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MINI REVIEW

Simultaneous spine extradural and intradural teratomas in a pediatric patient: A rare presentation with insights in the flawed migration of germ cells theory

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Abstract
Spinal teratomas are infrequent lesions in the pediatric population. These lesions can be extradural, intradural or intramedullary. We present a case of an 8-month-old boy that was assessed for underdevelopment of motor milestones. The neurologic examination revealed hyporeflexia, decreased sensation and flaccid paraplegia. MRI of the spine revealed two simultaneous and independent lesions in the extradural and intradural compartment. A laminectomy was performed for the T4-T7 vertebrae with total resection of both lesions. The histopathological analysis confirmed both lesions to be mature cystic teratomas. At the 1-year follow-up, the patient remained with no recovery of neurological function. A debate takes place regarding the etiology of formation of these lesions in the spine. The simultaneous presentation of two independent lesions in this patient could contribute to define the flawed migration of germ cells theory as the etiology for formation of teratomatous lesions in the spine.

Keywords Teratoma · Extradural · Intradural · Spine · Pediatric

Introduction

Teratomas are lesions comprising tissues arising from all three germ cell layers: endoderm, mesoderm, and ectoderm, scarcely seen in the pediatric central nervous system (CNS) accounting for only 0.1% of tumors [1]. Spinal teratomas are rarely seen in children with only a few cases reported (Table 1). They can be located in the intradural, extradural or intramedullary regions with neurological symptoms arising from compression and displacement of nervous tissue [1, 2]. Whereas the morphological and histological composition of the lesions is very variable, most teratomas contain fatty tissue making it an imaging hallmark for its diagnosis and positioning magnetic resonance imaging (MRI) as the preferred tool for the evaluation of its features [3, 4]. The emergence of technology, surgical techniques and the growing experience of surgeons makes complete resection as the main goal for most spinal tumors and teratomas are no exception to this [5]. Although an established mechanism for the formation of spinal teratomas has not been elucidated, some theories are more prone to match its clinical and anatomical characteristics. The purpose of this paper is to present the case of an 8-month-old boy with two teratomas in the thoracic spine and to the best of our knowledge, this is the first report of two independent simultaneous extra and intradural teratomas in a pediatric patient. With this article, we aim to contribute to the available cases in the literature and seek to collaborate in the consolidation of its formation.

Case presentation

An 8-month-old male with a previous history of three episodes of urinary tract infections was admitted for in-patient care. During hospital stay, the patient was noted with underdevelopment of motor milestones, as he dragged his legs while crawling and was unable to stand. The rest of developmental milestones were appropriate according to age. The patient was referred to our department for further evaluation after hospital discharge. The neurologic examination revealed hyporeflexia,

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Table 1 Spinal teratomas previous reports

Author	Age	Sex	Level and location	Preoperative symptoms	Approach	GTR	Diagnosis	Outcome
Quon 2014 [17]	21 m	M	T3–T10 Epidural	• Spasticity • Failure to ambulate • Undeveloped motor milestones	Bilateral laminectomies from T3–T9	Y	Mature teratoma	Restored motor function 6 months after surgery and physical therapy
Basmaci 2011 [9]	5 yrs	M	T12 Intradural Extramedullary	• Pain and numbness in lower extremities • Gait disability • Paraparesis and loss of sensitive function under T12 dermatome • Hyporeflexia	Laminectomy and laminoplasty	Y	Mature teratoma	Neurological recovery
Khan 2021 [26]	27 d	F	T12 Intramedullary	• Poor feeding • Irritability • Progressive hypotonia	Laminectomy and laminoplasty	N	Immature teratoma	Recurrence at 9 months Chemotherapy treatment without recurrence at 5-year follow-up Scoliosis without neurological deficits
Ishiguro 2011 [27]	5 m	M	T1–S5 Intramedullary Associated with spina bifida occulta and lumbosacral lipoma	• Progressive paraparesis • Hyporeflexia in lower extremities • Urinary tract infections and urinary retention • Constipation	Laminotomy and laminoplasty	N	Mature teratoma	Without neurological recovery in the 3-year follow-up
Deckey 2021 [18]	7 m	M	T5–T6 Paravertebral and epidural with spinal stenosis	• Thoracic scoliosis • Asymptomatic thoracic spinal prominence since birth • Hypotonia	Laminectomy and decompression with three level neurotomy	Y	Mature teratoma	Improved tone in lower extremities Normal motor development at follow-up
Yoshioka 2018 [28]	64 d	F	C7–L1 Epidural Extended bilaterally through foramina to the mediastinal region	• Dyspnea • Paraparesis	Laminotomy	N	Immature teratoma	Reincidence No neurological improvement
Shimauchi 1988 [29]	4 yrs	M	C7–T6 Intradural extramedullary Associated with spina bifida	• Gait disturbances • Paraparesis • Hyperreflexia of lower extremities • Positive Babinski bilaterally	Laminectomies	N	Mature and immature teratoma	Recovery of motor function at follow-up
Acar 2003 [13]	7 m	F	T6–T8 Extradural	• Flaccid paraparesis • Loss of sensitive function	Laminectomy	Y	Mature teratoma	Gradual motor and sensitive improvements

Table 1 (continued)

Author	Age	Sex	Level and location	Preoperative symptoms	Approach	GTR	Diagnosis	Outcome
Kaneko 1999 [15]	34 d	M	C4–L4 Extradural Invasion to the foramina and extending to the mediastinal region	• Flaccid paraplegia • Dyspnea	Laminotomy and thoracotomy	N	Mature teratoma	Reintervention Spinal deformity Pigeon chest deformity due to intercostal nerve lesion Neurogenic bladder No recovery of neurological motor function
Murovic 1986 [16]	3 d	M	T11 Extradural with invasion to the retroperitoneal compartment	• Palpable abdominal mass • Flaccid paralysis in lower extremities • Decreased sensation in lower extremities	Laminectomy and laparotomy	Y	Mature teratoma	Tumor recurrence to the lumbosacral region with reintervention needed Outcome not specified
Choi (2004) [14]	8 m	M	T2–T4 Extradural	• Progressive paraparesis	Hemilaminectomy	Y	Mature teratoma	Motor improvement at 3-month follow-up
Senayali 2004 [30]	1 d	M	Thoracic (level not specified) Intramedullary	• Mass in the back • Exstrophic bladder	Not specified	Y	Mature teratoma	Paraplegia 2 days post operatively Gradual recovery with intact motor function at 8-month follow-up CSF leakage Hydrocephalus needing shunt
Gressot 2014 [31]	5 d	F	C6–T2 Intradural Associated with Chiari malformation type 2 and myelocystocele Detected in utero	• Mass in the cervicothoracic junction • Peter´s anomaly • Motor and sensitive function normal	Repair of myelocystocele and resection of the mass	Y	Mature teratoma	Asymptomatic in the 3-month follow-up
Mpayo 2014 [19]	7 yrs	M	T9-T11 Intradural Extramedullary	• Presented as recurrent aseptic meningitis due to rupture of spinal teratoma	Endoscopic resection	Y	Mature teratoma	Recovery with no further episodes
Rainey 2018 [32]	1 m	F	C2–T5 Intramedullary	• Irritability • Poor feeding • Somnolence • Absent Moro reflex • Decreased bilateral grasp reflex	NS	NS	Immature teratoma	Chemotherapy Gross developmental delays Neurological function is slowly improving
Sancak 2001[33]	2 d	F	T4–T5 Extradural Associated with Taussig-Bing malformation	• Cyanosis • Respiratory distress • Mass on back	Laminectomy	Y	Mature teratoma	No neurological symptoms

NS not specified, *Y* yes, *N* no, *M* male, *F* female

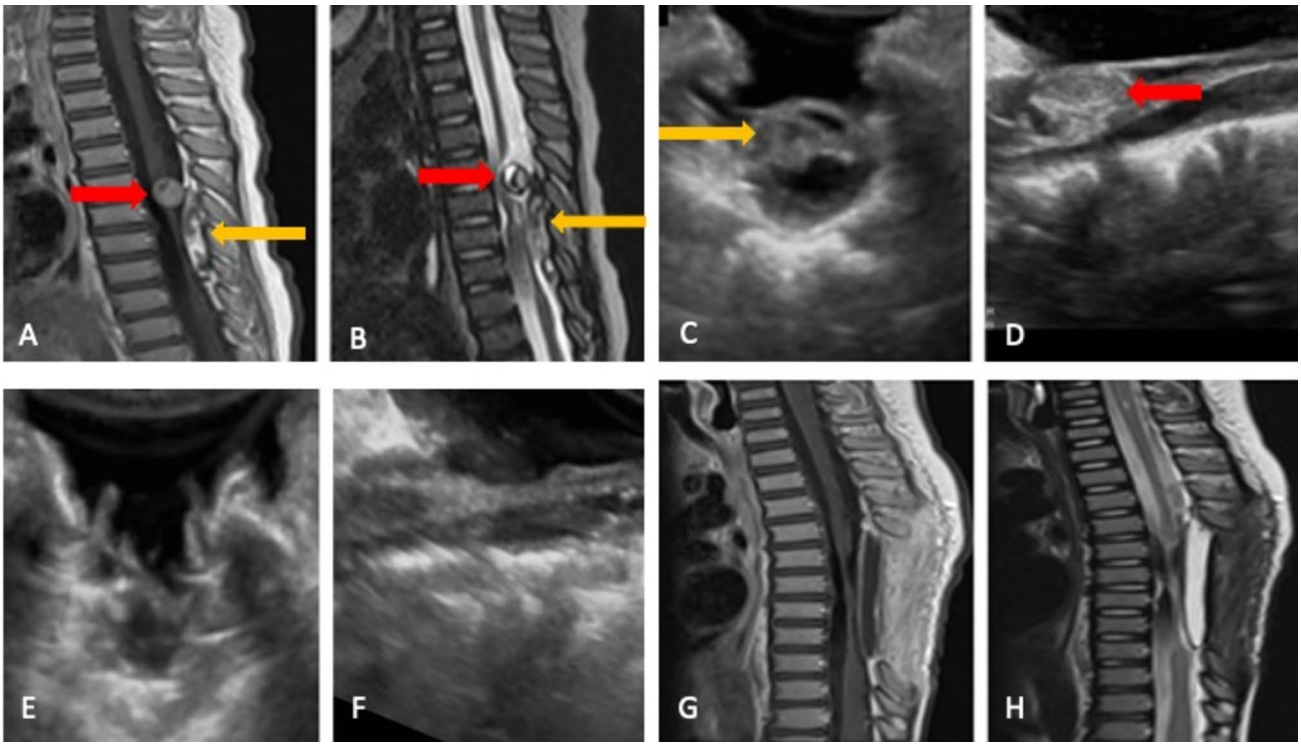

Fig. 1 Preoperative (**A**) T1-weighted and (**B**) T2- weighted images of the thoracic spine with evidence of the extradural (yellow arrow) and subdural (red arrow) lesions. Intraoperative ultrasonography depicting (**C**) extradural lesion in axial plane and (**D**) intradural lesion in sagittal plane. (**E**) Axial and (**F**) Sagittal planes with evidence of GTR of both lesions through intraoperative ultrasonography. (**G**) T1 and (**H**) T2-weighted postoperative images showing GTR of both lesions

decreased sensation and flaccid paraplegia besides urinary symptoms. Magnetic resonance imaging (MRI) showed two medullary lesions in the thoracic region from T5–T8 with heterogeneous enhancement after administration of intravenous (IV) gadolinium (Fig. 1) and were described as two independent lesions. The first mass was described as an extradural fusiform lesion with fatty tissue components (Fig. 1). The intradural lesion appeared as an intradural extramedullary lobulated neoplasm with spinal cord, foraminal and dentate ligament compression. The lesion showed hyperintense regions in T1 sequence and heterogeneous characterization in T2 with no suppression in fat saturation imaging (Fig. 1).

A laminectomy was performed for the T4–T7 vertebrae revealing an extradural lesion composed of sebaceous material, hair and dental organs extending from T4 to T7 enclosed in a fibrous capsule (Fig. 2). Subsequent durotomy

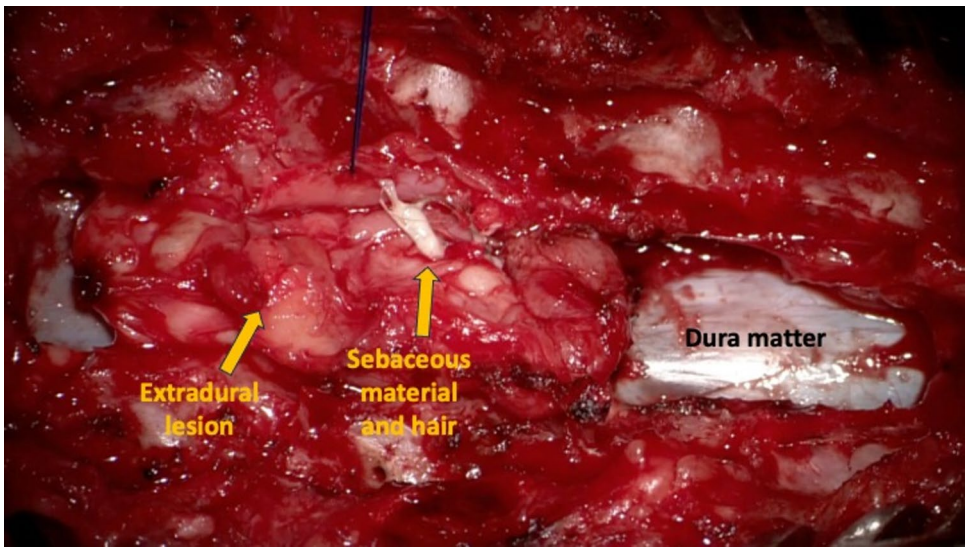

Fig. 2 Extradural lesion comprised of sebaceous material and hair

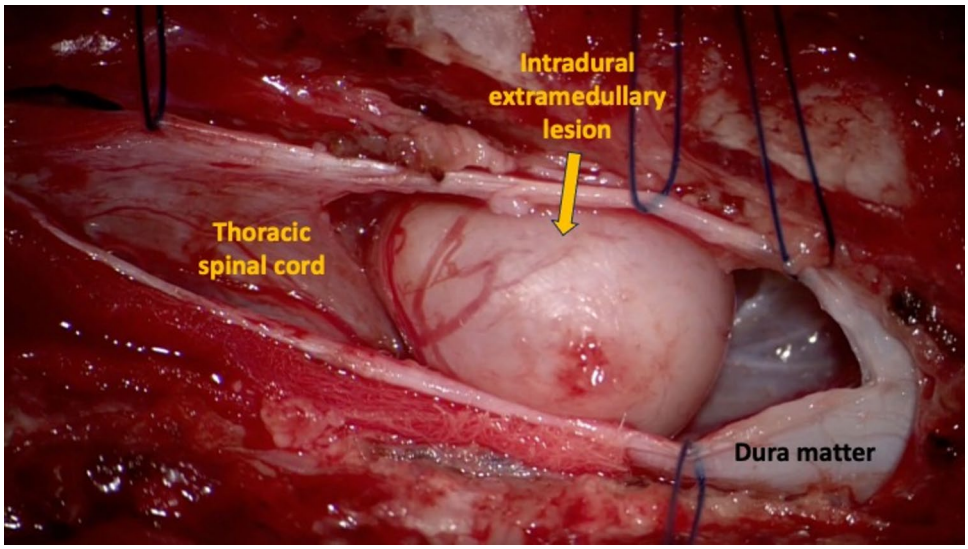

Fig. 3 Round capsulated intradural extramedullary lesion with compression and displacement of spinal cord

exposed a round sebaceous lesion of 10 mm diameter with compression of the spinal cord (Fig. 3).

Gross total resection (GTR) was achieved for both lesions with the aid of intraoperative ultrasonography (Fig. 1). Histopathological analysis of the tissue revealed both lesions to be comprised of mature elements from the three germ cell layers. None of the lesions showed features of malignancy. Final diagnosis was established as mature teratoma for both the extradural and intradural tumors. During hospital stay, the patient remained with no changes and was discharged 4 days later without complications. In the 1-year-follow-up, he remained with no motor nor sensitive function in the lower extremities; however, spasticity and hyperreflexia were noted. Recovery of neurological function in the lower extremities is yet to be determined in a longer-term follow-up.

Discussion

Spinal tumors account for 5.1% of all brain and other CNS neoplasms in the 0–19 years age group [5, 6]. Teratomas are rare neoplasms in the pediatric population and can arise in any location near the midline; still, patients in infancy or early childhood most often present these lesions in the sacrococcygeal region and only a few reported in the thoracic spine [7, 8]. Computed tomography (CT) can identify calcifications, fat and soft tissue but MRI remains as the gold standard imaging tool allowing better assessment of their morphology [9]. Teratomas need to go under histological differentiation to be classified in mature or immature [4], with the great majority of lesions exhibiting benign characteristics with 1 to 2% of these tumors undergoing malignant transformation [10]. The age of presentation is diverse and an early onset of the symptoms may be due to a congenital origin which would strictly imply its presence since gestation; however, its definition remains arbitrary and its use has been variable, utilizing it for lesions between 28 days to 12 months after birth [11].

We performed an analysis with available cases in the literature comparing age, location, degree of invasion, clinical manifestation, course of treatment and prognosis (Table 1).

Only one article was excluded due to incomplete clinical information. Children with thoracic teratomas presented with motor or sensitive neurological deficits that can improve completely or partially after surgical resection [9, 12–18]. Motor symptoms in infants were found to manifest as underdevelopment of motor milestones. In contrast, older children referred pain, numbness, and gait disturbances. Only one case was reported to present as recurrent aseptic meningitis due to rupture of the lesion [19]. Different variables influence the prognosis and presentation in each patient, such as age at onset of symptoms, size of the lesion, location and extra-spinal invasion of the lesion and concomitant malformations which may be directly related to spinal teratomas, but independent causality cannot be excluded (Table 1) [15, 16, 18, 20–25]. To the best of our knowledge, their relationship with teratomas is yet undetermined and we consider that further investigations with an accurate statistical methodology are still impeded due to the low number of cases reported to achieve statistical power and great heterogeneity between these. As in adults, functional prognosis in children is relatively good following surgery regardless of intramedullary, extradural or intradural location [20]. Preoperative assessment of invasion to nearby structures and degree of compression to neurovascular structures with MRI could help predict motor and sensitive outcomes in patients. Surgery is still the mainstay treatment for

teratomas as histological typification is necessary. GTR is encouraged to seek remission and clinical recovery. Remaining lesions have a risk of relapse going up to 18% [22] and chemotherapy or radiotherapy should be reserved for tumors exhibiting immature or malignant components [3].

The etiology of spinal teratomas is still under discussion by different authors and no established cause is available. According to the notion of the misplaced germ cell theory, the improper location of multipotent germ cells during the neural tube migration may prompt the development of spinal teratoma; however, a second hypothesis establishes a dysembryonic causality for its origin [12, 23, 25]. Midline position, occasional intramedullary location and determination of the sex chromatin of these lesions make the misplaced germ cell theory the most likely etiology but certain limitations, such as the number of cases available for genomic studies, make it extremely difficult to determine [21, 24]. Our case is the first report of simultaneous teratomatous lesions in the spine of a pediatric patient in both the intradural and extradural compartment. This presentation makes us consider flawed migration of germ cells more likely to give origin to this lesion due to the presence of both lesions at the same level of the spine with structural independency between them giving an anatomical explanation rather than pure chance of pluripotential cell rests at the same level of the spine as a dysembryonic origin would require [25]. Dysembryonic theory is more supported by the presence of dysraphism and lesions in proximity with structures originating from the caudal cell mass [12]. Upper lesions without dysraphisms, as in this case, do not match with the associated characteristics and are better supported by flawed migration of germ cells.

We acknowledge that the discussion of a single case with these characteristics does not confirm nor reject any theory for its etiology, but we consider that it can open new lines of discussion for its resolution. Limitations to this study include the interpretation of possible outcomes with a limited pool of patients as well as the lack of a statistical analysis. Further reports in the field may contribute to a more accurate evaluation of the data and the elaboration of more robust reviews. Multicentric collaborations of possible nonreported cases could help to pool a greater number of patients and analyze the characteristics of these lesions to explore the relationship between spinal teratomas and the cell migration theory, or in the other case, explore alternative mechanisms.

Conclusion

Spinal teratomas are rare neoplasms in children with high heterogeneity in its location and extension. The lack of a proper statistical analysis precludes the objective analysis of outcomes and prognosis among patients. Surgical treatment should be performed as it can provide improvement and allow histological analysis for its typification. This is the first reported case of an extradural and intradural teratoma in a pediatric patient. The simultaneous presentation of two lesions in the extradural and intradural compartment with no relationship or connection between them can help to elucidate and further analyze the theory of its etiology.

Author contributions Omar R. Ortega-Ruiz wrote the original draft as well as edition of the manuscript. Performed the research and analysis of the data. Carlos D. Acevedo-Castillo: writing of the original draft. Luis Alejandro Pérez-Ruano edited and reviewed the manuscript and the preparation of the figures shown in the manuscript. Enrique Caro-Osorio edited and reviewed the manuscript and should be recognized for the administration of the project.

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Data Availability Data is provided within the manuscript or supplementary information files

Declarations

Conflict of interest The authors declare no financial nor any non-financial competing interests.

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