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CASE REPORT

A Rare Case of Intramedullary Dermoid with Atypical Presentation

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ABSTRACT

Background: Intraspinal dermoid cysts are rare and benign tumors that occur primarily due to the defective closure of the neural tube, an ectodermal derivative, during the process of development. They can appear subdurally, extramedullary or intramedullary, with intramedullary presentations being relatively rare.

Case presentation: We present here a 24-year-old lady with low backache radiating to both lower limbs, with tingling and numbness of both lower limbs for 3 years. MRI study of the lumbosacral spine revealed a heterogeneous intradural mass lesion in the conus medullaris region at L2-L3 level (low lying cord) displacing the traversing nerve roots. Total excision of the tumor was tone and sent for biopsy which revealed keratinocytes with keratin flakes,based on which a diagnosis of dermoid was made.

Conclusion: The intramedullary location of a primary dermoid cyst an adult is rare and thus makes this case a very unique and rare entity Keywords : dermoid cysts, intramedullary lesions, spinal cord tumours.

Keywords: Dermoid cysts, Intramedullary cysts, Spinal cord tumour.

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INTRODUCTION

First described in 1745 by Verrattus, dermoid tumors uncommonly occur in the spine, especially in an intramedullary location.[1] They are rare, benign, slow-growing tumors arising from more than one of three primitive germ cell layers.[2] The prevalence of these lesions according to recent studies is about 0.3%.[3] Presented here is a case of intramedullary dermoid at the conus medullaris level in a 24-year-old female with magnetic resonance imaging (MRI) features and histopathological findings and relevant scientific review of the literature.

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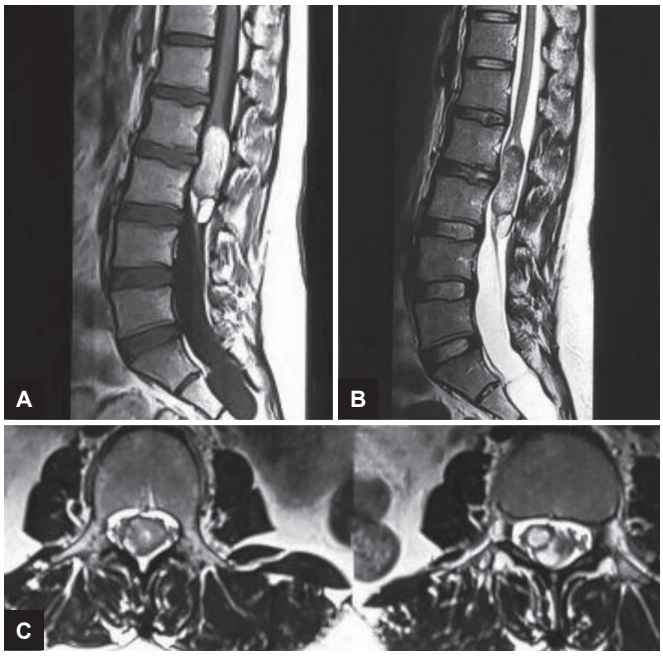
CASE REPORT

A 24-year-old lady presented to our hospital with a low backache radiating to both lower limbs, with tingling and numbness of both lower limbs for 3 years. There was no history of urinary incontinence, infection, lumbar puncture, spinal trauma, or previous spinal surgery.

On clinical examination, the skin over the neck and back was normal with no evidence of any sinus, hairy patch, or any cutaneous mark. Power in right lower limb was 4/5 and left was 5/5. Reflexes were normal. Sensory examination revealed hypoesthesia with diminished pinprick and touch sensation over L4-L5 dermatome. Plain radiograph of lumbosacral spine was normal.

Magnetic resonance imaging study of the lumbosacral spine revealed a heterogeneous intradural mass lesion in the conus medullaris region at L2-L3 level (low-lying cord) displacing the traversing nerve roots measuring 56 (craniocaudal) × 14 (anteroposterior) × 22 (mediolateral) mm (Figs 1A to C). It was heterogeneously hyperintense on T1- and T2-weighted images. On fat-suppressed



Figs 1A to C: Magnetic resonance imaging of lumbosacral spine depicting the intramedullary dermoid

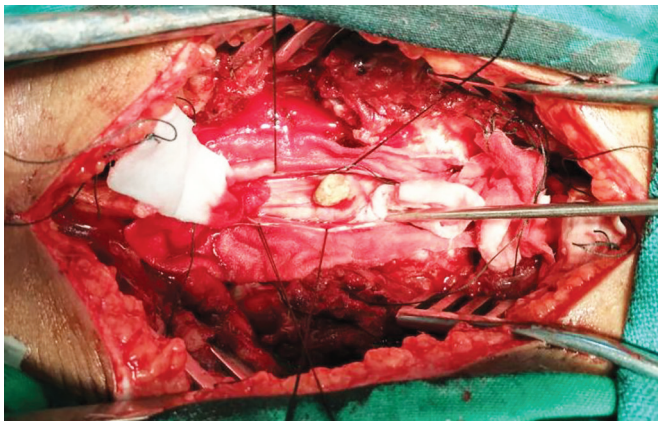


Fig. 2: Intraoperative view of the dermoid following myelotomy

images, the caudal component of the lesion showed a signal drop. The cranial part showed restricted diffusion with no postcontrast enhancement.

Treatment

The patient was managed surgically. In the prone position, under all aseptic and antiseptic conditions, a vertical midline incision was made from L2 to L4 level.

Following subperiosteal dissection, L2, L3, and L4 spinous processes and laminae were exposed. L2 and L3 laminectomy was done. Bulge and stretching of the dura were noted. Dura was opened and a swelling was noted over the underlying, low-lying cord. A midline dorsal myelotomy was done. Dirty white, flaky, sticky, and avascular tumor was visualized (Fig. 2). Removal of the entire tumor followed by adequate irrigation was done and the specimen sent for histopathological evaluation (Fig. 3).

Histopathologic evaluation revealed aggregates of keratinocytes with keratin flakes with degeneration, which confirmed the diagnosis of the dermoid cyst (Fig. 4).

DISCUSSION

Spinal dermoids account for 0.8 to 1.1% of primary spinal tumors,[1,4,5] with the majority occurring in the extramedullary or subdural juxtamedullary lumbosacral region, usually in the conus or cauda equina.[4] Various hypotheses have been advanced to explain the pathogenesis of spinal dermoids. Van Gilder and Schwartz produced dermoids experimentally in young rats by implantation of skin fragments.[6] This "traumatic theory" was supported by several authors (Choremis et al, Hetzel and Kloss) where repeated lumbar punctures were sometimes responsible for these tumors, especially in children who have undergone numerous lumbar punctures for the treatment of tuberculous meningitis.[5] According to the "developmental theory," desmoids arise from the cell rests that result from defective closure of the neural tube between the 3rd and 4th week of embryonic life.[6-8] Many of the intraspinal dermoid cysts that arise spontaneously are seen in association with other congenital anomalies of the spinal cord, vertebrae, and the soft tissues overlying them dorsally.[5] No such history or finding was noted in our patient.

These cysts are often diagnosed when presenting clinically as neurologic manifestations, ranging from paresthesias to paralysis and sphincter complications.[9] Our case presented with a low backache radiating to both lower limbs, with tingling and numbness. The case series of epidermoid and dermoid tumors of the spinal cord presented by Bradford[10] evidenced most of the cases with urinary symptoms, either retention or incontinence. However, the patient in our case denied any such symptoms.

Magnetic resonance imaging represents the test of choice in demonstrating intramedullary lesions. The two major components of fluid and fat appear hyperintense on T1-weighted images, being most consistent with the similar findings mentioned by Do-Dai et al.[11] The areas

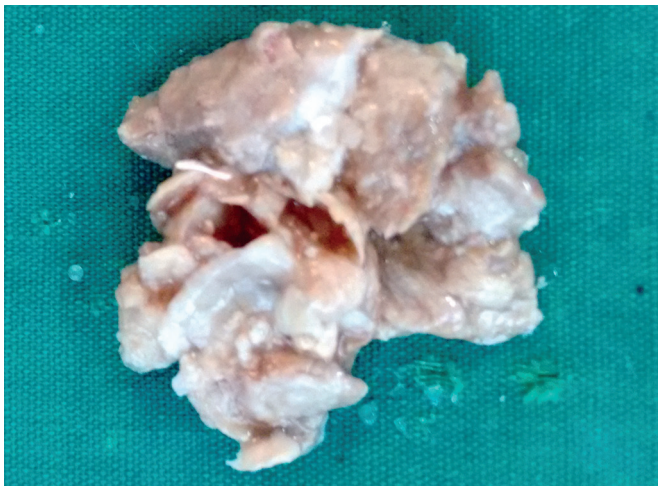


Fig. 3: Excised specimen of the dermoid

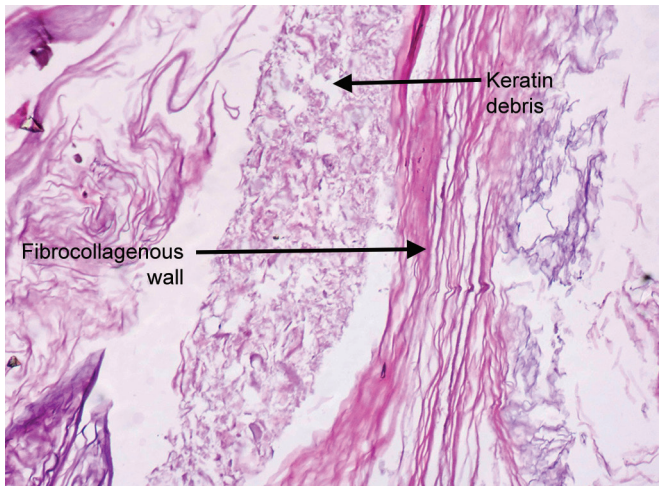


Fig. 4: Histopatholgical view of the dermoid

of hyperintensity are reflecting secretions of sebaceous glands and liquid lipid metabolites. Solid portions of the tumor are slightly hypointense to isointense compared with the spinal cord.

The dermoid cyst is usually filled with a soft, whitish yellow waxy substance with hairs and glandular secretions in addition to areas of induration.[1,5] This substance is produced by the secretion of sebaceous glands, progressive desquamation of the epithelium, and the transformation of keratin into fatty acids and cholesterol.[1,5] If the contents of the cyst are released into the subarachnoid space, a sterile chemical meningitis will result, presumably in response to the fatty acids present.[5,12] On microscopy, these tumors are usually surrounded by a multilayered, cornified epithelium with a collagen stroma lying in the dermis and hypodermis with cutaneous appendages, such as hair follicles, hair, sebaceous and sweat glands, as well as sebum.[1] The blood vessels were only detected in the connective tissue surrounding the tumor but never penetrating the epithelial wall of the dermoids.[1]

The treatment of spinal dermoids is usually surgical. Special care not to spill the cyst contents is taken while evacuating the cyst. A conservative approach is advised regarding the cyst wall.[1,5] Typically, the capsule is adherent to the adjacent neural tissue, and attempts at total resection are hazardous. Cyst recurrence that may result after incomplete removal occurs years after the initial procedure and does not justify the risk involved in attempting complete capsule removal.[1,6,12,13] Postoperative results have been variable in the literature, ranging from improvement in symptomatology to worsened postoperative status in certain cases. In a series of 16 cases of spinal dermoids and epidermoids reported by Lunardi et al,[4] most of the patients had improved, with two of them getting worse and three developing chemical meningitis.

CONCLUSION

Dermoid cysts are rare lesions in the spinal canal that tend to occur in the lumbosacral area. True intramedullary dermoids are even less common. Our case is unique as she did not present with the typical clinical features of an intramedullary dermoid. Appropriate diagnosis and careful meticulous excision of these tumors can produce the best outcomes.

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