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Case Report

Primary intradural sacral epidermoid in a nondysraphic spine: Case report and review of literature

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Abstract

The occurrence of epidermoids within the spinal canal is uncommon. Most of the reported spinal epidermoids (SEs) have been described in the thoracic or lumbar regions. They occur either following trauma or in the setting of coexistent spinal dysraphism. The authors describe an unusual case of a 28-year-old lady who presented with long-standing back pain and urinary incontinence. Magnetic resonance imaging (MRI) of her spine demonstrated a sacral SE without any coexistent spinal dysraphism. The diagnosis of an epidermoid was confirmed by histopathological examination following laminectomy and excision. To the authors' best knowledge, this is the third case of a sacral SE occurring in a non-dysraphic spine. The case is discussed in the light of a relevant literature review.

Key words: Non-dysraphic, sacral, spinal epidermoid (SE)

INTRODUCTION

Spinal epidermoids (SEs) are uncommon tumors, constituting less than 1% of all intraspinal tumors.[1] Their occurrence in the sacral region is unusual.[1-9] Most of the sacral SEs have been reported either in conjunction with anterior sacral meningoceles, as a component of Currarino syndrome (CS),[10-14] or as sequelae to lumbar punctures.[9,15] This report of a patient with a primary, nonsyndromal sacral SE in the absence of coexistent spinal dysraphism or preceding trauma is the third of its kind in indexed literature.

CASE REPORT

Clinical presentation and examination

A 28-year-old lady presented with low backache since childhood and urinary incontinence of a duration of 1 year. There was no history of weakness or numbness in the limbs, meningitis, lumbar puncture, or trauma to the spine. Her neurological examination revealed impaired sacral sensations. There was no evidence of spinal dysraphism. Urological evaluation revealed a neurogenic bladder with chronic cystitis.

Neuroimaging

Magnetic resonance imaging (MRI) of the spine revealed a 5.1 cm × 4.3 cm × 2.9 cm sized intradural tumor from L5 to S2 levels. It was isointense on T1-weighted images, hyperintense on T2-weighted images, and demonstrated mild peripheral contrast enhancement [Figure 1a-h]. It had an extraspinal extension through the right S1-2 foramen. Computed tomography (CT)

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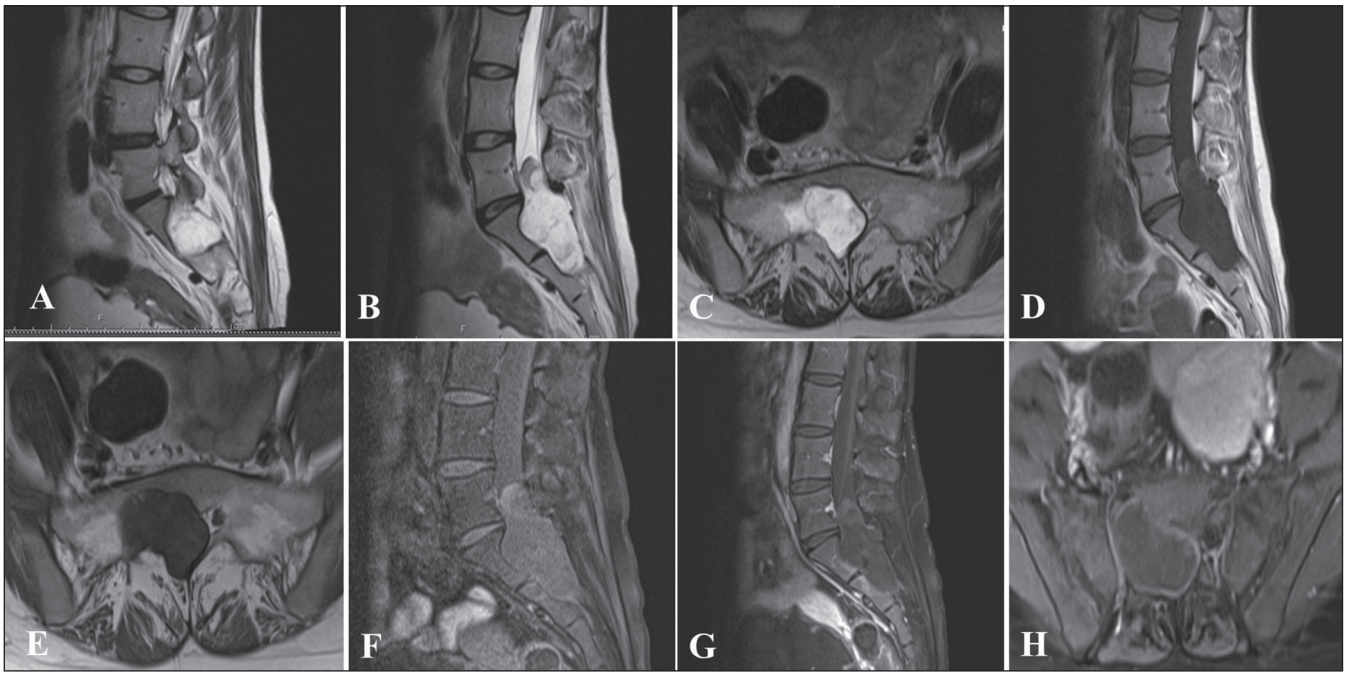

Figure 1: T2-weighted sagittal (A and B) and axial (C) MR images showing a predominantly hyperintense tumor extending from L5 to S2 levels. Scalloping of the S1 and S2 vertebrae is noted. T1 weighted sagittal (D) and axial (E) images showing a hypointense tumor. Postcontrast fat suppression images (G and H) showing mild peripheral enhancement (F)

of the lumbosacral spine showed a widened right S1-2 neural foramen and scalloping of the posterior portions of the S1 and S2 vertebrae [Figure 2a-f]. Based on these findings, a radiological diagnosis of a long-standing benign nerve sheath tumor (schwannoma/neurofibroma) was considered.

Operation and histopathological examination

At surgery, a defect in the right S1 lamina was noted overlying the bulged-out dura. There was no evidence of spinal dysraphism. Following partial L5, S1, and S2 laminectomies and durotomy, a soft, pearly white lesion was encountered [Figure 3a-c]. It had pushed the roots to the left side. The filum terminale appeared normal. The lesion and its capsule were excised totally. Histopathology was reported as epidermoid [Figure 4a and b].

Postoperative course

Postoperatively, she did not develop any new neurological deficit. A postoperative MRI with diffusion-weighted (DW) sequences done showed no obvious residue [Figure 5a and b]. She was discharged on a tapering dose of oral steroids to prevent aseptic meningitis. At 6 months of follow-up period, the patient was doing well; however, her urinary incontinence had remained the same.

DISCUSSION

Epidermoid cysts (ECs) are uncommon benign lesions with an incidence of less than 1% in the spine.[16,17] They commonly present in the fourth decade with a slight male preponderance.[18] Described by Cruveilhier as tumeurs perlées (pearly white tumors)[18] due to their gross appearance, they consist of soft, whitish, keratin material without any element of skin appendage.[19] Congenital ECs are frequently found in association with spinal dysraphic conditions such as tethered cord, low lying conus, dermal sinus, or *spina bifida*,[20] while acquired ECs occur, following repeated lumbar punctures or trauma.[19,20]

Among the SEs, the thoracic region is the commonest site of occurrence, followed by the sacral and cervical regions.[21,22] In addition to the few reports of sacral SEs in indexed literature [Table 1], there are reports as well of presacral ECs occurring as a component of CS or Currarino triad (anorectal stenosis, defect in the sacral bone and a presacral mass). Additionally, there have been reports of the lesion occurring with an anterior sacral meningocele,[13,14] a presacral meningeal cyst that occurs due to agenesis of a portion of the anterior sacrum.

Clinically, sacral SEs often remain asymptomatic due to the accommodative capacity of the sacral spinal canal. They generally exhibit extensions in a cephalad direction into the lumbar canal or laterally through the neural foramina. They may extend ventrally as well, breaching the anterior or posterior sacral wall.[23] Urinary dysfunction with or without back pain may be the presenting symptom, as in our case. Other cases may present with cutaneous manifestations of spinal dysraphism or meningitis.[24]

MRI is the imaging modality of choice for the diagnosis of a sacral SE. The tumor is classically a nonenhancing lesion of cerebrospinal fluid intensity that is isointense or hypointense on T1-weighted sequences and hyperintense on T2-weighted

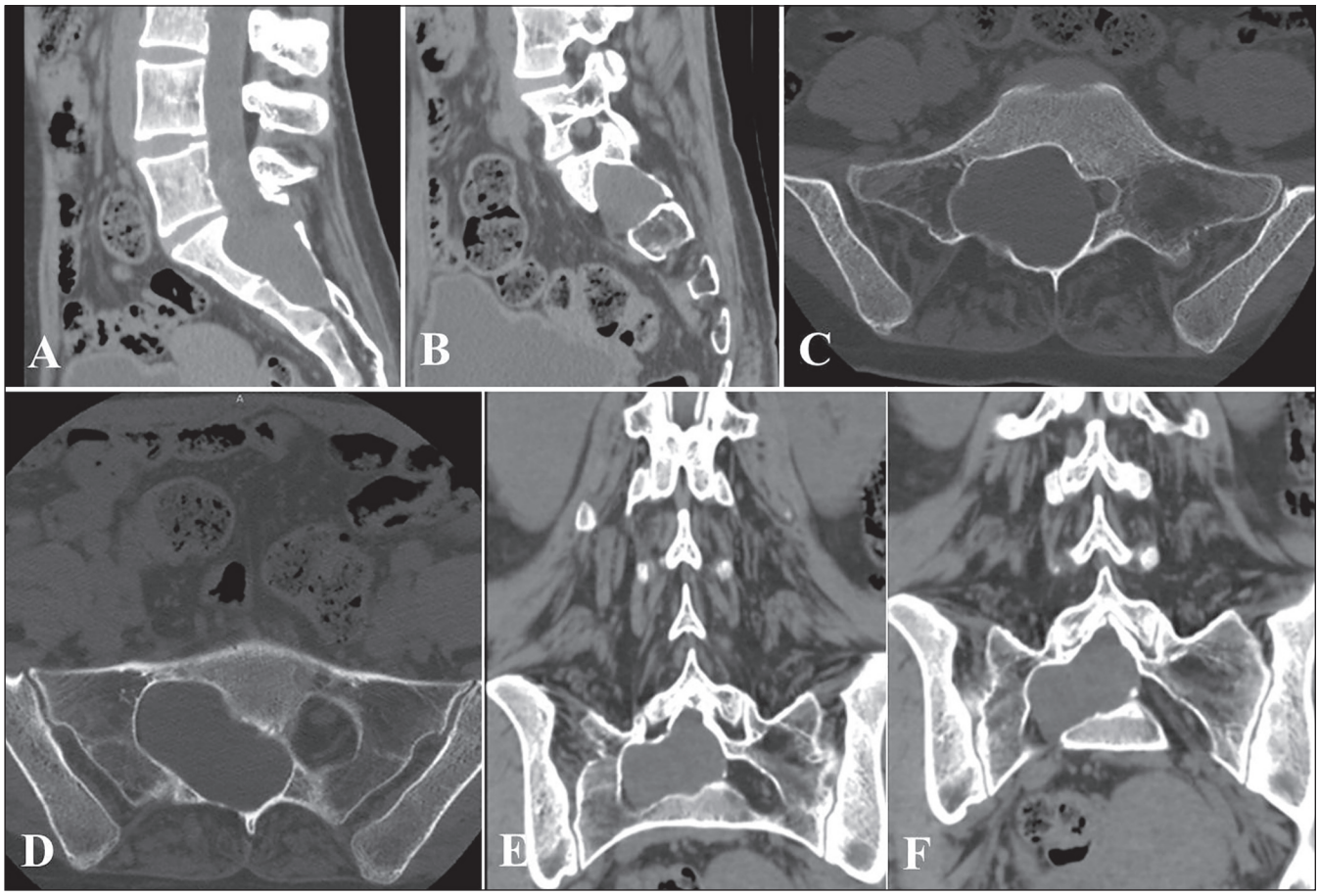

Figure 2: CT sagittal (A and B) axial (C and D) and coronal (E and F) images showing an isodense tumor with bony erosion and remodeling

Table 1: Intra spinal sacral epidermoid tumors reported in literature

Author	Age/sex	Associated with spinal dysraphism	Level
Liu *et al.*[1]	35/M	Dermal sinus, spina bifida	L5-S2
Liu *et al.*[1]	52/F	None	L4-S1
Liu *et al.*[1]	20/M	Dermal sinus, spina bifida, diastematomyelia	L3-S2
Manara *et al.*[2]	25/F	None	L5-S2
van Aalst *et al.*[3]	18 months/M	Dermal sinus	Sacral
van Aalst *et al.*[3]	14 months/F	Myelomeningocele	Lumbosacral
van Aalst *et al.*[3]	12 years/F	Myelomeningocele	Sacral
van Aalst *et al.*[3]	14 months/F	Dermal sinus	Lumbosacral
Hamby *et al.*[4]	3 years/M	Dermal sinus	Lumbosacral
French *et al.*[5]	4 years/F	Sacral spina bifida	T12-S2
Manno[6]	30 months/M	Sacral spina bifida	L5-S1
Matera[7]	28 years/M	Not known	T11- S1
Aulbach[8]	7 years/F	Dimple over lumbar area	L2-S5
Tipton[9]*	Not known	Not known	Sacral canal

*History of prior lumbar puncture present

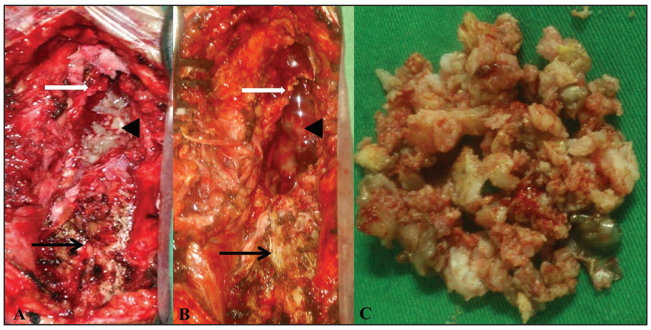

Figure 3: Intraoperative image demonstrating (A) The tumor cavity with a pearly white tumor (arrow head) (B) The tumor bed after excision (white arrow indicating the remaining L5 lamina and the black arrow pointing at the caudal spinal laminae) (C) Gross specimen of the whitish avascular tumor

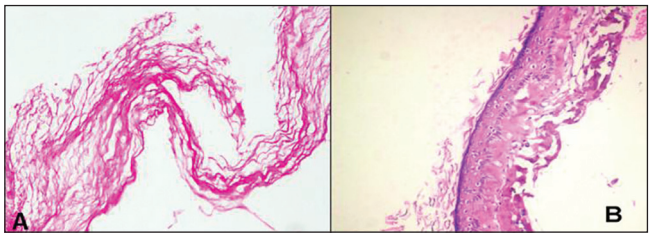

Figure 4: Paraffin section of the lesion demonstrating (A) Anucleated squames with (B) Stratified squamous lining epithelium. [Hematoxylin & Eosin (A, B) 100×]

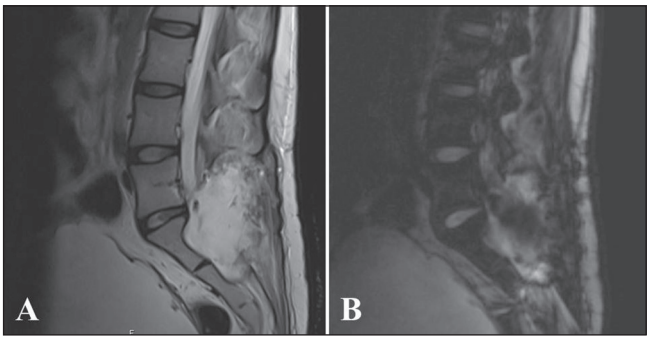

Figure 5: Postoperative T2-weighted (A) and diffusion-weighted (B) Sagittal MR images demonstrating total excision of the tumor with no obvious residue

sequences.[20] Discrepancy in the intensity characteristics occur due to varying lipid and protein components. Abnormalities in vertebral bodies or posterior elements may be evident in long-standing lesions. DW imaging demonstrates restricted diffusion such as in ECs elsewhere, and may be of benefit while evaluating a cystic sacral lesion.[2]

Total excision remains the treatment of choice in symptomatic SEs compressing the thecal sac.[20] However, a densely adherent capsule may preclude total excision,[21] resulting in early relapses. Residual cystic contents predispose to the occurrence of aseptic meningitis in the postoperative period, a major complication that can result in the development of normal pressure hydrocephalus.[17,22,24]

CONCLUSIONS

SEs in the sacral region are rare. This is the third report of a sacral intradural SE occurring in a non-dysraphic spine. Epidermoid should be considered in the differential diagnosis of an intradural sacral tumour. When suspected, the radiological evaluation of such a lesion should include diffusion weighted imaging.

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Conflicts of interest

The authors declare that they have no conflict of interest.

REFERENCES

1. Liu H, Zhang JN, Zhu T. Microsurgical treatment of spinal epidermoid and dermoid cysts in the lumbosacral region. J Clin Neurosci 2012;19:712-7.
2. Manara R, Severino M, Mandari R, Mattisi G, Dal Pozzo S, Carollo C. Chronic cystic lesion of the sacrum: Characterisation with diffusion-weighted MR imaging. Radiol Med 2008;113:739-46.
3. van Aalst J, Hoekstra F, Beuls EA, Cornips EM, Weber JW, Sival DA, *et al.* Intraspinal dermoid and epidermoid tumors: Report of 18 cases and reappraisal of the literature. Pediatr Neurosurg 2009;45:281-90.
4. Hamby WB. Tumors in the spinal canal in childhood. II. Analysis of the literature of a subsequent decade (1933-1942); report of a case of meningitis due to an intramedullary epidermoid communicating with a dermal sinus. J Neuropathol Exp Neurol 1944;3:397-412.
5. French LA, Peyton WT. Mixed tumors of the spinal canal. Arch Neural Psychiat 1942;47:737-51.
6. Manno NJ, Uihlein A, Kernohan JW. Intraspinal epidermoids. J Neurosurg 1962;19:754-65.
7. Matera RF, Martino A. Epidermoide gigaate infectado de cola de caballo. Arch Neurochir 1945;2:87-96.
8. Aulbach R. Beitrag zur Xtiologie und zum klinisehen Bild der fixierten Ledenlordose. Z Orthop 1950;79:645-50.
9. Tipton W Jr, D'Ambrosia RD, Corkill G. Intraspinal epidermoid tumor of the sacral canal presenting with urinary retention. J Urol 1975;113:880-3.
10. Shoji M, Nojima N, Yoshikawa A, Fukushima W, Kadoya N, Hirosawa H, *et al*. Currarino syndrome in an adult presenting with a presacral abscess: A case report. J Med Case Rep 2014;8:77.
11. Nakamura S, Wakamatsu K, Tsubokawa T, Moriyasu N. Sacral epidermoid cyst communicating with the spinal CSF canal. Childs Brain 1980;6:103-11.
12. Li GD, Chen K, Fu D, Ma XJ, Sun MX, Sun W, *et al.* Surgical strategy for presacral tumors: Analysis of 33 cases. Chin Med J (Engl) 2011;124:4086-91.
13. Shamoto H, Yoshida Y, Shirane R, Yoshimoto T. Anterior sacral meningocele completely occupied by an epidermoid tumor. Childs Nerv Syst 1999;15:209-11.
14. Kansal R, Mahore A, Dange N, Kukreja S. Epidermoid cyst inside anterior sacral meningocele in an adult patient of Currarino syndrome manifesting with meningitis. Turk Neurosurg 2012;22:659-61.
15. Nica DA, Strambu VE, Roşca T, Cioti D, Copaciu R, Stroi M, *et al.* Acquired epidermoid cysts of the cauda equina. J Med Life 2011;4:305-9.
16. Chandra PS, Manjari T, Devi BI, Chandramouli BA, Srikanth SG, Shankar SK. Intramedullary spinal epidermoid cyst. Neurol India 2000;48:75-7.
17. Guidetti B, Gagliardi FM. Epidermoid and dermoid cysts. Clinical evaluation and late surgical results. J Neurosurg 1977;47:12-8.
18. Roux A, Mercier C, Larbrisseau A, Dube LJ, Dupuis C, Del Carpio R. Intramedullary epidermoid cysts of the spinal cord. Case report. J Neurosurg 1992;76:528-33.
19. Penisson-Besnier I, Guy G, Gandon Y. Intramedullary epidermoid cyst evaluated by computed tomographic scan and magnetic resonance imaging: Case report. Neurosurgery 1989;25:955-9.
20. Yin H, Zhang D, Wu Z, Zhou W, Xiao J. Surgery and outcomes of six patients with intradural epidermoid cysts in the lumbar spine. World J Surg Oncol 2014;12:50.
21. Kansal R, Mahore A, Dange N. Giant intramedullary epidermoid extending from the brain stem to the upper thoracic spinal cord. Turk Neurosurg 2012;22:452-3.
22. Ogden AT, Khandji AG, McCormick PC, Kaiser MG. Intramedullary inclusion cysts of the cervicothoracic junction. Report of two cases in adults and review of the literature. J Neurosurg Spine 2007;7:236-42.
23. Feldenzer JA, McGauley JL, McGillicuddy JE. Sacral and presacral tumors: Problems in diagnosis and management. Neurosurgery 1989;25:884-91.
24. Lunardi P, Missori P, Gagliardi FM, Fortuna A. Long-term results of the surgical treatment of spinal dermoid and epidermoid tumors. Neurosurgery 1989;25:860-4.