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Case report

Recurrent lumbar intraspinal epidermoid cyst presenting as lower back pain, a case report

Fajr M.A. Sarhan [a,*], Ahmed Mahmoud Daqour [b], Zahra Hosam Abu-Harb [a], Asaad Al-Darawish [b], Fidaa Zakaria Ganaim [a]

[a] Al-Quds University-School of Medicine, Abu-Dis, East Jerusalem-West Bank, Palestine
[b] Al-Makassed Islamic Charitable Hospital, East-Jerusalem, West Bank, Palestine

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ABSTRACT

Introduction: Intraspinal Epidermoid Cyst (IEC) is a very-rare, yet one of the most benign tumors of the Central Nervous System (CNS). Very few cases of IEC were reported world-wide. Presentations depend on the location of the tumor with pain being the most common presenting symptom.

Case presentation: A-35-year-old patient presented to the neurosurgical department for a regular follow up. Her history dates back to four years ago, when she first presented with back pain, which was later accompanied with fecal and urinary incontinence. Magnetic Resonance Imaging (MRI) showed a tumor at the level of L4-L5 which was surgically removed. Three years later, the patient presented to the clinic with similar symptoms. A repeat MRI showed a mass at L4-L5, which was surgically removed. Histological findings of both surgeries showed findings consistent with IEC. Currently the patient has no complaints.

Clinical discussion: IEC has various presentations. Our case presented with back pain, and urinary and fecal incontinence. Diagnosis depends on imaging and histology. MRI is the imaging modality of choice for those tumors in the CNS. Surgical removal is associated with high recurrence as a result of the suboptimal resection due to the strong adhesions between the capsule and the spinal cord.

Conclusion: Pain is the most common presenting symptom for IEC. Adjuvant radiotherapy, which should be implemented in the standard of care, is associated with a lesser degree of recurrence, in addition to regular follow-ups.

1. Introduction

Intraspinal epidermoid cyst is one of the most benign and very rare tumors of the central nervous system [1–3]. Two etiologies for the intraspinal epidermoid cysts were documented, the first being congenital, which is due to ectodermal tissue displacement during the neural tube formation, while the second being an acquired form from the repeated traumas like the lumbar puncture, and previous surgeries [1–8].

Presentations depend on the site of the tumor, while pain is the most common presenting symptom. Urinary and fecal incontinence can happen too [6,9,10].

In this article, we present a case of a recurrent case of intraspinal epidermoid cyst presenting as back pain, urinary and fecal incontinence. To our best knowledge, this is one of the few reported cases world-wide concerning this rare entity.

2. Case presentation

A 35-year-old female patient presented to the hospital for a follow-up after her third spinal surgery. The patient's history dates back to 4 years ago, when she presented to the neurosurgical clinic complaining of lower back pain of four-month-duration. The patient is a non-smoker and is not taking any medications. The patient doesn't have any chronic medical condition. No other family member has similar findings to the patient. The pain was not associated with weakness, sensory deficits or urinary and fecal incontinence. X-ray was done, and no findings of clinical significance were seen. The patient was then treated as a case of lower back pain with ibuprofen. The symptoms kept on worsening, as the patient mentioned, and had her presenting to the clinic two months later with pain accompanied by urine and fecal incontinence. Physical examination showed a conscious, oriented alert patient with equal pupils and both similarly reactive to light. Cranial

* Corresponding author.
E-mail address: fajr.sarhan@students.alquds.edu (F.M.A. Sarhan).

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nerves are intact, upper limbs power is 5/5 and reflexes are +2, sensations are normal. Lower limb muscle power examination findings showed bilateral findings of the following: extensor hallucis longus power of 4/5, dorsiflexion power of 4/ 5, plantar flexion power of 4/5, and proximal hip flexion power of 4/5. Rest of the muscles are 5/5. Sensation is decreased at the S1 dermatome. The patient had both urinary and stool incontinence. Hoffman and Babinski signs were negative. The patient's past medical and surgical history is positive for spina bifida occulta at the level of L4, which was surgically corrected at the age of 12. A decision for Magnetic Resonance Imaging (MRI) showed intradural space occupying lesions at the L4-L5. Fig. 1(A, B, C) shows the perioperative MRIs of the patient's spinal cord.

Surgical evacuation of the mass was discussed, and the patient showed positivity towards the surgical approach. Surgery was performed by the chair of the neurosurgical department. The surgical course was conducted with a midline incision, with resection of the intradural space. No intraoperative complications were noticed. The mass was sent to the pathology department. It was received in formalin, and consists of two pieces of soft tissues, measuring 2.5 and 1.5 cm.

The patient was then monitored at the neurosurgical department.

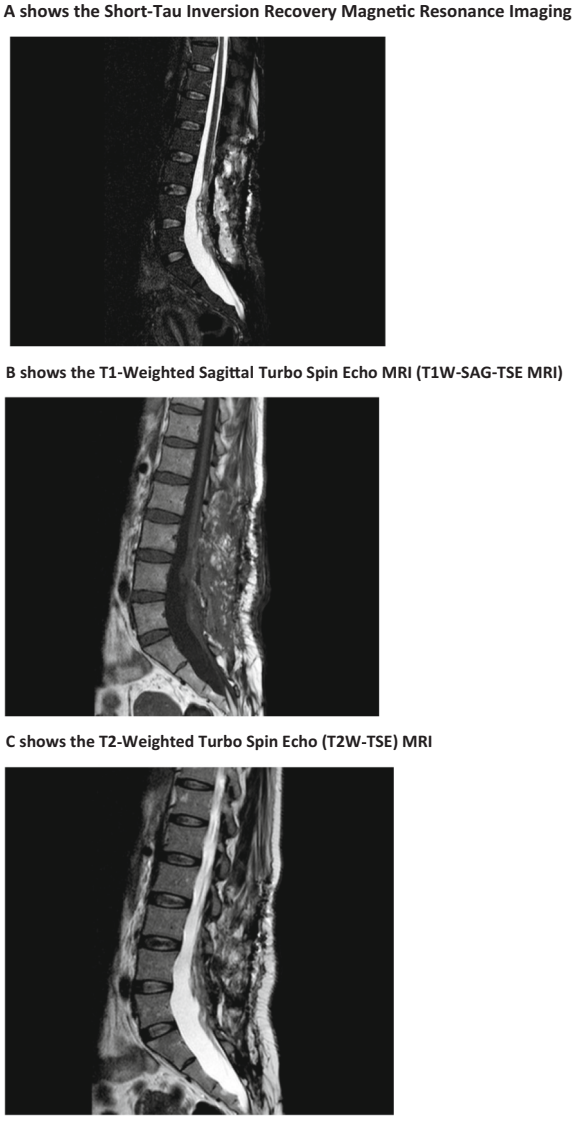


Fig. 1. A shows the Short-Tau Inversion Recovery Magnetic Resonance Imaging (STIR-MRI).
B shows the T1-Weighted Sagittal Turbo Spin Echo MRI (T1W-SAG-TSE MRI).
C shows the T2-Weighted Turbo Spin Echo (T2W-TSE) MRI.

The post-surgical course showed a healing scar. Physiotherapy was started and the pain started to elude. Urinary and fecal incontinence started to improve as well as the sensory findings. Histological description of the specimen confirmed the finding of epidermoid cyst. The patient was informed of the diagnosis, and advised for interval follow-ups at the neurosurgical department.

Three years later, the patient returned to the clinic complaining of weakness, paraparesis, urinary and fecal incontinence, and lower back pain radiating to both legs. Physical examination findings were similar to the time when she presented three years ago. MRI was repeated and showed a mass at the L4-L5 region. Fig. 2(A, B, C, D) shows perioperative spinal cord imaging.

Vital signs were within the normal range. A decision for a surgical resection was made after a thorough discussion with the patient. The patient showed frustration due to the need for a new surgery. Consent for the surgery was taken. The mass was evacuated and sent to pathology which showed findings consistent with epidermoid cyst.

Currently the patient is symptom free, and follows up regularly at the neurosurgical clinic. Discussion on the necessity of regular-timed follow ups, and non-timed visits if symptoms happen were discussed with the patient. The patient is adherent to the follow up measures and timing.

3. Clinical discussion

Intraspinal Epidermoid cyst is a very rare tumor that is considered to be one of the most benign tumors of the CNS [1,2]. In addition to being a slow growing tumor, it accounts for less than 1 % of the tumors in the CNS. Octavian-Mihai Sîrbu et al. estimated that in the year 2019, there have been a very limited number of cases world-wide with a slight female predominance [3].

Two documented etiologies were found for the Intraspinal epidermoid cysts; a congenital form that happens from the anomalous displacement of the ectodermal tissue during the closure of the neural tube. Spinal dysraphism, dermal sinus, spina bifida and hemi-vertebrae are associated with the congenital cause. The acquired form is from repeated traumas, such as lumbar punctures [1,2,4–6]. In our case, congenital etiology is suggested as the patient underwent repair for spina bifida occulta at the age 12.

If anatomically dividing the cause of the epidermoid cyst to anterior to and posterior to the spinal cord, it has been seen that the anterior to the cord lesions are due to congenital malformation, and those posterior to the cord are due to the failure of surgical repair [7,8]. There has been some documentation that malignant transformation can happen, but is rare [1]. Malignant transformation can cause rapid onset of various neurological symptoms depending on the location of the tumor [2].

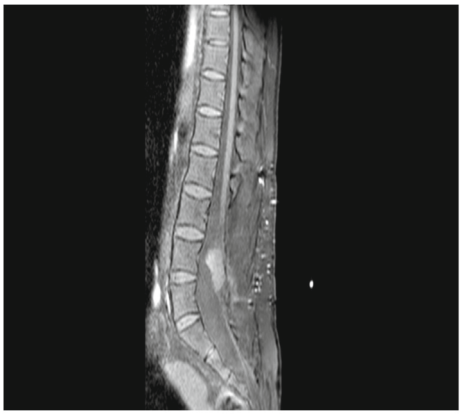
The way that epidermoid cyst presents depends on the location of the tumor. The presentation can take up to years from the onset of tumor to the time it presents [6]. While pain is the most common presentation, bowel and urinary incontinence and other neurological symptoms can also occur, separately or concurrently [9,10]. Our case presented with back pain radiating to the lower limbs, associated with sensory and motor deficits, and urinary and fecal incontinence.

Magnetic Resonance Imaging (MRI) is the imaging modality of choice [4]. Epidermoid cyst on MRI can show up as a hyperintense area compared to the hypo-intense CSF, in addition to lobulation; Tumor's degree of hyperintensity is different as the composition of the tumor is made of both proteins and lipids [1,6,11]. This benign tumor is well demarcated and GAD may show peripheral enhancement [6]. Refer to Figs. 1 and 2 for our patient's imaging.

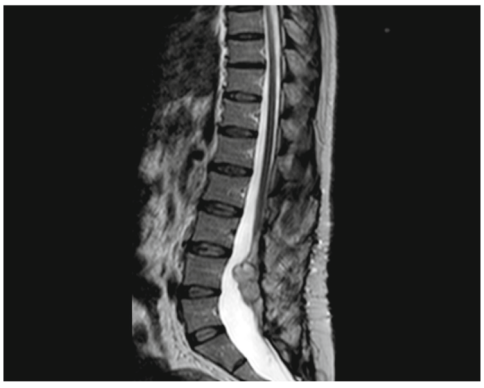
Gold standard for the diagnosis is a biopsy that shows a collagenous layer supporting a lined stratified squamous epithelium. Keratin shedding is the cause of cholesterol crystals formation. [1,6,11] The main differentiation in histology between the epidermoid and dermoid is the presence of the skin adnexa in the dermoid tissues [6]. Grossly, the cyst contains a considerable amount of fat compared to the cholesterol [1].

Managing Intraspinal epidermoid cysts can be of considerable

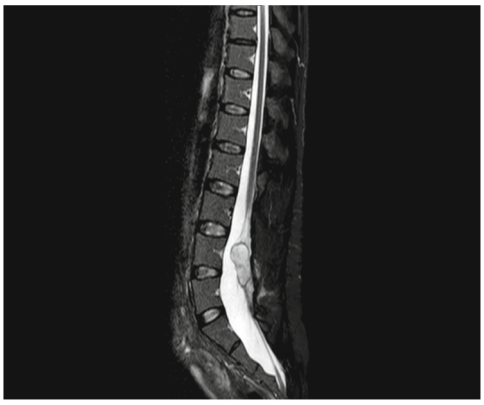
A shows T1-Weighted Turbo Spin Echo-Fat Saturation (T1W-TSE-FS) MRI



B shows T2-Weighted Turbo Spin Echo (T2W-TSE) MRI



C shows Short Tau Inversion Recovery Turbo Spin Echo (STIR_TSE) MRI



D shows a T1-Weighted Turbo Spin Echo-CR (T1W-TSE-CR) MRI

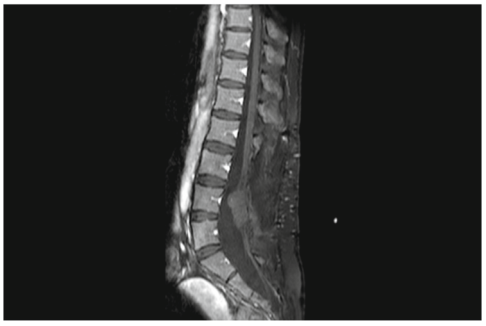


Fig. 2. A shows T1-Weighted Turbo Spin Echo-Fat Saturation (T1W-TSE-FS) MRI.
B shows T2-Weighted Turbo Spin Echo (T2W-TSE) MRI.
C shows Short Tau Inversion Recovery Turbo Spin Echo (STIR_TSE) MRI.
D shows a T1-Weighted Turbo Spin Echo-CR (T1W-TSE-CR) MRI.

difficulty. For asymptomatic tumors, watchful waiting is needed, while surgery is needed for the symptomatic tumors. Surgical treatment can cause recurrence of the cyst. The reason for recurrence is the suboptimal resection due to the strong adhesion between the capsule and spinal cord, most commonly, or between the capsule, and the arachnoid membrane, or nerve roots. Recurrent surgeries do increase the difficulty in treatment of the recurrence as the adhesiveness and scar tissues can be seen on the epidermoid cysts [1,11]. Our patient had a recurrent epidermoid tumor after the first surgery. The cyst occurred at the same location as the previous one, which was L4-L5.

Radiotherapy is to be recommended in those with many recurrences as it can slow down the recurrence [1,7,8]. Regular follow ups are needed to detect early recurrence [7,8].

4. Conclusions and take home messages

In this article we present a case of intraspinal epidermoid cyst, which is a rare tumor with a high recurrence rate after the surgery. To our knowledge, very few cases were reported world-wide. Surgical resection is associated with a high recurrence rate. Current literature indicates that adjuvant radiotherapy slows down the recurrence. More investigations are needed for better surgical therapy.

5. Methods

This case has been reported in line with SCARE 2020 guidelines [12].

Consent

Written informed consent was obtained from the patient and the family for Publication of this case report and accompanying images. A copy of the informed consent is available for review by the Editor-in-Chief of this journal on request.

Ethical approval

The study is exempt from ethnical approval in our institution.

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Guarantor

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Research registration

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Author contribution

Study concept or design: Fajr M A Sarhan, Zahra Hosam Abu-Harb.
Data collection: Asaad Al-Darawish, Ahmed Mahmoud Daqour, Fidaa Zakaria Ganaim.
Writing the paper: Fajr M A Sarhan, Ahmed Mahmoud Daqour, Zahra Hosam Abu-Harb, Asaad Al-Darawish, Fidaa Zakaria Ganaim.

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Declaration of competing interest

The authors declare no conflict of interest.

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