Child's Nervous System (2019) 35:1411–1413
https://doi.org/10.1007/s00381-019-04144-w

CASE REPORT

Cervical intradural extramedullary epidermoid cyst at the background of congenital scoliosis with a semi-segmented C6 hemivertebra

Veli Umut Turgut[1] · Murat Şakir Ekşi[2] · Ahmet Özak[1] · Emel Ece Özcan-Ekşi[3]

Received: 28 February 2019 / Accepted: 29 March 2019 / Published online: 5 April 2019

Abstract
In the present case study, we present a female adolescent patient harboring a rare clinical presentation of spinal intradural extramedullary epidermoid cyst concomitant with congenital scoliosis. A 17-year-old female patient was admitted to the clinic with long-lasting neck pain. She was neurologically intact yet had cervicothoracic scoliosis. Cervical MRI and CT depicted a right C6 hemivertebra, fused to the lower endplate of the C5 vertebra. At the same vertebra level, she had an intradural extramedullary mass lesion anterior to the spinal cord. We planned to excise the mass lesion first. We used neuromonitoring during the surgery and made the surgery via posterior approach. We observed a pearl-like mass lesion anterolateral to the spinal cord. We excised the mass lesion with its capsule microsurgically via peace-meal route. She was neurologically stable following the surgery. Histopathological diagnosis was epidermoid cyst. Most of spinal inclusion cysts occur secondary to spinal dysraphism or iatrogenic inoculation. Isolated spinal inclusion cyst located anterior to the spinal cord concomitant with vertebral anomalies should be kept in mind before making proper surgical planning. Surgery is the modality of choice for spinal inclusion cyst and should be performed under the guidance of neuromonitoring, especially in cases with lesions located at higher spinal levels.

Keywords Congenital · Epidermoid cyst · Scoliosis · Cervical · Surgery

Introduction

Dermoid and epidermoid cysts are derived from skin appendage and grouped under inclusion cysts. Most of inclusion cysts are congenital in origin. Some inclusion cysts develop after birth. Congenital inclusion cysts develop from ectodermal tissue remnants during neural tube closure period between the 3rd and the 5th weeks of gestation [1–3]. Spinal inclusion cysts are mostly detected in the intradural extramedullary zone (60%) with concomitant dysraphic anomalies such as dermal sinus tract, spina bifida, or enterogenous cyst.

Congenital scoliosis is lateral curvature of the spinal column in coronal plane more than 10° and is present since birth. Incidence of congenital vertebral anomalies is 0.5–1/1000 live birth [4]. Abnormalities of vertebral formation or segmentation appear within 6 weeks of gestation [5].

In the present case study, we present a female adolescent patient harboring a rare clinical presentation of spinal intradural extramedullary epidermoid cyst concomitant with congenital scoliosis.

Case report

A 17-year-old female patient was admitted to the clinic with long-lasting neck pain. She had no skin stigmata. She was neurologically intact. However, she had cervicothoracic scoliosis. On plain anterior-posterior radiograph, coronal Cobb was 41.8°. Cervical MRI and CT depicted a right C6 hemivertebra, fused with the lower endplate of the C5 vertebra

✉ Murat Şakir Ekşi
muratsakireksi@gmail.com

[1] Clinic of Neurosurgery, Antalya Atatürk State Hospital, Antalya, Turkey

[2] School of Medicine, Department of Neurosurgery, Acıbadem University, Göztepe Mah, Mesire Sok, Tütüncü Mehmet Efendi Cad, No:3/34, Tepekule Apart., Kadıköy, Istanbul, Turkey

[3] School of Medicine, Department of Physical Medicine and Rehabilitation, Bahçeşehir University, Istanbul, Turkey

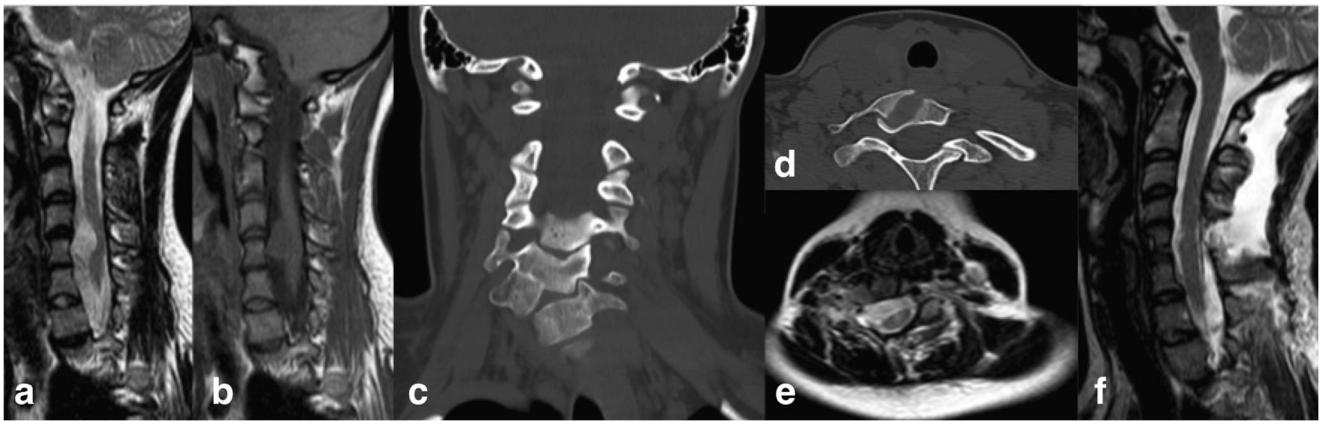

Fig. 1 Cervical sagittal MRIs depict an intradural extramedullary lesion hyperintense on T2- (**a**, **e**), and isointense on T1-weighted (**b**) scans. Coronal (**c**) and axial (**d**) plane CT scan revealed a right C6 semi-segmented hemivertebra fused to the lower endplate of the C5 vertebra. Post-operative T2-weighted cervical sagittal image (**f**) reveals that the lesion has been totally excised

(Fig. 1a–e). At the same vertebra level, she had an intradural extramedullary mass lesion anterior to the spinal cord. The mass lesion was isointense and hyperintense on T1- and T2-weighted MRI scans, respectively. She had no organ anomaly. We planned to excise the mass lesion first. We used neuromonitoring (somatosensory- and motor-evoked potentials) during the surgery and made the surgery via posterior approach. No abnormal neuromonitoring signals were present throughout the surgery. Immediately after opening the dura, cerebrospinal fluid flushed out of the subarachnoid space with much higher pressure than expected. We observed a pearl-like mass lesion anterolateral to the spinal cord. We excised the mass lesion with its capsule microsurgically via peace-meal route. She was neurologically stable following the surgery (Fig. 1f). Histopathological diagnosis was epidermoid cyst with unique appearance of squamous cell lining under lamellated keratin debris (Fig. 2). We discharged her to home in a good condition with a cervical collar and referred her to a tertiary spine center for her congenital scoliosis.

Discussion

Inclusion cysts namely "epi-/dermoid cysts" are derived from cutaneous ectoderm [6]. There is confusion about nomenclature of inclusion cysts. Despite the name "cyst," most of mass lesions in this category are solid or partly cystic. Nomenclature is based upon contents of inclusion cysts whether to call them as dermoid or epidermoid cysts. Dermoid and epidermoid cysts have similar clinical presentation and similar treatment protocols with similar prognoses [6–10].

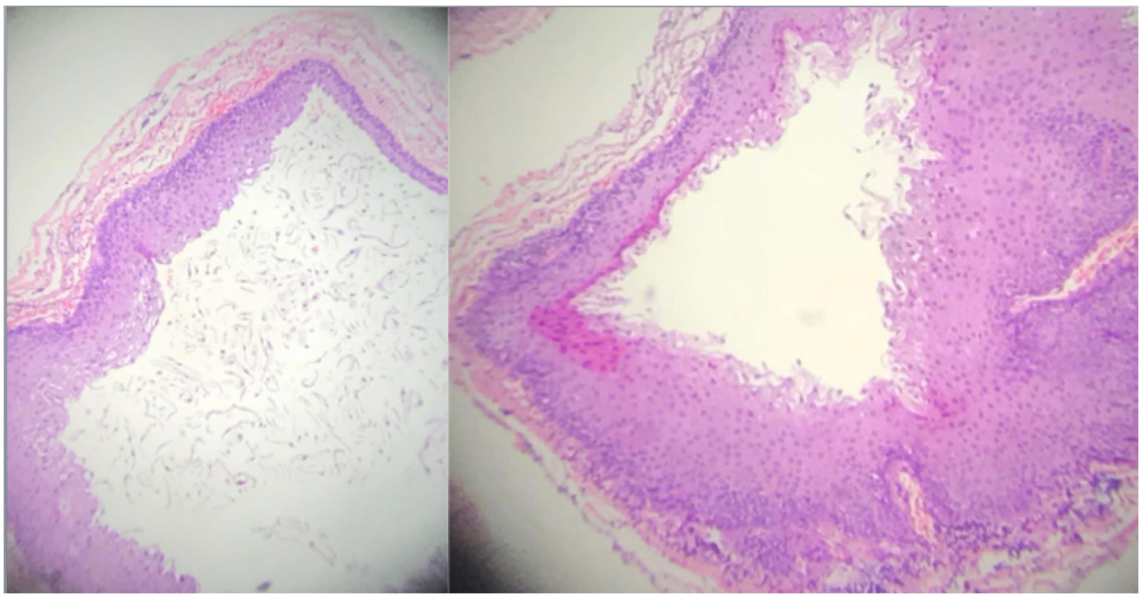

Fig. 2 The histopathological figures show lamellated keratin debris and squamous cell lining of the epidermoid cyst

Spinal inclusion cysts compose 1–2% of spinal tumors [6, 11, 12]. Most of spinal inclusion cysts are diagnosed in pediatric patients, constituting 8% and 17% of spinal tumors seen in children and infants, respectively [6]. Epidermoid cyst predominates intracranial compartment compared with dermoid cyst, whereas vice versa in the spinal canal [6, 7, 13, 14]. Clinical presentation is rather earlier in patients with dermoid cysts compared with those with epidermoid cysts (1.5 years vs. 35 years). Our patient presented at late adolescence period [6, 8, 14]. The lumbosacral region is the most common location of spinal inclusion cysts [6, 8, 11, 15]. Epidermoid cyst in the present case was located at the lower cervical spine, which is a rare spinal location for inclusion cysts [16]. Dermal sinus tract is observed in 73% of patients with spinal inclusion cysts [6]. Our patient had no skin stigmata.

Underlying vertebral anomalies leading to congenital scoliosis are categorized as failure of formation, failure of segmentation, and mixed failure [5]. Failure of formation is named as "wedge vertebra," "hemivertebra," and "butterfly vertebra" according to the type of malformation. Our patient had a C6 vertebra formation anomaly with semi-segmentation, fused with lower endplate of the C5 vertebra on the right side.

Spinal dysraphism is the underlying pathology for spinal inclusion cysts. There are multiple closure sites of the neural tube and boundary zones, especially in the mid-thoracic and lumbosacral regions, with significant risks due to anomalies of dysjunction at the boundary zones. Our patient had cervical intradural extramedullary epidermoid cyst concomitant with spine bony anomaly adjacent to the mass lesion with subtle clinical findings. This scheme makes us think that the pathology might have developed due to a partial defect in the early period of neural tube formation, since there was no dermal sinus tract despite the presence of spinal bony defect. What makes our case further unique is the cyst location anterior to the spinal cord. Only spinal inclusion cysts presented in Currarino syndrome are located anteriorly, yet in only the sacral region. Our patient had no other findings of Currarino syndrome.

Surgery is the choice of treatment, even though there is no obvious sign and symptom of cyst enlargement and cyst infection [6]. Maintenance of spinal cord function is more important than total removal of the cyst, in patients with severe adhesions. It should be kept in mind that tumor recurrence could only be prevented by total resection of the cyst with its capsule. Thus, total resection of the cyst and maintenance of spinal cord function should be balanced using intraoperative neuromonitoring.

Conclusion

Isolated spinal inclusion cyst located anterior to the spinal cord and concomitant with vertebral anomalies could rarely be detected. Surgery is the choice of treatment in patients with spinal inclusion cyst. Surgery should be performed under the guidance of intraoperative neuromonitoring, especially in patients with lesions at higher spinal levels.

Acknowledgments We thank Dr. Aslı Demir, M.D., for her efforts in the preparation of the pathological figures.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

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