Case Report

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Thoracic Spinal Intradural Intramedullary Epidermoid Cyst: Case Report

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Abstract

Background Data: Epidermoid cysts grow slowly so the symptoms would present with slowly progressive course and delay in the diagnosis that may extend up to years.[4] Presence of neurological symptoms and signs is usually detected that might be associated with local mild but persistent back pain**.**

Purpose: To describe a case presentation of spinal epidermoid cyst.

Study design: A case report of intradural thoracic an epidermoid cyst.

Patients and Methods: Our case was a forty years old male patient presenting with left lower limb motor weakness. He had a 5 month history of slowly progressive left lower limb weakness and not associated with sphincter disturbance. Magnetic resonance imaging of the spine demonstrated an intradural lesion of the thoracic part of the spinal cord.

Results: The lesion was excised totally and 14 days after the surgical removal, the motor power of the left lower limb showed marked improvement up to grade 4 on MRC scale.

Conclusion: The diagnosis of intradural epidermoid cyst could be considered in patients with slowly progressive lower limbs weakness and mild persistent back pain. (2016ESJ121)

Keywords: Epidermoid cyst, thoracic spinal cyst, intradural cyst

Introduction

The first description of epidermoid cysts in the literature is attributed to Cruveilhier in 1835. He characterized them as pearly tumors because of their appearance.[5] Epidermoid cysts or tumors are benign lesions that may arise intracranially or in the spine.[16] Intracranial epidermoid cysts account for 0.2% to 1.8% of brain tumors.[13] It is much less amongst the spinal tumors with incidence of 0.6 - 1.1%.[9] Most spinal epidermoid tumors are located in intradural extra medullary position.

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Intramedullary spinal epidermoid cysts are extremely rare.[22] It is encountered commonly in lumbosacral and lower thoracic regions.[14]

About 40% of intraspinal epidermoid cysts are acquired and are considered to be late complication of multiple lumbar punctures or after history of meningomyelocele repair.[7] Presence of other congenital abnormalities like dermal sinus, spina bifida and hemivertebrae augments the congenital origin and are mostly associated with congenital epidermoid cysts.[4]

In this report we would present a case report of thoracic intradural epidermoid cyst

Case report

A Forty years old male patient presented with left lower limb weakness with slowly progressive course along 5 month duration and abnormal sensation all over the lower half of the abdomen till the bilateral feet. The weakness started in the form of loss of dorsiflexion of the left foot then knee flexion and extension and continued to progress and included the whole left lower limb. Condition is associated with bowel disturbance but no bladder disturbance was reported till time of surgery. No history of fever and no previous history of any back surgical intervention or lumbar puncture.

Clinical examination showed no cutaneous abnormalities were present on the back, no palpable masses on the back or localized tenderness and a normal range of motion of both back and lower limbs. Neurological examination showed no abnormalities detected all over the right lower limb but the left lower limb showed motor power grade zero according to the Medical Research Council scale (MRC) involving hip (flexor, extensor and adductor muscles), knee (flexor and extensor muscles) and absent ankle (planter and dorsal flexion). A sensory level has been detected corresponding to Thoracic 6 (about hand breadth below the level of nipples) and was associated with partial loss of superficial and deep sensation on both lower limb. Spasticity and exaggerated knee and ankle reflexes with extensor planter reflex were reported.

The patient was admitted into the Neurosurgery department and neuroimaging studies were done. MRI revealed an intradural lesion on the dorsal aspect of the thoracic spinal cord that extended from T4 to T5 levels. The lesion was showing low signal intensity in T_1 and T_2 weighted MRI with faint contrast enhancement. Computed tomography (CT) showed hyperdense spindle shaped lesion within the spinal canal behind the vertebral body T4. (Figure 1 and 2)

The patient has been scheduled for surgical intervention through a posterior spinal approach for excision of the lesion. Patient has been also formally consented regarding the possible complications and expectation of the recovery.

Under general anesthesia and in prone position, with guidance of intraoperative fluoroscopy the surgery was performed. Electrophysiological monitoring was not available. Through posterior approach, the team did posterior spinal canal decompression laminectomy from T4 to T6. Dura was opened and revealed distended spinal cord content and firm consistency rather than the below and above levels. Through a limited myelotomy and cautious dorsal spinal cord splitting a bright white yellowish capsule was identified. Through a gentle Microscopic dissection, the well-defined capsulated mass was totally excised after meticulous separation from surrounding neuronal elements. Histopathological examination revealed an epidermoid cyst.

Immediate postoperative examination revealed added deficits by occurrence of right

lower limb weakness at levels of hip, knee and ankle. Motor power grade 4 on MRC scale, and the same left lower limb weakness as pre-operative status. Receiving conservative treatment with close continuous physiotherapy, the patient showed gradual motor power improvement on the right lower limb till retaining the full motor power as pre-operative status. Meanwhile the left lower limb motor power showed gradual improvement along the post-operative 4 weeks till reaching grade 4 according to MRC in all limb muscle groups. After removal of the urinary catheter, the patient showing normal bladder functions as pre-operative status.

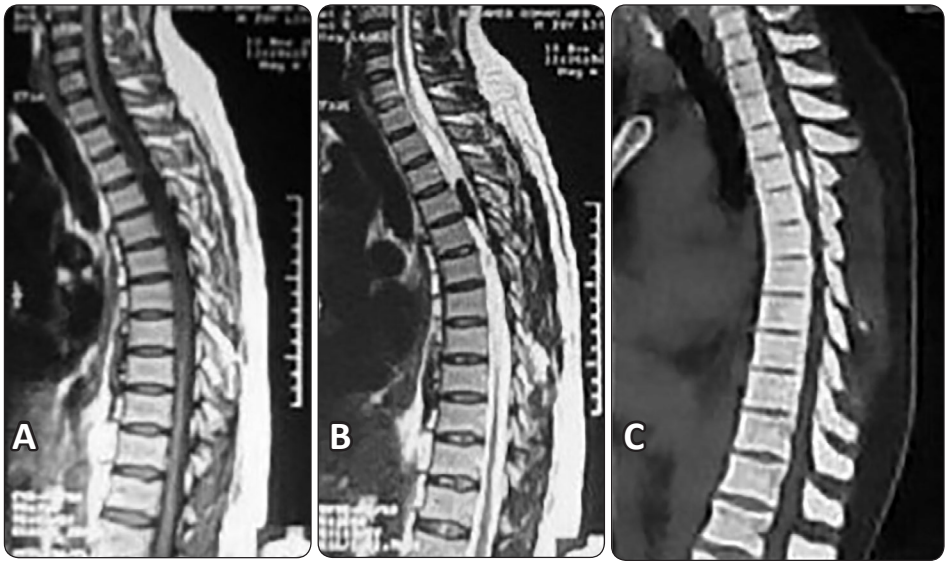

Figure 1: Preoperative imaging, (A) Sagittal T1-weighted MRI showing ill-defined hypo to isointense intradural mass. (B) Sagittal T2-weighted MRI showing hypointense intradural mass. (C) Sagittal thoracic CT spine showing hyperdense intraspinal lesion.

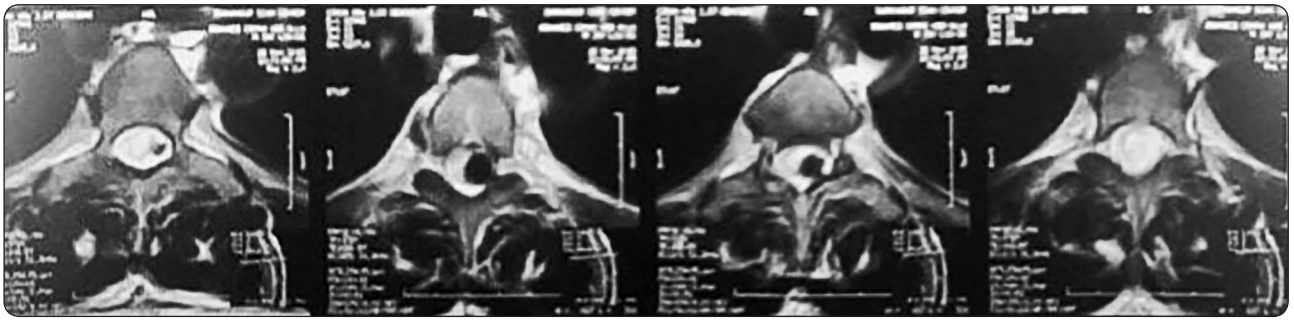

Figure 2: Preoperative axial T2-weighted MRI showing hypo intense rounded intradural mass on the left side.

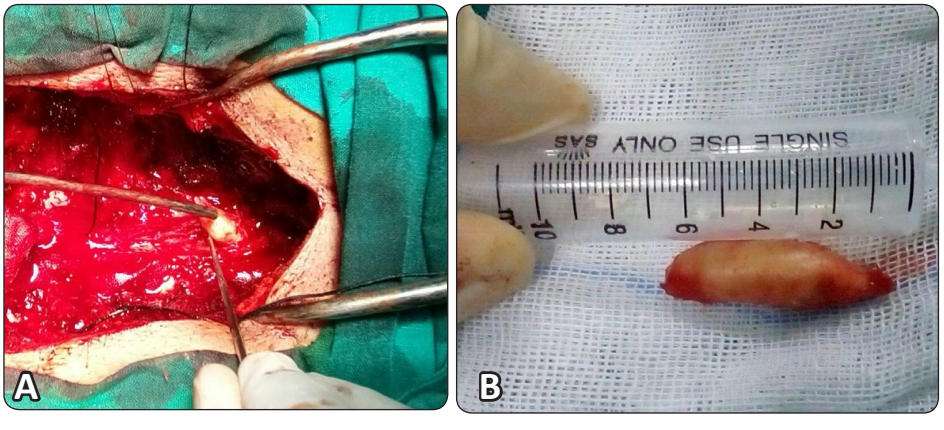

Figure 3: (A) Intra operative imaging showing white yellowish mass while excised from the spinal cord. (B) Showing excised yellowish longitudinal cylindrical well capsulated mass about 3 cm in length.

Discussion

The first intramedullary epidermoid cyst was described by Chiari in 1883, and the first successful surgical removal of an intramedullary epidermoid cyst was reported by Gross in 1934.[19,21,27] In 1992, Roux et al,[21] found 46 cases with spinal intramedullary epidermoid cysts in the literature and reported another case from their clinic in 1992. In our literature review, we found 11 more reports since then.[2,3,7,9,10,14,15,22,24,26,27]

Epidermoid cyst can be either congenital or acquired.[3] In 1936, Love and Kernohan[17] first described epidermoid cysts as congenital epithelial tumors. Traditionally, however, epidermoid cysts had been described as resulting from a primary failure of neurulation, that is these tumors derive from ectopic inclusions of epithelial cells during neural tube closure. This theory, however, could not account for many of the associated anomalies.[11] More recently, Dias and Walker[6] explained their cause as a problem at the gastrulation stage of development, with primary disruption of tissues derived from surface ectoderm. This primary gastrulation dysembryogenesis, leads to a secondary disruption of neural tube closure, during the 3rd to 5th week of gestation. Epidermoid cyst is frequently associated with developmental abnormalities such as spina bifida, dermal sinus, meningomyelocele, diastematomyelia and enterogenous cysts or one or more of the mesodermal malformations, particularly those involving the vertebrae (e.g., hemivertebrae, absent vertebrae, fused vertebrae, butterfly vertebrae, midline bony spurs).[6,11,18] Acquired epidermoid cysts have been found years after single or multiple lumbar spinal punctures or meningomyelocele repairs and are thought to result from iatrogenic penetration of skin fragments.[12] The epidermoid cyst is lined by simple stratified squamous epithelium resting on an outer layer of connective tissue. It is filled with soft and waxy material rich in cholesterol crystals, but contains no skin appendages.[19]

The slow growth of epidermoid cysts often leads to a delay in their diagnosis. Duration of symptoms before diagnostic recognition may extend for many years. The symptoms and signs at presentation of intramedullary epidermoid cysts are not specific, may be present in any myelopathic process and dependent on the level of the lesion. In the thoracic and upper lumbar regions they are usually neurologic, such as progressive paraparesis and motor–sensory complaints. Back pain may be present and may be local or radiating. Sphincter troubles may occur later.[9]

Magnetic resonance imaging is the diagnostic method of choice. In the last decade, MRI has undoubtedly revolutionized the diagnosis of intramedullary tumors, reducing the interval between onset of symptoms and diagnosis. Of all the 58 cases of intramedullary epidermoid cysts reported, there have been only 15 cases with MR imaging reported in the literature earlier.[2-4,7,9,10,14,15,19,20-22,24,26,27] Magnetic resonance imaging of epidermoid cyst is characterized by the absence of perifocal edema and being a well circumscribed lesion. Its lobulated aspect helps differentiating it from arachnoid cyst. It is hypo intense in T1-weighted, hyperintense in T2-weighted images and shows minimal peripheral enhancement with contrast material.[14,19]

The treatment of an intramedullary epidermoid cyst of the spinal cord is surgical. The surgical approach to intramedullary tumors is described by Epstein and Wisoff.[8] Emptying of the cyst material is normally performed without difficulty, but the thin tumor capsule is usually tightly adherent to the surrounding cord neural tissue and its complete removal can cause

neurological deficits. This intimate adherence of the capsule to the cord is confirmed by experimental work on animal models. Van Gilder and Schwartz[25] demonstrated that, in four rats with intramedullary implants with skin, the connective tissue of the cyst invaded the cord parenchyma locally. So when the cyst wall is intimately attached to the spinal cord, it should be left in place.[2,14,24] Intraoperative evoked potentials, motor and sensory, may be of value in maximizing patient outcomes.

In our case, great concern was taken to not compromise the blood supply of the cord because the area T1-T4 is vulnerable area for vascular insult due to decreased interconnections of the vascular supply.

Epidermoid cyst is well known by its high rate of recurrence due to the adherence of its capsule to the surrounding neural tissue.[23] Total removal of the epidermoid cyst is seldom accomplished in the majority of cases.[2] It is thought that if as little as a single cell remains the patient is at risk for recurrence. The risk exists for a duration period equal to the patient's age at diagnosis plus 9 months.[1]

There were 58 cases reported in the literature with intramedullary epidermoid cysts in the spinal cord. The majority of these cases underwent surgical intervention with improvement of the preoperative neurological deficits. We have reviewed the literature and we have not found any description of a recurrent intramedullary spinal cord tumor. In most cases, even partial removal of the intramedullary epidermoid resulted in total remission of symptoms for a long period, enabling patients to enjoy good function. If recurrence does occur, another surgery may relieve the symptoms again.[3]

Conclusion

The diagnosis of intradural epidermoid cyst could be considered in patients with slowly progressive lower limbs weakness and mild persistent back pain. The case reported is very unusual because of its type, intramedullary location, and not associated with other congenital anomalies. The surgeon role is to remove the cyst content as well as the safe part of the capsule.

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الملخص العربى

حويصلة بشرية بداخل الأم الجافية للحبل الشوكى بمنطقة الفقرات الصدرية: تقرير حالة

البيانات الخلفية: هناك العديد من الأجسام الغريبة التى تنمو بداخل الأم الجافية للحبل الشوكى, و منها ما هو حميد و ما هو خبيث. وجود الحويصلة البشرية أمر غير معتاد اطلاقا خصوصا فى أناس لم يخضعوا لعمليات بالظهر سابقا. و تمتاز الأعراض المصاحبة للحويصلات البشرية أنها تحدث بشكل بسيط ثم تزداد و تتطور على مدار يصل الى سنوات.

الغرض: عرض حالة نادرة الحدوث لوجود حويصلة بشرية داخل الأم الجافية للحبل الشوكى

تصميم الدراسة: تقرير حالة

المرضى والطرق: هذه الورقة البحثية هى عرض لحالة مريض عمره أربعون عاما أصيب بضعف بالقوة الحركية للطرفين السفليين بشكل تدريجى متزايد لمدة خمسة أشهر سبقه وجود آلم مزمن بسيط بوسط العمود الفقرى لمدة حوالى عام و كان يستجيب للعلاج الدوائى كالمسكنات. تم عمل فحص بالرنين المغناطيسى و الآشعة المقطعية أوضحت وجود كتلة غريبة داخل نسيج الحبل الشوكى فى منطقة الفقرات الصدرية أمام الفقرة الرابعة و الخامسة

النتائج: تم تهيئة المريض بماهية المرض و تهيئته نفسيا لتقبل أية مضاعفات من الوارد حدوثها و التعامل معها و خصوصا عدم توقع حدوث تحسن و استرداد بالقوة الحركية مباشرة بعد التدخل الجراحى و أن ذلك يتطلب عدة عوامل من أهمها المواظبة على برامج العلاج الطبيعى و التأهيل. أجريت جراحة لقطع الصفائح العظمية للفقرات الصدرية من الفقرة الثالثة و حتى السادسة ثم فتح الأم الجافية ثم فصل تشريحى لنسيج الحبل الشوكى و استئصال الكتلة الغريبة و التى أثبت تحليل الأنسجة كونها كيس بشرى.

الاستنتاج: يمكن النظر في تشخيص الكيس البشرى داخل الام الجافية داخل الحبل الشوكى في المرضى الذين يعانون من ضعف ببطء تقدمي في أطرافه السفلي وآلام الظهر المستمر الخفيفة

ينبغى أن يشتبه فى وجود الحويصلات البشرية داخل أنسجة الحبل الشوكى و ذلك كما توقعه طاقم الجراحة نظرا للشكل الخارجى الأملس و المحتوى اللين لتلك الكتلة الغريبة. بعد أربعة عشر يوما من الجراحة تحسنت القوة الحركية للطرفين السفليين بشكل ملحوظ. و هذه الورقة البحثية تعرض و توثق حدوث وجود الكيس البشرى فى داخل نسيج الحبل الشوكى بالفقرات الصدرية.