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Sacral extradural arachnoid cyst: a rare cause of low back and perineal pain

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Abstract Sacral extradural arachnoid cysts are rare. The clinical and radiological features of this condition are characteristic. One such rare case with low back and perineal pain is presented and the literature is reviewed. This patient presented with pain in the low back and perineal region, which was aggravated by standing, walking and straining. The patient also had numbness in both lower limbs, precipitated by standing and walking. Both the symptoms were relieved by lying down. Magnetic resonance imaging (MRI) re-

vealed a sacral extradural arachnoid cyst. Sacral laminectomy with opening of the arachnoid cyst and ligation of the fistulous tract was done. Postoperatively, there was complete clinical recovery. Though rare, this entity should be considered in the differential diagnosis of low back and perineal pain. Surgical treatment is curative.

Keywords Arachnoid cyst · MRI · Occult intrasacral meningocele · Occult spinal dysraphism · Sacrum

Introduction

Sacral extradural arachnoid cysts are rare. When present, they produce low back pain and pain in the perineal region, which is aggravated by Valsalva maneuvers and relieved by lying down. A high index of suspicion is required to diagnose this condition. To date, 41 cases have been reported in the English language literature (Table 1).

Case report

A 25-year-old man presented with a 2-year history of pain over the low back and perineal region, which had become severe 1 month prior to admission. The pain was aggravated by standing, walking and straining, and was relieved partially by lying down in the prone position. The patient also complained of numbness of both lower limbs on prolonged standing or walking, which was relieved by lying down. There was no other positive history. Examination revealed normal higher functions and cranial nerves. The patient had weakness of the extensor hallucis longus on both sides, left more than right. There was also tenderness over the upper sacral region. Bladder, bowel and sexual functions were normal. There were no other focal neurological deficits. Plain radiographs of the

lumbosacral region revealed a suspicious bony erosion in the mid-sacral region. Magnetic resonance imaging (MRI) revealed a cystic lesion opposite the second and third sacral segments, which was isointense to cerebrospinal fluid (CSF) on T1- and T2-weighted images (Fig. 1). There was evidence of communication of the cyst with the caudal end of the thecal sac. There was no communication with the pelvis. Scalloping of the sacrum was evident. At surgery, laminectomy of S1–S4 was done, and a large, thin-walled bluish cystic mass containing CSF was encountered, which was opened. The cystic mass was found to be communicating with the thecal sac through a defect in the theca. The fistulous communication was obliterated and the defect in the theca was closed with purse-string sutures. Postoperatively, the patient noted marked relief of pain and did not have numbness while standing and walking. Two years following surgery, the patient was neurologically normal without any deficits.

Discussion

The term “occult sacral meningocele” was first introduced by Enderle in 1932 [19]. It has come to mean a

sac composed of fibrous tissue resembling dura mater that is usually lined by arachnoid, lies within an enlarged sacral spinal canal, and is attached to the distal termi-

Table 1 A summary of previously reported cases of sacral extradural arachnoid cysts

Serial no.	Authors	Year	No. of cases	Associated conditions
1	Young et al. [31]	1969	1	Nil
2	Joseph & McKenzie [15]	1970	1	Nil
3	Kak et al. [16]	1972	1	Nil
4	Reddy et al. [28]	1974	1	Nil
5	Florez & Ucar [9]	1976	1	Intrasacral and presacral neurofibroma
6	Gelmers & Go [10]	1977	1	Nil
7	Lamas et al. [19]	1977	1	Nil
8	Holness et al. [14]	1978	1	Anterior sacral meningocele and suprasellar teratocarcinoma
9	McCreath & Macpherson [20]	1980	1	Nil
10	Fardon [8]	1980	1	Transverse fracture of sacrum
11	Kumar et al. [18]	1980	1	Nil
12	Grivegneee et al. [12]	1981	1	Nil
13	Agrillo et al. [1]	1983	1	Lumbar disc prolapse
14	Genest [11]	1984	1	Nil
15	Roosen et al. [29]	1985	1	Nil
16	Mink [21]	1985	1	Nil
17	Doty et al. [6]	1989	4	Nil
18	Cole et al. [4]	1989	1	Nil
19	Harken & el-Khoury.[13]	1990	1	Marfan syndrome
20	North et al. [23]	1990	1	Bilateral anterior sacral and perineurial cysts
21	Bayar et al. [2]	1991	2	Lumbar disc prolapse
22	Rabb et al. [25]	1992	1	Nil
23	Boukoba et al. [3]	1993	6	Nil
24	Tatagiba et al. [30]	1994	1	Lumbar disc prolapse
25	Raftopoulos et al. [27]	1995	1	Anterior sacral meningocele and Marfan syndrome
26	el Mostarchid & Bellakdhar [7]	1995	1	Nil
27	Doi et al. [5]	1995	1	Tethered cord
28	Raftopoulos et al. [26]	1995	3	Nil
29	Okada et al. [24]	1996	1	Spina bifida
30	Kim et al. [17]	1999	1	Intradural arachnoid cyst

Fig. 1 A,B Sagittal T1- and T2-weighted sequences of the lumbosacral spine, showing a cystic lesion in the sacral spinal canal with the intensity of cerebrospinal fluid and scalloping of the sacrum

Fig. 2 A Type Ia spinal meningeal cyst (extradural meningeal cyst). B Type Ib spinal meningeal cyst (occult intrasacral meningocele). C Type II spinal meningeal cyst (Tarlov's perineurial cyst). D Type III spinal intradural meningeal cyst (intradural arachnoid cyst)

extradural meningeal cysts with nerve root fibers (Tarlov's perineurial cysts) (Fig. 2C), and type III are intradural arachnoid cysts (Fig. 2D) [22]. According to the Nabors et al. classification, the lesion described in this patient is a type Ib meningeal cyst.

nation of the dural sac by a narrow or broad pedicle that usually permits a free flow of cerebrospinal fluid from the distal thecal sac into the meningocele [6].

However, traditionally a meningocele is believed to consist of a herniation of the meninges outside the confines of the spinal canal. Since, in this particular condition, there is no herniation of the meningeal elements outside the confines of the spinal canal, and there is no evidence of associated spinal dysraphism, and the clinical as well as the radiological picture is similar to the spinal extradural arachnoid cysts that occur at higher spinal levels, it would be more appropriate to name this condition "sacral extradural arachnoid cysts" than "occult intrasacral meningocele". In addition, there is no evidence that this lesion is developmental. This author, therefore, believes that the term "intrasacral meningocele" should not be used to refer to these lesions.

Nabors et al. [22] classified spinal meningeal cysts into three types. Type I are extradural meningeal cysts without spinal nerve root fibers; these were further subdivided into type Ia, extradural arachnoid cysts (Fig. 2A) and type Ib, occult intrasacral meningoceles (Fig. 2B). Type II are

Clinical presentation

The clinical features of sacral extradural arachnoid cysts vary from being asymptomatic to sensory disturbances in the sacral dermatomes and, occasionally, sphincter disturbances [2, 6, 19, 30]. Patients usually present in the third or fourth decade of life with dull, intermittent pain in the low back of insidious onset. Pain is usually aggravated by activity and Valsalva maneuver. The pain may radiate to the posterior thigh or calf. Sphincter disturbances may be present. Sensory loss, if present, usually involves the sacral dermatomes. Motor strength in the lower extremities is normal. Since the clinical presentation is similar to that of lumbar disc prolapse, this condition is frequently underdiagnosed. However, several authors have reported that sacral extradural arachnoid cysts can co-exist with lumbar disc prolapse, and in such cases the lumbar disc prolapse and not the cyst was considered to be the cause of the symptoms [1, 2, 30]. In our patient, even though the clinical features were suggestive of disc prolapse, MRI did not reveal any associated disc lesion.

Mechanism of formation and enlargement of sacral extradural arachnoid cysts

The exact mechanism of formation and enlargement of these cysts is not known. However, it is believed that the arachnoid herniates through a small, probably congenital, dural defect in the caudal end of the thecal sac. This arachnoidal "diverticulum" progressively increases in size because of easy egress of CSF into it in the erect posture. Valsalva maneuvers that occur during daily activities might increase the volume of the cyst. Increase in pressure and volume of the cyst during erect posture and during Valsalva maneuvers might produce traction on the roots of the cauda equina by the arachnoid "diverticulum", and might be responsible for the sensory and motor disturbances that are often seen in these patients. The passage of CSF back into the thecal sac while lying down might explain the relief of symptoms that occurs in this posture. Even though intuitive, the above theory explains the symptomatology of these lesions.

Radiology

Plain radiographs may reveal widening of the sacral canal with scalloping of the sacral vertebral bodies. Previously, myelography and computed tomographic (CT) myelogra-

phy with water-soluble contrast media were used to diagnose this condition [6, 19]. In both these investigative procedures, delayed studies were used to demonstrate the fistulous communication as well as the contrast filling of the meningocele sac. However, these studies have now been replaced by MRI, which clearly delineates the nature of the lesion as well as the associated anomalies [3, 6, 17]. MRI shows a cystic lesion with signal characteristics of CSF in both T1- and T2-weighted sequences.

Treatment

Sacral laminectomy, with opening of the cyst and identification and closure of the fistulous communication, is the treatment of choice. Recently, endoscopic treatment of these lesions has been reported [26, 27]. Improvement of symptoms following surgery is the rule. To date, there has been no case of recurrence of the cyst.

Conclusions

Though rare, sacral extradural arachnoid cysts should be considered in the differential diagnosis of pain in the low back and perineal region. MRI is the investigation of choice and surgery is curative.

References

1. Agrillo U, Vangelista S, Pirrone R, Simonetti G (1983) Intracanal meningocele associated to lumbar disk rupture. A case report. *J Neurosurg Sci* 27:265-267
2. Bayar MA, Yidiz B, Buharali Z (1991) Management problems in cases with a combination of asymptomatic occult intracanal meningocele and disc prolapse. *Acta Neurochir (Wien)* 108: 67-69
3. Boukobza M, Sichez JP, Roland E, Cornu P (1993): MRI evaluation of sacral cysts. *J Neuroradiol* 20:266-271
4. Cole GP, Flannery AM, Gulati AK (1989) Intracanal meningocele. Case report and review of the literature. *Spine* 14:1418-1420
5. Doi H, Toyoda I, Matsumoto K, Imai S, Sekimizu M (1995) Occult intracanal meningocele with tethered cord: case report. *Neurol Med Chir (Tokyo)* 35:321-324
6. Doty JR, Thomson J, Simonds G, Ren-gachary SS, Gunby EN (1989) Occult intracanal meningocele: clinical and radiographic diagnosis. *Neurosurgery* 24:616-625
7. el Mostarchid B, Bellakhdar F (1995) Intracanal meningocele. Report of a case. *Rev Rhum Engl Ed* 62:212-214
8. Fardon DF (1980) Intracanal meningocele complicated by transverse fracture. A case report. *J Bone Joint Surg Am* 5:839-841
9. Florez G, Ucar S (1976) The occult intracanal meningocele. *Neurochirurgia (Stuttg)* 19:46-53
10. Gelmers HJ, Go KG (1977) Intracanal meningocele. *Acta Neurochir (Wien)* 39:115-119
11. Genest AS (1984) Occult intracanal meningocele. *Spine* 9:101-103
12. Grivegnee A, Delince P, Ectors P (1981) Comparative aspects of occult intracanal meningocele with conventional x-ray, myelography and CT. *Neuroradiology* 22:33-37
13. Harken KL, el-Khoury GY (1990) Intracanal meningocele in a patient with Marfan syndrome. Case report. *Spine* 15:610-612
14. Holness RO, Hoffman HJ, Mancer K, Armstrong D (1978) Intracranial teratocarcinoma in a child with anterior sacral and intracanal meningocele. *Neurosurgery* 2:143-147
15. Joseph RA, McKenzie T (1970) Occult intracanal meningocele. *J Neurol Neurosurg Psychiatry* 33:493-496
16. Kak VK, Chugh KS, Sodhi JS (1972) Occult intracanal meningocele. *Neurochirurgia (Stuttg)* 15:148-152
17. Kim CH, Bak KH, Kim JM, Kim NK (1999) Symptomatic sacral extradural arachnoid cyst associated with lumbar intradural arachnoid cyst. *Clin Neurol Neurosurg* 101:148-152
18. Kumar S, Kaza RC, Nagpal S, Kaza R, Sehgal H (1980) Occult intracanal meningocele (a case report). *Indian Pediatr* 10:839-842
19. Lamas E, Lobato RD, Amor T (1977) Occult intracanal meningocele. *Surg Neurol* 8:181-184
20. McCreath GT, Macpherson P (1980) Sonography in the diagnosis and management of anterior sacral meningocele. *J Clin Ultrasound* 2:133-137
21. Mink JH (1985) Case report 313. Diagnosis: intracanal cyst ("occult intracanal meningocele"). *Skeletal Radiol* 13:314-317

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22. Nabors MW, Pait GT, Byrd EB, Karim NO, Davis DO, Kobrine AI, Rizzoli HV (1988) Updated assessment and current classification of spinal meningeal cysts. *J Neurosurg* 68:366–377
 23. North RB, Kidd DH, Wang H (1990) Occult bilateral anterior sacral and intrasacral meningeal and perineurial cysts: case report and review of the literature. *Neurosurgery* 27:981–986
 24. Okada T, Imae S, Igarashi S, Koyama T, Yamashita J (1996) Occult intrasacral meningocele associated with spina bifida: a case report. *Surg Neurol* 46:147–149
 25. Rabb CH, McComb JG, Raffel C, Kennedy JG (1992) Spinal arachnoid cysts in the pediatric age group: an association with neural tube defects. *J Neurosurg* 77:369–372
 26. Raftopoulos C, Baleriaux D, Hancq S, Closset J, David P, Brotchi J (1995) Evaluation of endoscopy in the treatment of rare meningoceles: preliminary results. *Surg Neurol* 44:308–317
 27. Raftopoulos C, Pierard GE, Retif C, Braude P, Brotchi J (1995) Endoscopic cure of a giant sacral meningocele associated with Marfan's syndrome: case report. *Neurosurgery* 30:765–768
 28. Reddy DR, Sathyanarayana K, Krishnamurthi D (1974) Occult intrasacral meningocele: report of a case. *Aust N Z J Surg* 44:273–275
 29. Roosen N, VanVyve M, De Moor J (1985) Intrasacral meningeal cyst demonstrated by sacral epidurography. *Neuroradiology* 27:123,124
 30. Tatagiba M, Aguiar PH, Samii M (1994) Management of occult intrasacral meningocele associated with lumbar disc prolapse. *Neurosurg Rev* 17:313–315
 31. Young IS, Bruwer AJ (1969) The occult intrasacral meningocele. *Am J Roentgenol Radium Ther Nucl Med* 105:390–399