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Conus medullaris dermoid with secondary rupture into the central canal: A case report and imaging findings

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Abstract

Spinal dermoid cysts are benign, congenital tumors that typically form along the spinal cord. These cysts are characterized by their cystic structure, often being either unilocular (one cavity) or multilocular (multiple cavities). The cysts are lined with squamous epithelium, and they contain various skin appendages, such as hair follicles, sweat glands, and sebaceous glands. These features are indicative of the cyst's origin from ectodermal tissue during embryonic development. The tumors can be classified based on their location within the spine as Intramedullary dermoid cysts that occurs within the spinal cord which are rarer and can cause more direct compression on the spinal cord and surrounding neural structures and Extramedullary dermoid cysts that are located outside the spinal cord but within the spinal canal. They typically cause less severe direct compression but can still lead to symptoms through mechanical pressure on adjacent nerves or tissues. A ruptured spinal dermoid cyst is a rare but serious condition that occurs when the cyst breaks open, releasing its contents (such as keratin, hair, and sebaceous material) into the surrounding spinal structures. This can lead to a variety of complications, as the contents of the cyst can cause inflammatory reactions, chemical meningitis, or even spinal cord damage.

Keywords: Conus medullaris; Dermoid; MRI; Spinal.

Introduction

Dermoid cysts were first identified in the 18th century by Serratus and later termed "pearly tumors" by Cuvelier. Spinal dermoid cysts are rare, comprising less than 1% of all spinal cord tumors [1]. They are uncommon, slow-growing, and benign lesions that can be found in intramedullary, intradural-extramedullary, and extradural locations [2]. The lumbosacral spine is most commonly affected with a spinal dermoid cyst; cervical and upper thoracic regions are the least commonly affected segments. These cysts can also be associated with various spinal abnormalities, including split cord malformation, where the spinal cord is divided into two segments; tethered cord, a condition where the spinal cord is abnormally attached and limited in movement and myelomeningocele. Additionally, they can also be associated with bony malformation, dermal sinus tract or hypertrichosis. These associations complicate the clinical presentation of spinal dermoid cysts, requiring careful assessment and management to address both the cyst and any coexisting conditions. These cysts develop from ectopic ectodermal remnants during embryonic neural tube closure, resulting in the formation of cystic structures [3]. In rare cases, ab-

normal dermal seeding can occur as a result of spinal surgery, lumbar puncture, or trauma, which may lead to the development of spinal dermoid cysts. Approximately 20% of spinal dermoid cyst cases are associated with other conditions such as dermal sinus, spinal dysraphism, and a low-set tethered cord, which may contribute to the complexity of their presentation and treatment. Spinal dermoid cysts are often asymptomatic and detected incidentally. However, when symptomatic, they may present with motor disturbances, pain, sensory deficits, or bowel and bladder dysfunction [4]. If a rupture occurs, they can become acutely symptomatic [5]. Although benign and slow growing, dermoid cysts carry a risk of rupture which is associated with a high morbidity and mortality. A dermoid cyst can rupture during surgery, after a trauma or spontaneously. Early diagnosis through key imaging techniques such as MRI and surgical excision is the treatment of choice for ruptured spinal dermoid cysts. MRI allows for the accurate identification of the cyst's location, rupture, and potential complications, such as spinal cord compression or chemical meningitis. Early intervention being key to a favorable prognosis. Delayed diagnosis or treatment can increase the risk of permanent neurological damage, highlighting the importance of prompt medical attention.

Case report

We present the case of a 21-year-old patient diagnosed with an intraspinal dermoid cyst, a rare congenital lesion resulting from ectodermal inclusion during neural tube closure. The patient presented with progressive lower back pain, radiating lower limb weakness, and sensory disturbances over several months. MRI of the spine revealed a well-defined, non-enhancing, heterogeneous lesion with fat suppression characteristics, suggestive of a dermoid cyst. Surgical excision was performed, and intraoperative findings confirmed a cystic mass containing hair, sebaceous material, and keratinous debris. Postoperative recovery was uneventful, with significant improvement in neurological symptoms. This case highlights the importance of early imaging and surgical intervention in symptomatic intraspinal dermoid cysts to prevent irreversible neurological deficits. Two intramedullary lesions are observed along the conus medullaris, characterized by a predominant cystic component with a small fat signal intensity component. No significant post-contrast enhancement is noted. The first lesion is located at L1–L2 level, measures 1.3 × 1.7 × 3.9 cm (AP × TR × CC). The second lesion is at the L3 vertebral level, measures 1.9 × 1.3 × 1.8 cm (AP × TR × CC). Both lesions nearly completely occupy the spinal canal, leading to severe central canal stenosis, displacement of the thecal sac, and compression of the nerve roots. Additionally, a few fat droplets are seen within the central canal at D9, D10, and D11 levels. Mild posterior cortical scalloping of the D12 vertebral body is also noted.

Discussion

Dermoid tumors account for approximately 1.1% of all primary intraspinal tumors. They originate from the ectopic inclusion of embryonic ectodermal remnants within the spinal canal during neural tube closure in embryonic development. The human spinal cord develops through primary and secondary neurulation. During primary neurulation, the cutaneous and neuroectoderm separate following neural tube fusion. Once primary neurulation is complete, the caudal neuropore closes, forming

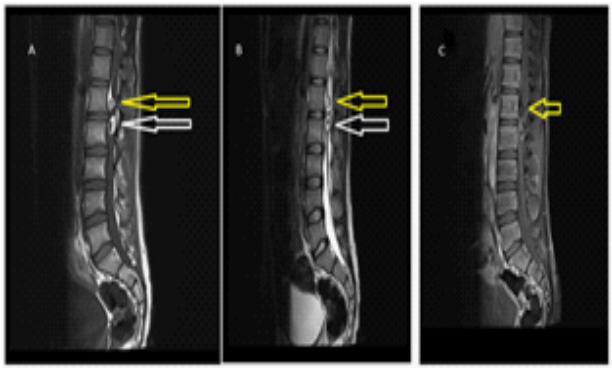

Figure 1: Sagittal T1, T2 and T1 fat supressed MRI images **(A,B,C)** of lumbar spine showing two conus medullaris dermoid cysts.

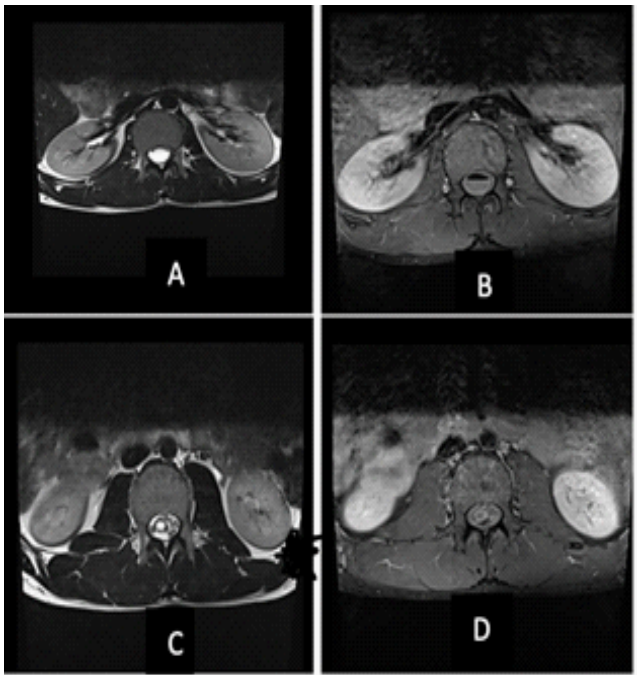

Figure 2: Axial T2 pre-contrast, T2 post-contrast at L1-L2 vertebral level **(A,B)** and axial T2 pre-contrast, T2 post-contrast at L3 vertebral level **(C,D)** showing spinal dermoid cyst with fluid-fluid level.

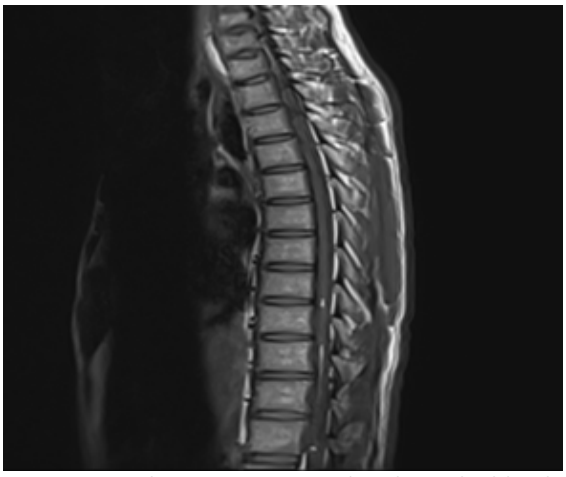

Figure 3: Few T1 hyperintensities at dorsal vertebral levels – fat droplets (indicating rupture).

the caudal cell mass distal to the neural tube. In secondary neurulation, the caudal cell mass undergoes cavitation and subsequently connects to the central canal of the neural tube at the junctional region, forming the secondary neural tube [6]. Dermoid tumors primarily develop in the lumbosacral region, accounting for nearly 60% of cases, where they commonly affect the cauda equina and conus medullaris. Their occurrence in the upper thoracic spine is significantly less frequent, comprising only about 10% of cases, while their presence in the cervical region is even rarer, with an incidence of approximately 5%. These tumors tend to be congenital in origin, arising from ectodermal remnants that become trapped during neural tube closure. Due to their slow-growing nature, symptoms may not appear until the lesion reaches a size large enough to exert pressure on surrounding neural structures, leading to neurological deficits such as radiculopathy, gait disturbances, or sensory impairments. Advanced imaging techniques, particularly MRI, are crucial for precise localization and characterization of these lesions, aiding in surgical planning and management.

Conclusion

A ruptured spinal dermoid cyst is a medical emergency that requires immediate attention. While these cysts are typically benign and asymptomatic in their early stages, a rupture can lead to significant neurological and inflammatory complications. Early detection, often through MRI, and prompt surgical intervention are critical to preventing long-term damage and improving patient outcomes.

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