dermoids often contain skin appendages which if present will cause heterogeneity in all sequences. In addition, dermoids may contain considerable amount of fat component that apparently, if exists, will reflect hyperintensity in T1 WI.

MRI is superior to CT scan in detecting intramedullary lesions because of its ability to obtain sagittal and coronal planes. Furthermore, the different sequences used in MRI provide images that perfectly differentiate between cystic and solid lesions within the cord, and between the cord and outer cerebrospinal fluid.

Another important technical feature of MRI that makes it better than CT scan in the diagnosis of intramedullary lesions is the possibility to avoid any bony artifacts that could be produced by spinal vertebrae.

In conclusion, MRI has undoubtedly revolutionized the diagnosis of intramedullary tumors, reducing the interval between onset of symptoms and diagnosis, and it is the investigation of choice for intramedullary epidermoids. These benign tumors should be considered in mind whenever intramedullary cystic lesion is encountered, even in very young age group.

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Figure 1: CT scan at the level of the lesion shows widening of the spinal canal due to hypodense enlargement of the cord without demarcation of the lesion outline (arrow)

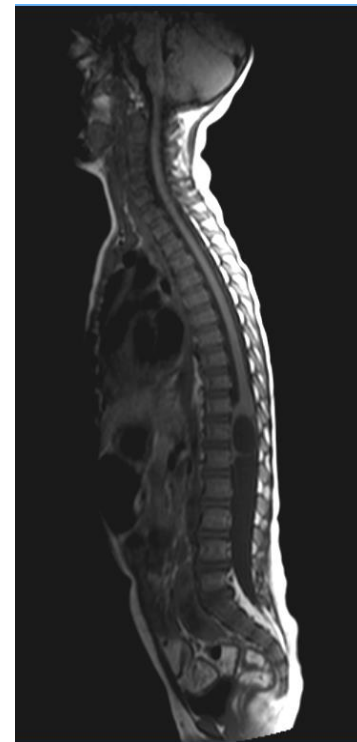


Figure 2(A): sagittal T1 WI of spine MRI shows a hypointense well defined expansile intramedullary lesion at the conus medullaris .



Figure 2(B): The lesion became hyperintense on T2 WI MRI.

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Figure 3: The lesion does not show any contrast enhancement (arrow).