Pediatric Dorsal Intradural Extra-Medullary Epidermoid Cyst (Non-Dysraphic): An Illustrative Case with Literature Review

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Abstract

Background: Spinal epidermoid cyst is a rare congenital tumour that may be associated with spinal dysraphism. Congenital spinal epidermoid cysts without spinal dysraphism are rare. Here we are reporting a rare and interesting case of thoracic Intradural extra-medullary epidermoid cyst not associated with spinal dysraphism. Epidermoid cysts (EC) represent less than 1.0% and are rarely seen as an intradural extra-medullary spinal cord tumor. Preoperative Clinico-radiological assumption is important for surgical planning. [Journal of National Institute of Neurosciences Bangladesh, January 2023;9(1):88-95]

Keywords: Epidermoid; Spinal dysraphism; Intradural Extra-medullary; Dorsal

Introduction

Congenital epidermoid cysts originate from displaced ectoderm inclusions arising in early fetal life, between 3 and 5 weeks of gestation¹. They typically occur in pediatric patients and may be associated with dysraphism and/or dermal sinus. Given the pathogenesis of acquired spinal epidermoid cysts, the lumbar spine represents the most common location, whereas epidermoid cysts of the thoracic and cervical spine have been less frequently described². The pediatric population is more frequently involved³. Barbagallo et al³ and Anuj et al⁴ revealed that only eight cases of isolated spinal intradural extra-medullary epidermoid cysts had been reported six cases of which were in the thoracic region. Akgun et al⁵ reported a case of intradural extra-medullary epidermoid cyst in a 3-year-old male at L1 to L2 level presenting with increasing leg weakness⁶. Ganapathy et al⁷ reported another case of spinal intradural extra-medullary epidermoid cyst in a 1-year-old female child at the level of D-10 presenting to the outpatient clinic with a brownish discoloured skin patch on the back with a slight swelling observed since birth⁷. This brings the total number of reported spinal intradural extra-medullary epidermoid cysts to 11, including ours out of which only 8 are reported in thoracic location. Spina bifida, spinal dysraphisms, scoliosis and cutaneous/dermal defects are often associated with congenital ECs⁸⁻¹¹.

Epidermoid cysts (ECs) are benign tumours, which are commonly observed in the intracranial region. Cruveilhier originally named ECs “tumeurs perlées (pearly tumours)” in 1829, because they had a pearl-like appearance and After extensive literature review we found this case is rare¹²⁻¹⁴. Although ECs rarely occur in the intraspinal region accounting for less than 1.0% of all primary spinal cord tumours spinal ECs may cause spinal cord or nerve root compression enlarging the spinal canal¹⁵. Since Chiari first reported an intramedullary EC in 1883¹⁶, over 100 cases of spinal ECs have been reported in the literature¹⁷⁻¹⁸. It has been reported that the etiologies of spinal ECs are both congenital and acquired. Congenital spinal ECs are thought to have arisen from aberrant ectodermal cells during the closure of the neural tube in the embryonic period, and spinal
ECs in the conus medullaris region were reported to occur as an intramedullary tumour\(^6\,18\,19\).

**Case Presentation**

A 3-year-old boy presented with paraparesis, MRC grade III in Rt lower limb and grade IV in Lt lower limb and urinary retention for 3 weeks, which were gradually increasing. Cutaneous signatures of spinal dysraphism were not observed.

**Neuro Imaging Study:** Magnetic Resonance Imaging (MRI) revealed Intradural Extramedullary (IDEM) SOL in D3 to D4 levels (Figure Ia, Ib, Ic, Id). Mixed intensity mass without any enhancement with gadolinium (GAD), compresses the spinal cord in the intradural extramedullary compartment.

**Operation Note:** A French door laminoplasty D3-5 was performed under the guidance of C-arm. A typical pearly white, avascular, lesion appeared under the dura, on the left side, covered by the arachnoidal sheet and entirely occupying the dorsal portion of the spinal canal (Figure IIa). Following arachnoidal opening over the most prominent tumour component, microsurgical debulking of the tumour was started. (Figure IIb). After its reduction in volume, careful identification and dissection of the tumour membranes were started, trying
involved 3-4. Barbagallo et al2 and Anuj et al5 revealed that reported a case of intradural extra-medullary epidermoid described2. The pediatric population is more frequently spinal epidermoid cysts, the lumbar spine represents the and/or dermal sinus. Given the pathogenesis of acquired 8 are reported in thoracic location. Spina bifida, spinal (pearly tumours)” in 1829, because they had a pearl-like commonly observed in the intracranial region. cord or nerve root compression enlarging the spinal of the neural tube in the embryonic period, and spinal arisen from aberrant ectodermal cells during the closure acquired. Congenital spinal ECs are thought to have in D3 to D4 levels (Figure Ia, Ib, Ic, Id). Mixed intensity (MRI) revealed Intradural Extramedullary (IDEM) SOL occur as an intramedullary tumour9,18-19. ECs in the conus medullaris region were reported to dissection of the tumour membranes were started, trying debulking of the tumour was started. (Figure IIb). After during the dissection of tumour membranes off the right capsule from the arachnoid membrane. The capsule was subarachnoid compartment was also exposed and ligaments were visualized on both sides. The anterior the presence of the postero-lateral arachnoid septum, inadvertent traction of neural structures was helped by arachnoid septum toward the midline. Avoidance of compartment is very important. A good preoperative differential diagnosis. Complete resection as well as extradural lesion an epidermoid cyst keeps as a uncommon, it is not rare. For any dorsal intradural compartment is rare, it tends to occur in the thoracic level. They typically septations were ever seen. The dorsolateral septa similar tortuosity on the midline dorsal septum (Figure extends from the mid-cervical area down to upper staggers from the mid-cervical area down to upper middorsal vein. This midline dorsal septum typically are two series of dorsolateral septa, one series on each compartment is very important. A good preoperative awareness of the arachnoid septations can allow for an easier recognition of a plane of dissection at operation. However, deviation by tumour mass may allow for knowledge of the arachnoid septations is thus helpful in planning the operative approach. Both the medial dorsal septum and right postero-lateral septum had been stretched by the tumour mass and dislocated over the right dorsal rootlets. Consequently, the dissection of tumour membranes from arachnoidal planes on the right side did not cause any traction on nerve roots (Figure IIIb). Conversely, on the medial side tumour membranes were adherent to the left postero-lateral arachnoid septum. This was widely fenestrated and pushed over the left dorsal nerve rootlets. A gentle dissection of left-sided residual tumour membranes was started from the postero-lateral arachnoid septum toward the midline. Avoidance of inadvertent traction of neural structures was helped by the presence of the postero-lateral arachnoid septum, albeit incomplete, covering the left dorsal rootlets. After removing the tumour and its membranes denticulate ligaments were visualized on both sides. The anterior subarachnoid compartment was also exposed and fenestrated at the end of the procedure. (Figure IIIa). The tumour was encapsulated, and there was mild adhesion between the tumour and the arachnoid membrane. Initially, we attempted to separate the capsule from the arachnoid membrane. The capsule was very thin, it was opened, and the content was pearly white. Then, we removed the pearly white content.
involved 3-4. Barbagallo et al 2 and Anuj et al 5 revealed that thoracic and cervical spine have been less frequently most common location, whereas epidermoid cysts of the and/or dermal sinus. Given the pathogenesis of acquired swelling observed since birth 7. This brings the total brownish discoloured skin patch on the back with a slight number of reported spinal intradural extra-medullary found this case is rare 12-14.  Although ECs rarely occur in Cruveilhier originally named ECs “tumeurs perlées canal 8,15.

mass without any enhancement with gadolinium (GAD), Neuro Imaging Study: dissection of the tumour membranes were started, trying fenestrated and pushed over the left dorsal nerve the dissection of tumour membranes from arachnoid dislocated over the right dorsal rootlets. Consequently, septum had been stretched by the tumour mass and side of the spine. (Figure IIc) It was fenestrated and very thin, it was opened, and the content was pearly membrane. Initially, we attempted to separate the subarachnoid compartment was also exposed and removing the tumour and its membranes dentate lamellated layers of keratin. The cyst wall is composed of fibro collagenous tissue. No glial, nerve sheath or meningotheial tumour is seen. No evidence of malignancy is seen.

Gross total removal of the lesion was achieved with the help of microscopy, the upper and lower end of the tumour were protected with cotton to prevent any spillage. Per-operative intravenous methylprednisolone was used. After the removal of the content, the capsule was meticulously separated from arachnoid matter. Gross total removal (GTR) achieved. The wound was closed in layers. No complications after surgery occurred. Motor power regained slowly, two months after surgery patient was running.

Histopathology Shows a cyst lined by stratified squamous epithelium. The cavity contains lamellated layers of keratin. The cyst wall is composed of fibrocollagenous tissue. No glial, nerve sheath or meningotheial tumour is seen. No evidence of malignancy is seen. (Figure IVa, IVb)

Post-Operative Follow Up: There were no residual or recurrence of lesions seen in MRI (Plain and contrast) on 6 months’ follow-up. No compression was detected. Lamina maintains its anatomical configuration (Figures Va & Vb). Patient motor and autonomic function was normal.

Discussion Since the clinical symptoms of spinal ECs are variable and nonspecific with slow progression, the diagnosis

Figure IVa & IVb: Photomicrograph Show a cyst lined by stratified squamous epithelium. The cavity contains lamellated layers of keratin. The cyst wall is composed of fibro collagenous tissue. No glial, nerve sheath or meningotheial tumour is seen. No evidence of malignancy is seen.

Figure Va, Vb: Post-Operative MRI, Post-Operative MRI. No tumour identified in post-operative MRI show spinal cord has no residual or recurrence of the tumour.
can be delayed. In this case report, the patient visited us two weeks after he noticed weakness in both lower limbs. However, the majority of patients with spinal ECs usually have long disease duration due to their slow-growing nature.

Roux et al., reviewed 47 cases of intramedullary ECs, and they reported that the mean disease duration was 6 years. Morita et al., reviewed the literature on 81 spinal ECs since 1962, and they found that the mean disease duration was 26 months in congenital ECs and 15 months in acquired ECs. Currently, the disease duration seems to be shorter because of the introduction of MRI15. To the authors’ best knowledge, this is the fifth case reported of a congenital pediatric spinal IDEM Epidermoid cyst.

Spinal ECs are difficult to diagnose on clinical symptoms alone. Penisson-Besnier et al. first reported an intramedullary EC diagnosed by MRI. MRI is useful for the diagnosis of spinal ECs, demonstrating that the tumour is hypo-intense on the T1-weighted view and hyper-intense on the T2-weighted view. However, it is sometimes difficult to detect small cysts, because spinal ECs show the same signal as the cerebrospinal fluid on MRI. Discrepancy in the intensity characteristics occurs due to varying lipid and protein components.

Histological examination is critical for differentiating ECs from other tumours, especially from a dermoid cyst. A specific feature of the histological finding is a lined stratified squamous keratinizing epithelium surrounded by the outer layer of collagenous tissue with the absence of skin adnexa. The desquamation of keratin from the epithelial lining creates numerous cholesterol crystals.

The etiologies of spinal ECs are thought to be both congenital and acquired. Acquired ECs were mostly reported in the region of the cauda equina below the L1 level. Among the spinal epidermoids, the thoracic region is the most common site of occurrence, followed by the sacral and cervical regions.

Surgical treatment is required when the patients develop neurological deficits. Complete excision of ECs is essential for surgical treatment. However, complete resection of the encapsulated tumour is difficult for all cases, because the EC’s capsules are very thin and the tumour often adheres to the arachnoid membrane, spinal cord, or nerve roots. Thereby, subtotal resection is also commonly performed. The surgical management of such lesions is highlighted and the importance of spinal arachnoidal space anatomy is discussed. Knowledge of “classical” microsurgical anatomy was useful to perform a safe and effective tumour dissection.

The recurrence rate of spinal ECs was reported from 10 to 29.0% in previous literature. Although ECs are benign tumours, local recurrence is reported especially after subtotal excision. It is suggested that incomplete excision of basal germinal cells of the tumour induces tumour recurrence. Because the tumour content includes fat and cholesterol, they may also produce an inflammatory reaction leading to meningitis. In our case, complete resection of the tumour’s capsule with emptying of the cyst material was achieved, although the tumour’s capsule opened during the procedure. Fortunately, the patient did not develop any inflammatory reactions or neurological deterioration postoperatively. We use cotton patties to prevent spillage as well as local and systemic methylprednisolone injection to prevent an inflammatory reaction. There was no recurrence at the Six-month follow-up with a good clinical outcome. However, long-term follow-up is required for the potential risk of tumour recurrence.

Anatomical Study: Key, Axel and Magnus Gustaf Retzius published a famous anatomical book “IN 1875 where they describe subarachnoid space. Later, Prof. Yasargil conducted an extensive study on the same issue, they found -the arachnoid membranes consist of an outer condensed layer that is continuous and watertight. From this outer, condensed arachnoid membrane, there are numerous strand-like attachments to the pial surface of the spinal cord which are particularly numerous along the dorsal aspect of the spinal cord and relatively sparser ventrally. In almost

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Table 1: Published Cases of Congenital Pediatric Dorsal Non-Dysraphic Spinal Intradural Extramedullary Epidermoid Cysts are Tabulated

<table>
<thead>
<tr>
<th>SL</th>
<th>Author</th>
<th>Patients Particulars</th>
<th>Location of Lesion</th>
<th>Year</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.</td>
<td>Chang et al.</td>
<td>9 Y/F</td>
<td>Upper thoracic</td>
<td>1996</td>
</tr>
<tr>
<td>2.</td>
<td>Akgun et al.</td>
<td>3 years/ Male</td>
<td>Conus</td>
<td>2018</td>
</tr>
<tr>
<td>3.</td>
<td>Sibhi Ganapathy et al</td>
<td>1 year/ Female</td>
<td>D10</td>
<td>2019</td>
</tr>
<tr>
<td>4.</td>
<td>Swamy et al.</td>
<td>13y/ Male</td>
<td>D5-D7</td>
<td>2002</td>
</tr>
</tbody>
</table>

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92
all specimens, there was a distinct longitudinal midline dorsal septum, variably fenestrated, which extended from the dorsal midline of the outer condensed arachnoid to attach to the subjacent spinal cord along a middorsal vein. This midline dorsal septum typically extends from the mid-cervical area down to upper lumbar cord levels. The midline dorsal vein appears to influence the attachment of the midline dorsal septum to the spinal cord, and when the vein is tortuous, it may impart a similar tortuosity on the midline dorsal septum (Figure III a,b). In addition to the midline dorsal septum, there are two series of dorsolateral septa, one series on each side, with each element of the series oriented along the oblique course of the dorsal rootlets clustered at each segmental level. These septa extend from the dorsal root entry zone to envelop the dorsal rootlets (grouped at each segmental level) and attach to the outer arachnoid membrane dorsolateral. In most cases, the attachment tethers the dorsal rootlets dorsolateral and ensures clearance between the dorsal rootlets and the lateral parts of the spinal cord. The arachnoid enveloping the dorsal rootlets continues laterally on the inner surface of the outer arachnoid membrane to extend into the root sleeve where it appears progressively more condensed as the nerve bundles themselves condense to form the definitive dorsal root. The dorsolateral septa were considerably more irregular than the generally smoother, compact, and distinct midline dorsal septum. Along the ventral surface of the spinal cord, no arachnoid condensations or septations were ever seen. The dorsolateral septa were most distinct at thoracic and low cervical levels but were still evident further rostral and caudal. The pia mater of the spinal cord appeared to condense laterally to form the dentate ligament, which is attached to the outer arachnoid and through it to the overlying dura mater approximately halfway between the root exits. A better appreciation of the spinal arachnoid can be pertinent to the operating neurosurgeon in the following ways.

Knowledge of the arachnoid septations can allow for a smoother opening and subsequent reclosing of the arachnoid. For instance, when the midline dorsal septum is well-formed, it will resist opening exactly on or across the middorsal line. It may be advantageous to open the arachnoid slightly lateral to the middorsal line and then, if necessary, break down the midline dorsal septum separately to preserve the coherence of the outer arachnoid for subsequent closure. Of course, at upper cervical and lower lumbar levels, the midline dorsal septum is rarely encountered in recognizable form, being largely fenestrated at those levels, and one can usually open the arachnoid there in the exact midline as desired. When the dorsal vein, visible through the outer arachnoid, is noted to be tortuous, the surgeon can be forewarned that the midline dorsal septum may be similarly tortuous and an arachnoid incision planned accordingly. Another similar advantage to the neurosurgeon is the knowledge that the arachnoid septations may guide the dissection of a benign tumour. Of course, a tumour may displace the septations, but anticipating the presence of septation. However, deviation by tumour mass may allow for easier recognition of a plane of dissection at operation. A potential advantage may relate to preventing the formation of an arachnoid cyst after surgery. If an arachnoid cul-de-sac is appreciated following the removal of a tumour, it may be advantageous to break down the arachnoid septations to ensure free communication of the spinal fluid compartments involved.

Conclusion
Here we report a case of isolated (non-dysraphic) intradural extramedullary spinal epidermoid cyst which is uncommon, especially in the dorsal area. Though uncommon, it is not rare. For any dorsal intradural extramedullary lesion an epidermoid cyst keeps as a differential diagnosis. Complete resection as well as maintaining spinal anatomy is the goal. Getting an idea about tumour location concerning the arachnoid compartment is very important. A good preoperative image gives an idea and thus helps in the operative decision. A thorough knowledge of spinal subarachnoid space anatomy is helpful to distinguish between tumour membranes and arachnoidal planes and achieve a safe and complete resection, to avoid recurrences.

Abbreviations
EC: Epidermoid cysts
IDEM: Intradural Extramedullary
MRI: Magnetic Resonance Imaging
SOL: Space Occupying lesion
MRC: Medical Research Council

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Contribution to authors: SKM, KNK and AAM did this surgery, SKM write this article, DMA- check grammar then editorial formatting done by JW.

Data Availability
Any inquiries regarding supporting data availability of this study should be directed to the corresponding author and are available from the corresponding author on reasonable request.

Consent for publication: Informed consent was taken from a patient guardian. All methods were performed in accordance with the relevant guidelines and regulations.

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In the literature 17-18, it has been reported that the most common location for ECs is the thoracic spine. Spinal dysraphism often associated with congenital ECs 8-11. ECs are reported in thoracic location. Spina bifida, spinal dysraphism, and/or dermal sinus. Given the pathogenesis of acquired ECs, the term "tumeur perlée" was used to describe these tumours, highlighting their characteristic appearance.

Spinal intradural extra-medullary compartment. ECs are reported in thoracic location. Spina bifida, spinal dysraphism, and/or dermal sinus. Given the pathogenesis of acquired ECs, the term "tumeur perlée" was used to describe these tumours, highlighting their characteristic appearance.

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Cutaneous signatures of spinal dysraphism are often associated with congenital ECs 8-11. ECs are reported in thoracic location. Spina bifida, spinal dysraphism, and/or dermal sinus. Given the pathogenesis of acquired ECs, the term "tumeur perlée" was used to describe these tumours, highlighting their characteristic appearance.

Intraaxial lesions were protected with cotton to prevent any spillage. Per-operative intravenous methylprednisolone were used in the surgical management of such lesions. Results of the surgical treatment of spinal dermoid and epidermoid tumour dissection 2.

Six-month follow-up with a good clinical outcome. (Figure IVa, IVb) MRC: Medical Research Council

MRI: Magnetic Resonance Imaging
EC: Epidermoid cysts

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