

CASE REPORT

A Case of Emerging Epidermoid Cyst After Excision of Benign Cystic Teratoma Located in the Conus Medullaris

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Abstract

Teratomas constitute 0.1% of all spinal cord tumors and contain tissues originating from the three germ layers. The aim of surgery in spinal teratomas is total excision. However, total excision of tumors in the conus medullaris may lead to potentially significant morbidities. We aimed to present an epidermoid cyst (EC) after subtotal resection of teratomas. A 19-year-old woman complained of low back pain and numbness in the posterior aspect of the left leg. Magnetic resonance imaging (MRI) showed a tethered cord and an intradural lesion at the L4 level. During surgery, part of the capsule was left. Histopathological examination was reported as a benign cystic teratoma. Thirty-three months later, a control follow-up MRI showed a cystic lesion at the L4 level. After the total excision of the lesion, a neuro deficit developed. Histopathological examination was reported as EC. Spinal ECs account for less than 1% of spinal tumors. These tumors are lined with stratified squamous epithelium, similar to the skin's epidermis, and arise from the pathological displacement of epidermal cells into the spinal canal. It should be remembered that EC may develop after incomplete resection of conus medullaris-localized teratomas, and complete resection of these masses may result in neurological deficits.

Keywords: Benign cystic teratoma; conus medullaris; epidermoid cyst; surgical treatment.

Spinal teratomas, which are extragonadal germ cell tumors, contain tissues originating from three germ cell layers.^[1,2] They are infrequent and constitute 0.1% of all spinal cord tumors.^[1,3-6] Two mechanisms have been proposed to explain the origin of spinal teratomas. According to the dysembryogenic theory, these masses result from the chaotic differentiation of pluripotent cells in a locally disrupted developmental environment.

^[2,6] According to the misplaced germ cell theory, the pluripotent germ cell of the neural tube is lost during its passage from the yolk sac to the gonads, leading to spinal teratomas. Furthermore, this mechanism is the most probable reason for the development of spinal teratomas that occur with spinal dysraphism in adults.^[1,2,6] Teratomas are classified as mature, immature, or malignant according to the degree of differentiation.^[2] While mature teratomas

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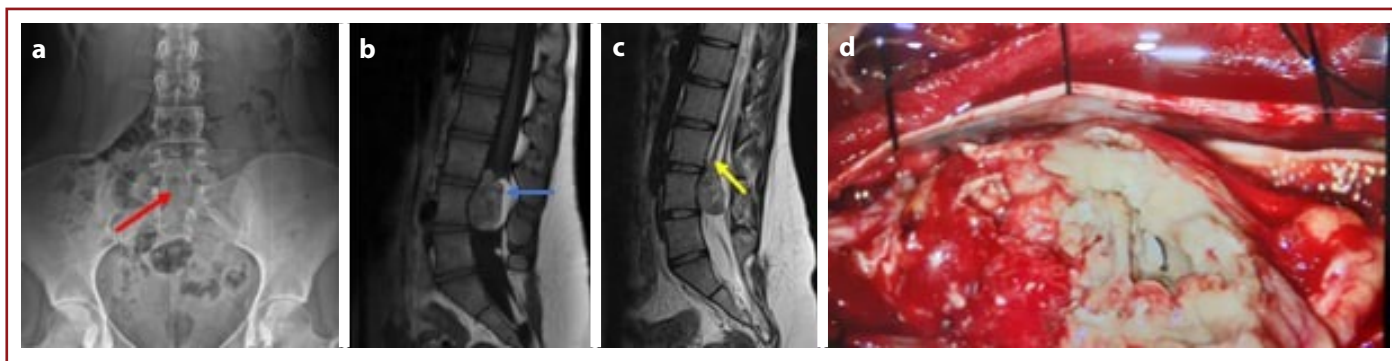


Figure 1. Closure defect in the L5 lamina and sacrum on lumbar radiograph (red arrow) (a); Initial preoperative lumbar MR imaging on T1 sequence: Craniocaudal dimension 42 mm, anterior-posterior diameter 30 mm, oval configuration solid mass lesion, located intradural at the level of L4, terminating at the level of the conus medullaris L3, with hypointense signal in the anterior part and hyperintense signal in the posterior wall (blue arrow) (b); The lesion has heterogeneous signal intensity on T2, there is a syringomyelic cavity cranial to the lesion (yellow arrow), the posterior wall of the lesion appears hyperintense on T2 (c); Image from the first operation showing an encapsulated mass containing hair, fat, neuronal tissue, and fine-white keratinized tissue (d).

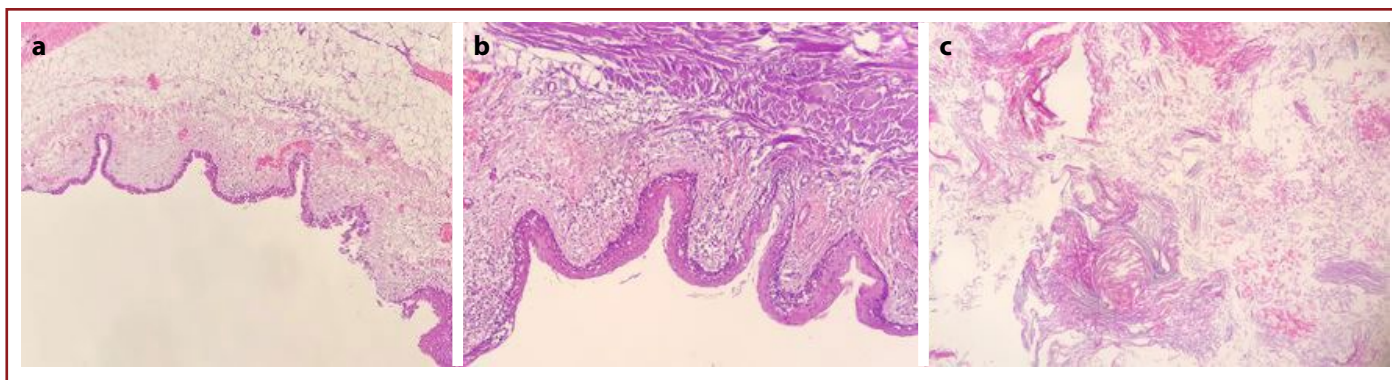


Figure 2. Cyst wall lined with keratinized squamous epithelium and ciliated pseudostratified epithelium, H&E x40 (a); Cyst wall lined with keratinized squamous epithelium, H&E x100 (b); Lamellar keratinous material, epidermoid cyst contents, H&E x40 (c).

contain well-differentiated tissue, immature teratomas contain poorly differentiated non-malignant tissue. Malignant teratomas are highly aggressive and have a poor prognosis.^[7] The aim of surgery in spinal teratomas is the total removal of the mass.^[5,8,9] However, total resection of approximately half of the teratomas is difficult due to their tendency to adhere to surrounding tissues. Subtotal resection is recommended for protecting the surrounding neuronal tissues. Since total and subtotal resection have similar recurrence rates (9% and 11%, respectively), aggressive resection is unnecessary.^[2,7] Recurrence rates also depend on the histopathological features of the tumors, and it has been found that recurrences are more frequent in immature and malignant teratomas.^[10] It has been reported that symptomatic recurrence rates of mature teratomas are very low, even in subtotal resections.^[10] Our case is the first in the literature to report the development of an epidermoid cyst (EC) after the subtotal excision of a mature teratoma localized in the conus medullaris.

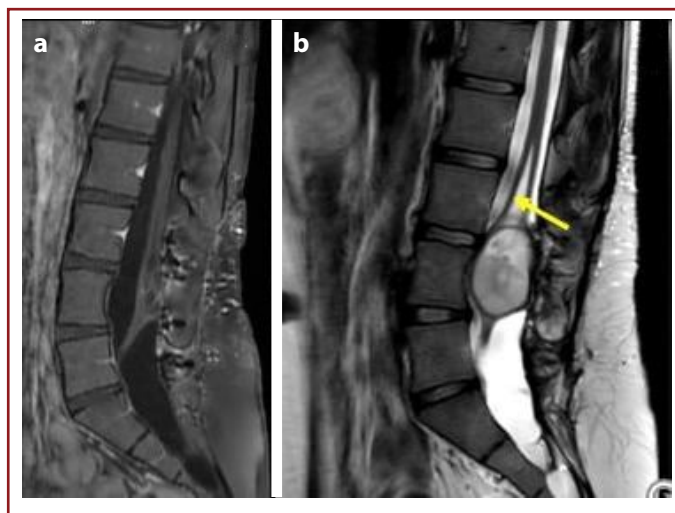


Figure 3. Control MRI examination 33 months after the first operation, non-contrast enhancing cystic mass lesion with a craniocaudal dimension of 40 mm and an anterior-posterior diameter of 25 mm at the L3-4 level on T1+C sequence (a) and oval configuration on T2 sequences, there is a syringomyelic cavity cranial to the lesion (yellow arrow) (b).

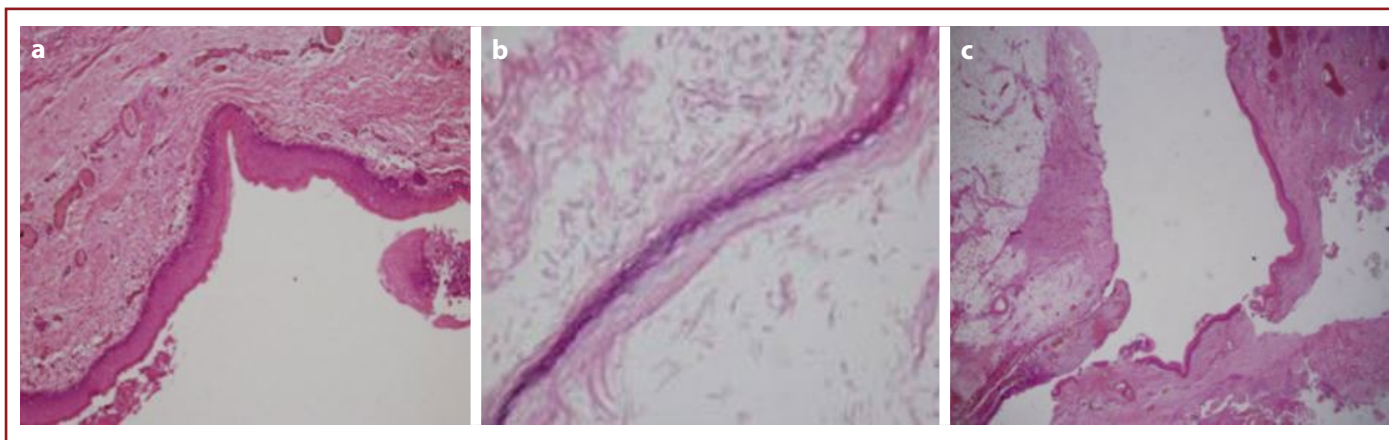


Figure 4. Epidermal cyst, lining with stratified squamous epithelium with granular layer, H&E x40 (a); Characteristic keratin material, H&E x40 (b); Epidermal cyst, lining with stratified squamous epithelium with granular layer, H&E x20 (c).

Case Report

A 19-year-old woman complained of low back pain and numbness in the posterior aspect of the left leg for five years. On neurological examination, she presented with hyperactive Achilles and patellar reflexes and positive Babinski sign bilaterally. Radiographic examination and spinal CT showed spinal dysraphism (Fig. 1a). Magnetic resonance imaging (MRI) showed an intradural lesion approximately 42x30 mm in size at the L4 level, which almost completely obliterated the spinal canal and had marked contrast enhancement posteriorly (Fig. 1b). The conus medullaris terminated at the L3 level. There was filum thickening as a tethered cord sign at this level. The anterior-posterior diameter of the spinal canal was increased at the L4 level (Fig. 1c). Intraoperative neurophysiologic neuromonitoring (IONM) guided L3-4 laminoplasty with mass excision and filum terminale release was performed.

Part of the capsule was not excised during surgery to avoid damage to neuronal tissues. Peroperatively, the mass contained hair, fat, neural tissue, and pearly white keratinized tissue (Fig. 1d). Postoperative neurological examination was normal. Histomorphologic findings in the tissue samples were evaluated in favor of benign cystic teratoma (Fig. 2). The patient was readmitted 33 months after the operation with complaints of pain in the lower back and lateral parts of both thighs, numbness in the legs, perianal and genital area, and frequent urination for the last three months. There was no history of spinal trauma or lumbar puncture. Muscle strength was normal on examination, and hypoesthesia was present in the perianal and genital regions. The bilateral Laseque test was positive, and the patella and Achilles reflexes were hyperactive. MRI revealed a non-contrast-enhancing cystic mass lesion with

an oval configuration with a craniocaudal dimension of 40 mm and an anterior-posterior diameter of 25 mm, which appeared adherent to the cord at the L3-4 level (Fig. 3). On MRI examination, the mass was primarily suggestive of recurrent teratoma. In addition, the patient developed recurrent tethered spinal cord syndrome due to adhesions. The patient underwent IONM-guided L3-4 laminoplasty with excision of the total recurrent mass, adhesions excision, and tethered spinal cord release. The perioperatively mass consisted of pearly white keratinized tissue. Postoperative neurological examination revealed left ankle extension muscle strength of 4/5, urinary incontinence, and perianal and genital hypoesthesia. Histomorphologic findings in the tissue samples examined were reported in favor of EC (Fig. 4).

Discussion

Cruveilhier was the first to describe spinal ECs in 1829 and called them tumors perlées, meaning pearl tumors.^[11] ECs arise from the pathologic displacement of epidermal cells into the spinal canal and are lined with stratified squamous epithelium similar to the epidermis of the skin.^[12-14] Spinal ECs are rare and constitute less than 1% of spinal tumors.^[11,12] Spinal ECs may be congenital or acquired. Congenital ECs are reported to be most commonly located in the conus medullaris region and occur due to accidental inclusion of the ectoderm during the closure of the neural tube.^[11,12,14] In addition, congenital ECs are frequently associated with hemivertebra, dermal sinus, spina bifida, and syringomyelia.^[11] Acquired ECs are frequently found at the L1 level. They are caused mainly by spinal trauma or any invasive procedure such as lumbar puncture or spine surgery that causes accumulation of ectopic epidermal cells in the intradural space and leads to cyst formation.

[11] We think the EC was formed by seeding the epidermal layer into the surgical area during teratoma excision or from the capsule section that could not be excised. Since total excision of tumors in the conus medullaris localization potentially causes significant morbidity,^[15] we attributed the development of neuro deficit after EC resection to the total resection of the capsule, which was tightly adherent to the surrounding tissues.

Conclusion

Our case report adds a new etiology for acquired EC to the literature and suggests that teratoma surgery in the conus medullaris is like a double-edged sword. Incomplete resection of a teratoma may result in EC, and trying to remove a teratoma or any intradural spinal cord tumor totally comes with the risk of postoperative neurological deficit. Neurological surgeons should consider the advantages and disadvantages of the degree of resection when performing this type of procedure.

Informed Consent: Approval was obtained from the patients.

Peer-review: Externally peer-reviewed.

Conflict of Interest: None declared.

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