Case Report
Intramedullary Epidermoid Cyst: A rare Intraspinal Tumor

Abdulkhaleq Ayedh Bin Nuhaid* **Wadea Saeed Bin Ghouth***

Abstract

We adduce this case of a very rare intraspinal tumor, an intramedullary epidermoid cyst in a male child of a 3 year-old, which is also a rare age presentation. This tumor was diagnosed through spine magnetic resonance imaging(MRI) examination. The child presented with spastic paraparesis of about three months' duration. The child was set for spine computed tomographic(CT) scan examination (prior to the MRI study) that failed to detect the lesion well, leading to delay of correct management. MRI should be carried out directly as the investigation of first choice whenever spinal lesion questionable in non-informative CT scan study. In this case report, the MR imaging characteristics of such rare benign tumor are debated with review of pertinent literature.

Key words: Epidermoid cyst, intramedullary lesion, magnetic resonance imaging.

Introduction:

Most of spinal canal epidermoid cysts are intradural extramedullary in location [1-9]. Epidermoid cyst within the spinal cord (intramedullary epidermoid cyst) is a very rare condition, less than 1% of all spinal tumors. It was firstly described by Hans von Chiari in 1883[11]. About 50 cases have been reported in the literature, of these, only seven cases have been submitted to magnetic resonance imaging (MRI) studies [12]. In this case report, we aim to create awareness of the role of MRI in the early diagnosis of intramedullary tumors, analyzing the MRI features of this rare lesion, and emphasize the possibility of early age presentation of such benign tumor.

Case Report:

A 3 year- old male child was referred to the MRI unit for spinal MRI study, with a history of progressive lower limbs weakness and arching of the back started since 3 months of age. There was no history of trauma, fever or convulsion. The child was vaccinated against polio virus. Physical examination revealed bilateral decrease of 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 previously (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 that period antibiotics and anti-inflammatory drugs were subscribed but without any benefit.

Under general anaesthesia, MRI for the whole spine was performed in sagittal and axial planes using closed type Phillips MRI machine of 1.5 Tesla. The used sequences were T1 and T2, as well as T1 after 2.5ml of Gadolinium contrast administration. STIR sequence was not used because no hyperintensity was found on the pre-contrast T1 WI Images. 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 12 mm. The lesion was hypointense on T1 WI and homogenous hyperintense on T2 WI (Figure2a&2b). There was no contrast enhancement at post-contrast images (Figure2c). The surrounded area of the cord didn't show edema or satellite focal lesions, and 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.

To opposing surgical procedures for intramedullary epidermoid cyst have been reported previously, one is radical resection of the cyst wall and the other is evacuation of the cyst contents [7]. The internal decompression by evacuation of contents as surgical procedures is chosen when the cyst wall is tightly adherent to the surrounding spinal cord tissue [3].

The case was operated and near total excision of the tumour was chosen [16], including the capsule was performed and sent for histopathological evaluation. The histopathological report declared 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 was recovering well and his motor power was improving.

* Department of specialized surgery, Radiology, College of medicine and health sciences, Hadhramout university-Mukalla-Yemen.Received on 5/2/2018 and Accepted for Publication on 7/5/2018

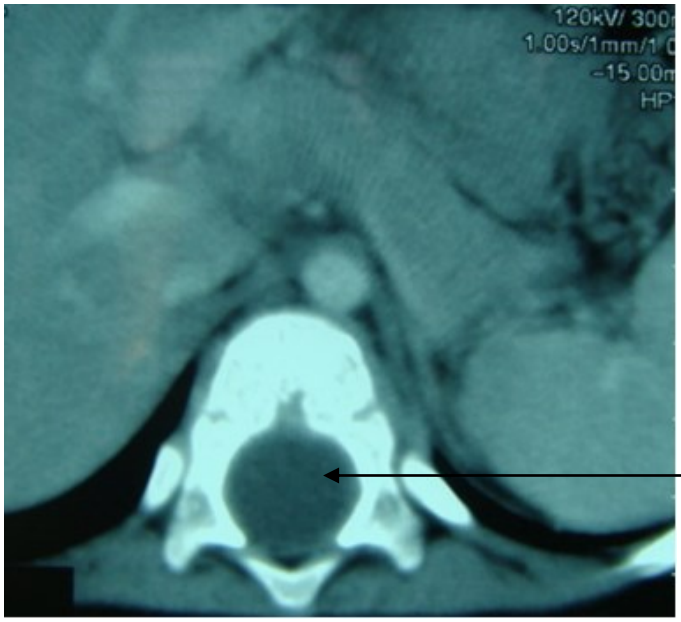
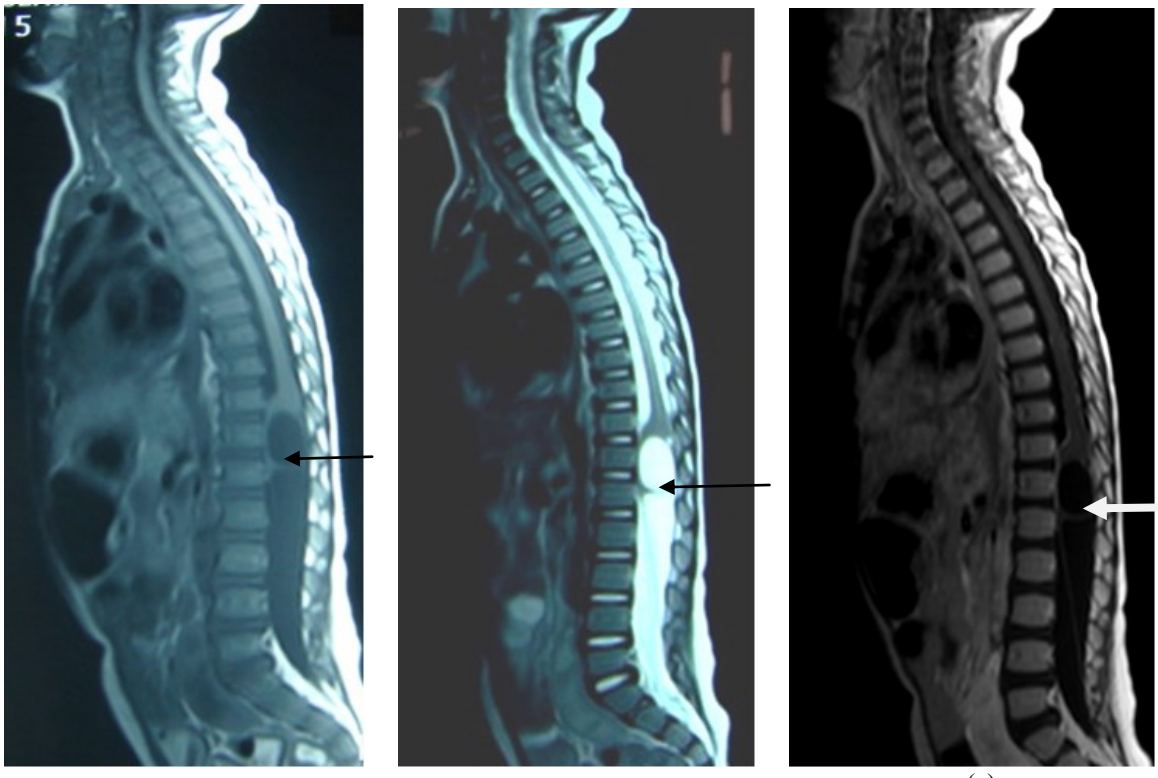


Figure 1: Axial CT scan at the level of the lesion shows widening of the spinal canal and hypodense expansion of the cord, outline of the lesion couldn't demarcated (arrow).



(a) (b) (c)

Figure 2-(a): Sagittal T1 WI of spine MRI; a hypointense well-defined, expansile intramedullary lesion at the conus medullaris (arrow). (b): The lesion turn into hyperintense on T2 WI MRI (arrow). (c): Sagittal T1W1 after Gadolinium administration: The lesion does not show any enhancement (see arrow)

Discussion:

The spinal canal tumours are traditionally classified into: 1-Extradural: including tumours arising from the vertebrae, 2-intradural extramedullary: meningiomas and neurofibromas are the main tumors encountered of this group, while epidermoid cysts and arachnoid cysts are less commonly found, 3-Intramedullary: The commonest of these are astrocytomas and ependymomas [4-5].

Intramedullary epidermoid cysts represent about 0.6-1.1% of all intraspinal tumors [6]. 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 defective closure of neural tube, or rarely duo to intraembryonal inclusion of this ectodermal tissue in early fetal life [14]. 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 [6,11].

The patient with this benign tumor usually presents with neurological deficits duo to compressive myelopathy. The sites affected depend on the segment of the cord involved. Sphincter troubles may occur later [6].

The radiological literature on spinal cord epidermoids is scarce. Chiari reported the first case in 1833 [10,13]. 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 duo 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 [16].

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 lumbo-sacral and thoracic regions while only three cases have been reported with cervical cord involvement [7]. Most of reports that evaluated the MRI features of spinal cord epidermoids are similar to 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 Swamy [15], 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 intramedullary epidermoid cyst. Firstly, the high sensitivity of MRI for medullary tissue detection allowed good visualization of the very thin compressed cord surrounding the cystic tumor, hence confirming its intramedullary location (Figure2a&2b). Secondly, the homogeneity in both T1 WI and T2 WI sequences indicates the absence of hemorrhage, 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 [1].

Considerable confusion still exists in differentiating between dermoid and epidermoid cysts. Dermoids often contains 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.

Finally, MRI has certainly revolutionized the diagnosis of intramedullary tumors, early diagnosis before onset of symptoms, and it is the investigation of choice for intramedullary epidermoids. These benign tumors should be considered in mind whenever intramedullary lesion be facing even in very early child age group.

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الكيس البشراني في النخاع الشوكي: ورم نادر داخل النخاع

عبدالخالق عايض بن نهيد وديع سعيد بن غوث

الملخص

هذه دراسة لحالة نادرة لورم داخل النخاع الشوكي يعرف بالكيس البشراني عند طفل يبلغ من العمر ثلاث سنوات، حيث يعد أيضا نادر الوجود في هذ العمر. تم تشخيص الورم عن طريق الرنين المغناطيسي للعمود الفقري، بينما كان الطفل يعاني من شلل جزئي تشنجي لثلاثة اشهر قبل تشخيصه ، وقد تم الكشف على المريض بجهاز التصوير المقطعي المحوسب لكنه فشل في الوصول إلى تشخيص نهائي للآفة ومن ثمّ تأخر علاج المريض. لذا ينصح بعمل رنين مغناطيسي دون تأخير لكل حالة يتوقع فيها أمراض النخاع الشوكي خاصة عندما لا تفي نتائج التصوير المحوسب. ويناقش في هذا التقرير صفات الرنين المغناطيسي لهذا الورم الحميد النادر مع مراجعه للحالات المسجلة ذات الصلة.

الكلمات المفتاحية: كيس بشراني ، آفة داخل النخاع الشوكي ، التصوير بالرنين المغناطيسي.