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Case Report **Neurosurgery**

Spinal Intradural Epidermoid Cyst with Dermal Sinus in a 2 – Year Old Child: A Rare Case Report

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Abstract

Introduction: Spinal epidermoid accounts for < 1% of all primary cord tumor. This rare benign lesion may be congenital or acquired. Invagination of epidermal elements into the neural tube during embryonic period leads to the development of epidermoid cyst. ***Case Presentation***: A 2 year old girl presented with a spontaneous intergluteal swelling with dermal sinus and lower limbs weakness. The lumber MR demonstrated an intradural lesion from L3 to L5 levels that compressed cauda equine or nerve roots. MR findings were compatible with an epidermoid cyst with dermal sinus without any coexistent spinal dysraphism. The patient underwent microsurgical laminectomy from L3 to L5, tumor was resected and sent for histopathological examination. A pearly white tumor was found and histopathology revealed an epidermoid tumor. Post-operative Magnetic Resonance Imaging (MRI) finding was total removal. ***Conclusion*:** Diagnosis of spinal epidermoid cyst is often delayed due to its obscure presentation. Here, we presented spinal lumber intradural epidermoid cyst with dermal sinus which was treated successfully with microsurgical total resection with preservation of spinal stability and neurological function. Post-operative follow up with MRI is helpful.

Keywords: Spinal, Intradural, epidermoid Cyst, dermal sinus, Child.

INTRODUCTION

Congenital (e.g., associated with spinal dysraphism) or iatrogenic epidermoid cyst (i.e., also called "pearl tumors") represent approximately 0.5-1% of all intraspinal tumors [1, 2]. They arise from epidermal cells implanted intradurally. This implantation may be either congenital or iatrogenic [3]. Iatrogenic spinal epidermoid cysts following lumber puncture have been reported in the literature [4]. As it is a slow-growing tumor with nonspecific clinical and radiological findings the preoperative diagnosis is often difficult.

The clinical symptoms/signs of these lesions reflect their location. Here, we present a rare case of a 2 year old girl with a congenital lumbosacral intradural epidermoid cyst with dermal sinus.

CASE PRESENTATION

Our patient was a 2 year old girl. She presented 2 months ago with a spontaneous intergluteal swelling fistulized to the skin since birth with lower limb weakness since age of 9 moths. Her attendant denied any history of lumbar puncture, traumatism, or surgery to the spine. On neurologic examination, she had motor deficit of the lower limbs: Medical Research Council (MRC) grading 1/5. Ankle jerks were absent with decreased anal tone. Local examination finding revealed a swelling with dermal sinus discharging pus in the intergluteal region but no inflammatory signs or tuft of hair or sacral dimple.

MRI finding of the spine with and without contrast showed an intradural lesion extending from L3 to L5 levels that was isointense on T1-weighted images and hyperintense on T2- weighted images which had taken contrast. After clinical evaluation and radiological investigation, diagnosis was consistent with epidermoid cyst with dermal sinus resulting in displacement of cauda equina.

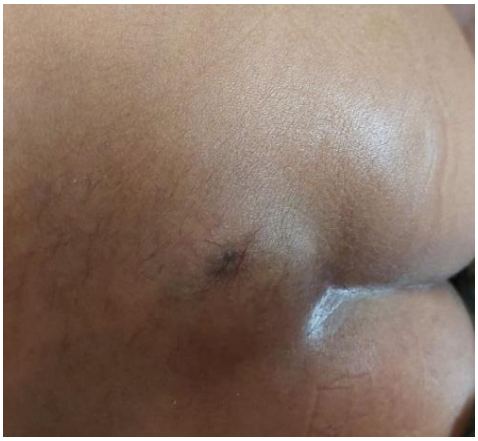

Figure 1: Epidermoid cyst with dermal sinus

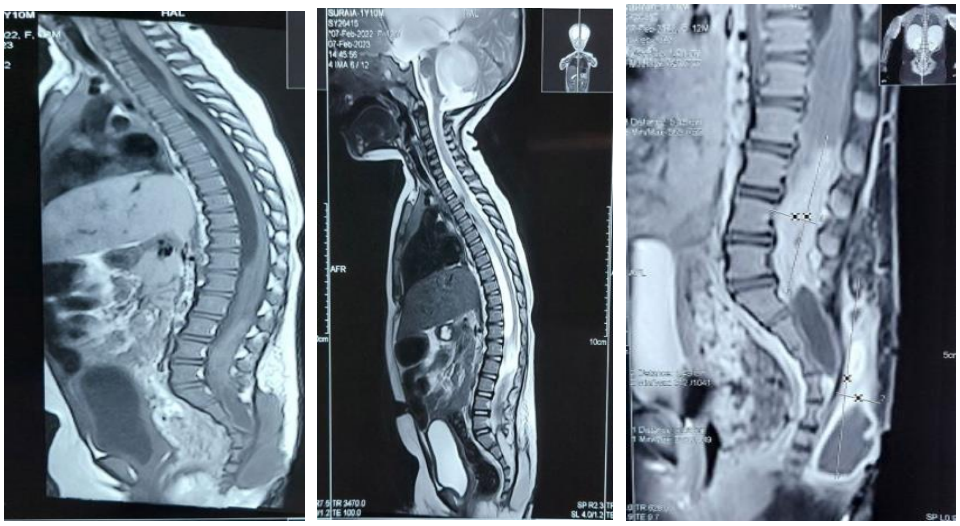

Figure 2: Noncontrast and contrast MRI of lumbosacral spine

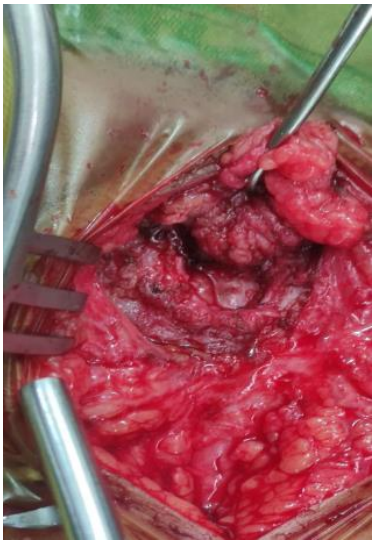

Figure 3: Per-operative picture

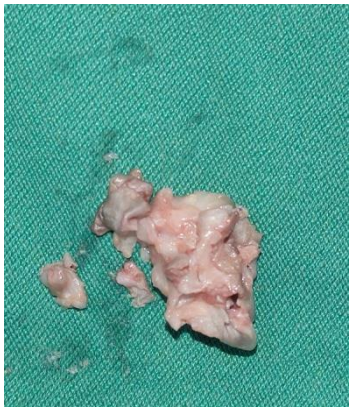

Figure 4: Epidermoid cyst

Surgery

She underwent total laminectomies from L3 to L5 for microscopic resection of epidermoid cyst with dermal sinus. After midline durotomy, an encapsulated "pearly white" tumor was found. Emptying of the cyst content and gross total removal of tumor was ensured. No spinal dysraphism was identified. An epidermoid cyst was confirmed by histopathological examination.

Immediate after operation her neurological deficit and sphincter tone improved. At 6 months follow-up period, her muscle power of lower limb was improved from 1/5 to 4/5.

DISCUSSION

Our patient was a rare case of epidermoid cyst with dermal sinus in non dysraphic spine. Congenital epidermoid cysts (ECs) are rare. Together with its slow growth and vague clinical menifestations, its diagnoses were often delayed [5, 6]. Our patient was symptomatic for 15 months with weakness of lower limbs and intergluteal swelling for 2 months fistulized to skin since birth.

Congenital ECs frequently associated with others spinal dysraphism following spina bifida, dermal sinus, and syringomyelia while acquired ECs occur following repeated lumbar punctures, trauma, or surgery [6, 7]. The number of acquired cyst has decreased significantly in recent years. They are attributed to the displacement of epithelial tissue secondary to a previous lumber puncture or trauma [8-10].

The signs and symptoms of these tumors vary with the level of Involvement but do not differ from other lesions in the spinal column [3, 11]. Owing to the characteristic of slow growing diagnosis of this Tumor is sometimes delayed. Our patient's symptoms had been evolving for 15 months. The MRI is the imaging of choice, the lesion appeared on T1-weighted images isointense and hyperintense on T2- weighted and contrast enhancement is uncommon [12-14].

Our patient's MRI of the spine demonstrated an intradural tumor from L3 to L5 levels. It was isointense on T1-weighted images, hyperintense on T2- weighted images with contrast enhancement after gadolinium injection. Nevertheless, in view of these radiological features, the pre- operative diagnosis of epidermoid cyst was not retained in the first instance but was based on the intraoperative macroscopic inspection and confirmed by histology subsequently. Per operatively, an encapsulated "pearly white "tumor was encountered. This macroscopic aspect of the tumor is the same as those reported in the literature [3, 5, 13, 15]. Surgical resection is the treatment of choice. Gross total resection is the goal of surgery to avoid the risk of recurrence and aseptic meningitis. However, when the tumor is tightly attached to the surrounding neural tissue a subtotal excision should be performed to preserve neural function [3, 5, 11, 13]. Although the epidermoid cysts are benign tumors, local recurrence (reported in up to 10-29% of cases) is reported, especially after subtotal excision because the incomplete excision of basal germinal cells [16-18].

CONCLUSION

Gross total resection of the tumor along with the capsule is important to minimize the risk for recurrence and results in resolution of prior neurological deficits.

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