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Case Report

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Radiographic and clinical features of thoracic intradural extramedullary epidermoid tumor: Illustrative case

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Abbreviations: CSF: Cerebrospinal fluid; CT: Computed tomography; DWI: Diffusion-weighted imaging; MRI: Magnetic resonance imaging.

Abstract

Background: Spinal epidermoid tumors are uncommon benign neoplastic lesions arising from heterotopic epidermal rest-cells. These lesions rarely occur in the thoracic region (<0.8%) and can present with symptoms of spinal cord and nerve root compression.

Case presentation: A 76-year-old female presented with a 2-year history of mid-thoracic back pain with radiculopathy to bilateral lower extremities refractory to conservative management. Magnetic resonance imaging showed an intradural extramedullary lesion. Repeat imaging (9 months later) showed increase in the size of the mass with imaging findings (iso-/hypointense T1, hyperintense T2 and increased signal on diffusion-weighted imaging) highly suggestive of epidermoid tumor. Patient underwent laminectomy and en bloc excision of the mass with histological findings later confirming the diagnosis. She was discharged home in a stable neurological condition and later made complete recovery during follow-up.

Conclusions: Epidermoid tumors should be considered in the differential diagnosis of intradural extramedullary lesions of the thoracic spine. Clinical course, presentation, and diagnosis can be delayed due to the slow growth pattern of these tumors. Contrasted magnetic resonance imaging is key for diagnosis and complete excision should be the goal of treatment. Good neurological outcomes are generally achieved, and serial radiological surveillance is recommended as these lesions can rarely recur.

Introduction

Spinal epidermoid tumors are uncommon benign neoplastic lesions (<1% of all intradural spinal tumors). That arise from heterotopic epidermal cells in the spinal canal [1]. These lesions can be congenital or acquired. Congenital lesions are often caused by premature dysjunction and entrapment of ectodermal cells during neural tube closure around the third to fifth weeks of development [2]. These cases tend to occur in the pediatric age group and are often associated with conditions such as spinal dysraphism, intradural lipomas and/or dermal sinus tract lesions [3]. On the other hand, acquired epidermoid tumors arise due to the implantation of cutaneous epithelial tissue into the intraspinal compartment after spinal trauma, lumbar puncture procedure, or following spinal surgery [4]. Often referred to as "pearly tumors" due to their smooth, glistening, white capsule [5], spinal epidermoids are most commonly intradural and extramedullary in their location. These lesions occur most commonly in the lumbosacral region and rarely involve the thoracic or cervical spine [6]. Ensuing symptomatology depends on the level of the lesion and extent of spinal cord and nerve root compression. The goal of treatment is safe, complete resection of the tumor and restoration of normal neurological function. Here we report a case of a thoracic intradural-extramedullary epidermoid tumor presenting with thoracic radiculopathy that was successfully managed with surgical excision.

Case presentation

Patient is a 76-year-old female former smoker (80 packs per year history) with a past medical history significant for bladder cancer, chronic kidney disease (stage III), myocardial infarction, morbid obesity, and osteoarthritis of both feet. She presented to the outpatient clinic with a 2-year history of mid-thoracic back pain with radiculopathy extending into the bilateral lower extremities. She noted that her pain was worse in her right lower extremity and was associated with intermittent non-dermatomal paresthesia worse on the right side. She had been using a cane or walker for approximately two years.

Following a trial of conservative management in the pain clinic and outpatient physical therapy, she was further worked up with magnetic resonance imaging (MRI) of the thoracic and lumbar spine. The MRI thoracic spine result was notable for a T2 hyperintense intradural extramedullary lesion measuring 7x5x8 mm in size eccentric to the left side at T10-11 spinal level (Figure 1). The patient was then scheduled to undergo further testing with MRI thoracic spine with gadolinium contrast to further characterize the lesion but was later lost to follow-up prior to acquiring the imaging workup. She later presented to the clinic nine months after the initial scan with worsening back pain symptoms and MRI thoracic spine with gadolinium contrast was obtained. Results were notable for a T1 iso-/hypointense, T2 hyperintense, non-enhancing lesion at T10-11 but with interval increase in size (15x5x9 mm), compared to prior MRI imaging. It had an increased signal on the diffusion-weighted imaging (DWI), sequence with a concurrent decrease in the apparent diffusion coefficient (ADC) values. The lesion was abutting the spinal cord at that level with moderate compression (Figure 2). The radiological differential diagnosis included a dermoid/epidermoid inclusion cyst or synovial cyst. Due to the lack of enhancement, meningioma or nerve sheath tumors were considered less likely. Given the imaging findings and persistent pain, the patient was counseled on the risks and benefits of alternative modes of management and opted to proceed with surgery to excise the lesion.

Following preoperative optimization, she underwent T10-11 osteoplastic laminectomy and en bloc excision of the underlying lesion. Under intraoperative microscope visualization, the lesion was identified following careful dissection of the arachnoid membrane. It was noted to be a light yellow well-demarcated pearly lesion surrounded by the nerve roots and was noted to be caudal to the conus medullaris (Figure 3). The mass had no dural or nerve root attachments. After careful dissection, it was freed from the overlying nerve roots and intradural venous vasculature and subsequently removed en bloc. Following satisfactory hemostasis, watertight dural closure was performed. Final surgical pathology confirmed the diagnosis of an epidermoid cyst. She was later discharged to home in stable condition. During her 6-week follow-up, she noted that her mid-thoracic back and radicular leg pain had improved, and MRI imaging confirmed complete excision of the lesion (Figure 4). She was able to ambulate independently and denied difficulties with balance or coordination. Informed consent to participate in this study was obtained.

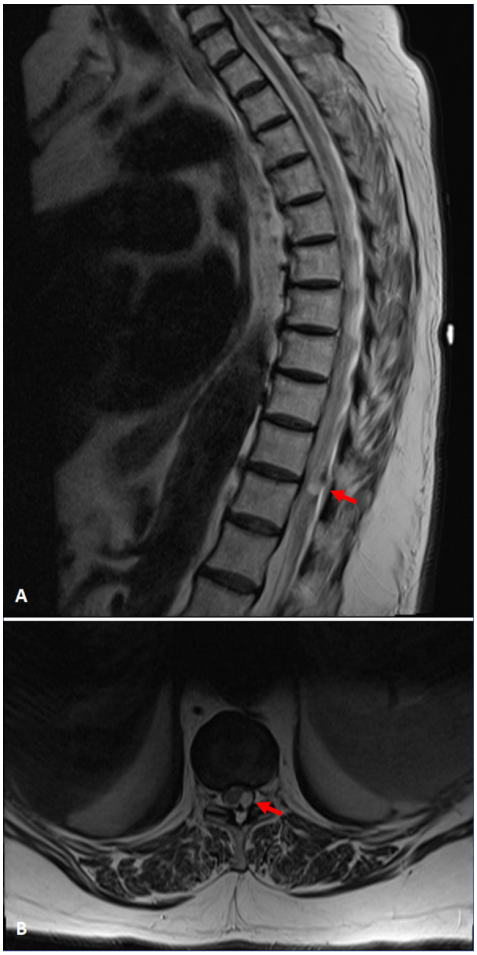

Figure 1: (A-B) Initial MRI. **(A)** Sagittal and **(B)** Axial T2-weighted MRI sequence demonstrating the intradural extramedullary lesion (red arrows) at the level of T10-11.

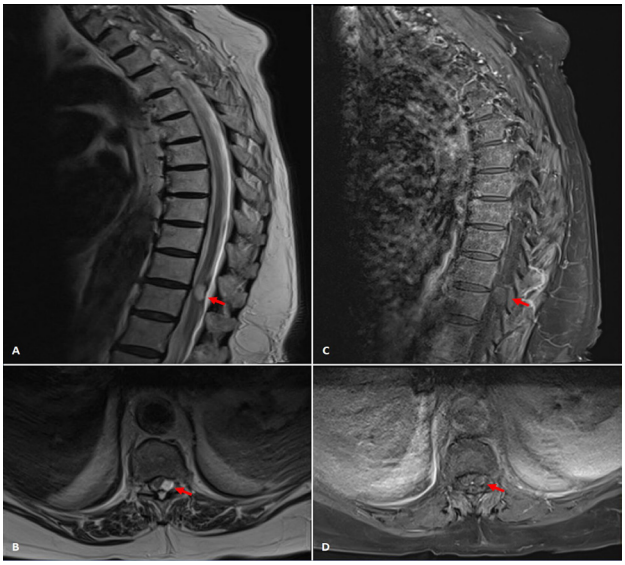

Figure 2: (A-D) Preoperative MRI. **(A)** Sagittal and **(B)** Axial T2-weighted MRI sequence demonstrating the interval growth of the intradural extramedullary lesion (red arrows) At the level of T10-11. **(C)** Sagittal and **(D)** Axial T1-weighted post gadolinium MRI sequence demonstrating lack of enhancement of the lesion (red arrows).

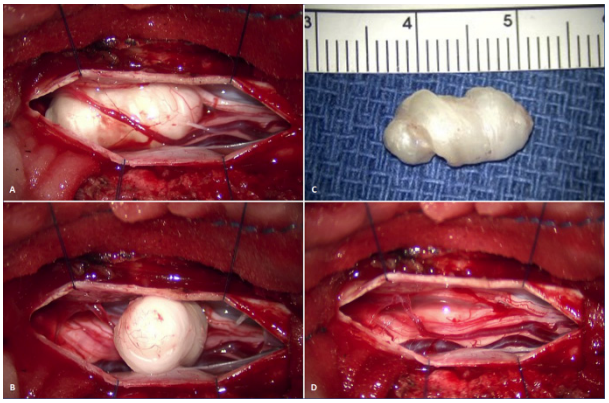

Figure 3: (A-D) Intraoperative gross appearance of the lesion. **(A)** After dural opening, the pearly mass was visualized. **(B)** The mass was not attached to the surrounding neural or vascular structures. **(C)** Gross morphology of the tumor after en bloc removal. **(D)** Final visualization of the surgical area showing complete tumor excision.

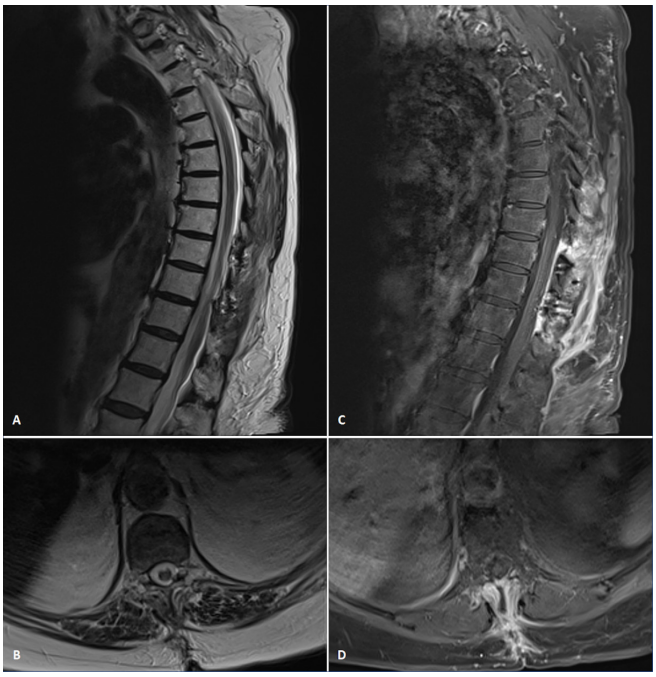

Figure 4: (A-D) Postoperative MRI. **(A)** Sagittal and **(B)** Axial T2-weighted MRI sequence confirming complete excision of the lesion. **(C)** Sagittal and **(D)** Axial T1-weighted post gadolinium MRI sequence confirming complete excision of the lesion.

Discussion and conclusions

Epidermoid tumors in the thoracic spinal cord are relatively the rare and account for <0.8% of all spinal tumors [4,7]. These lesions present more commonly in pediatric patients and can be caused by either a congenital or acquired etiology. Congenital epidermoid cysts are thought to arise from the abnormal inclusion of ectodermal rest cells due to premature dysjunction between the superficial and neural ectodermal layers between the third and fifth week of embryonic development [2]. This is confirmed by the association between spinal epidermoid tumors and developmental pathologies like spinal dysraphism, hemivertebra, diastematomyelia, and dermal sinus tract lesions. Congenital spinal epidermoids are often located along the midline, often dorsal to the cord and can be either intramedullary or extramedullary [8]. Conversely, acquired lesions develop due to traumatic or iatrogenic implantation of skin fragments into the intrathecal space following lumbar puncture or spinal surgery [4]. The tumors in these cases are usually extramedullary. Interestingly, since the advent of stylet lumbar puncture needles, the incidence of acquired epidermoid tumors has decreased though sporadic cases are still occasionally reported [9,10]. of note, in our patient there was no history of lumbar puncture or spinal surgery making it a sporadic spinal epidermoid tumor. Histological examination of these lesions shows stratified squamous epithelium, supported by an outer layer of keratin strands [11]. The progressive desquamation from the internal epithelial lining produces a soft white material giving these tumors their "pearly appearance" and accounts for their gradual but progressive increment in size [12]. These lesions often grow slowly, and clinical presentation is insidious and frequently delayed. The presenting symptoms are quite variable and vary according to the level and extent of spinal cord compression. Rarely, sudden onset or rapidly progressive symptoms have been reported in pediatric patients with congenital lesions [13]. The imaging modality of choice for spinal epidermoid tumors is MRI imaging of the spine with and without gadolinium contrast [1]. In cases with associated vertebral and/or spinal cord anomalies (mostly congenital cases), performing an additional CT scan or x-ray imaging can be valuable to evaluate the bony anatomy [14]. In general, epidermoids cause minimal displacement of the nerve roots and the spinal cord in the subdural space. They are often hypointense on T1-weighted images and hyperintense on T2 [15], though atypical signal intensity changes have been reported with hyperintense signals on T1 and hypointense signals on T2 [16]. In our patient, the pattern of signal intensity on MRI was characteristic with hypointense T1 and hyperintense T2 signals noted. Peripheral post gadolinium enhancement with well-defined borders has been reported, though in general these lesions are non-enhancing on contrasted scans [17]. Use of DWI sequences is key to differentiate epidermoid tumors from arachnoid cysts [18]. Epidermoids tumors show evidence of restricted diffusion that appears bright on DWI while arachnoid cysts appear dark on DWI with a signal intensity comparable to cerebrospinal fluid.

The mainstay of treatment for epidermoid tumors is gross total excision with removal of the capsule in order to minimize the risk of recurrence [6]. However, gross total excision may not be feasible in certain cases where the mass adheres to surrounding neural structures. In these cases, subtotal excision of the lesion can result in local recurrence with rates as high as 10% reported in the literature [19]. En bloc resection of the lesion is recommended when possible to avoid contamination of the surgical field or spillage of the tumor contents into subarachnoid space. Release of intralesional contents during piecemeal resection can result in prolonged aseptic chemical meningitis and subsequent arachnoiditis [15]. To reduce the risk for chemical meningitis, Mishra et al. recommend irrigation of proximal and distal arachnoid spaces with normal saline prior to dural closure [20]. In this case, complete en bloc excision could be performed with no signs or symptoms of postoperative meningitis noted. Follow-up imaging confirmed complete excision of the lesion and serial radiological surveillance is recommended for detection of recurrence. Epidermoid tumors must be considered in the differential diagnosis of intradural intra/extramedullary lesions of the thoracic spine. The clinical course, presentation, and diagnosis is often delayed due to the insidious growth pattern of these tumors. MRI imaging with and without contrast with DWI sequences is the imaging modality of choice and early surgical intervention targeting gross total excision is recommended. Given risk for recurrence even in the setting of en bloc resection of these lesions, serial radiological surveillance is paramount for

early detection of recurrence.

Declarations

Acknowledgments: None.

Conflicts of interest: The authors declare that they have no competing interests.

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