Case Report

Pediatric Symptomatic Sacral Extradural Arachnoid Cyst: Surgical Management Review

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Spinal arachnoid cyst (SAC) constitutes about less than 1% of all spinal tumors. It commonly occurs in third to fifth decades of life and mostly located in the thoracic region. Sacral extradural arachnoid cyst in the pediatric age group is an extremely rare location of occurrence. A such rare case of extradural arachnoid cyst of the sacral region, who presented with urinary incontinence is reported. The patient was managed surgically with laminectomy at S1–S3 vertebral level and complete excision of the extradural sacral arachnoid cyst with closure of transdural communication. The patient tolerated surgical procedure very well with regained bladder sphincter control at 6 months of follow-up. Early diagnosis and surgery are recommended for the treatment of symptomatic pediatric sacral arachnoid cysts.

**Keywords:** Extradural spinal cyst, management, surgery, sacral arachnoid cyst

**Introduction**

Spinal arachnoid cysts (SAC) are rare lesions.[1-7] They are most commonly observed in the thoracic region, and the sacral region is considered extremely uncommon.[1] SAC tends to affect men preferentially with peak incidence in the second to fifth decades of life and is extremely rare in the pediatric age groups.[1,2] Most patients present with low back pain and pain in the perineal region, aggravated by coughing, straining, or Valsalva maneuver and typically relieved by lying down.[4] Here the occurrence of a sacral SAC in a child and the successful management is reported.

**Case Illustration**

A 5-year-old male child presented to our outpatient services with complaints of nocturnal enuresis with occasional dribbling of urine and feeling of incomplete urinary emptying of bladder for the last 2 years. The patient was initially evaluated for any urological cause by a local urologist. Cystoscopy was performed to rule out any urological cause for the symptoms, but no local cause was noted and the child was then referred to our center for further management. On examination at admission, it was found that the child was average built with no cutaneous stigma of spinal dysraphism, neurofibromatosis, or spinal deformity. Neurological examination was essentially within normal limit.

Magnetic resonance imaging study revealed presence of a cystic lesion extending from the first to the third sacral segments [Fig-1] and showing isointense signal similar to cerebrospinal fluid (CSF) on T1- as well as T2-weighted images [Fig-2]; no contrast enhancement was observed, and a provisional diagnosis of sacral extradural arachnoid cyst was made.

He was taken up for elective surgery in the prone position under general anesthesia, and S1–S3 laminectomy was done. After laminectomy, an extradural large cystic lesion was noted, containing clear fluid-like CSF causing remodeling of bone. The cyst was completely extradural and a small communication through dura was seen, which ligated and reinforced along with excision of the cyst. A histopathologic study of the cyst wall showed fibrous connective tissue with an inner single-cell lining with the absence of any meningothelial...
or neural elements. He had uneventful postoperative course and was discharged from hospital on the third day. At a follow-up visit 6 months later, his bladder symptoms had resolved completely.

**DISCUSSION**

SACs constitute about 1% of all spinal tumors. SACs occurring in the spinal region are mostly noted in the third to fifth decades of life. They may be located either intradurally or extradurally, the latter being relatively uncommon. The extradural arachnoid cysts are mostly encountered in the thoracic spine (65%), with the other spinal region being extremely rare. Sacral cysts constitute about 5–7% of all adult SAC cases.

The etiology of spinal extradural arachnoid cysts is still ill-understood. Many postulates are put forward. Active fluid secretion from the arachnoid cyst wall, passive osmosis of water, and hydrostatic pressure of CSF, ball-valve mechanism with unidirectional flow of CSF and entrapment, cause gradual cyst enlargement.

Rohrer et al. observed the ball-valve mechanism intraoperatively. Ball-valve mechanism in the communicating pedicle is associated with pulsatile CSF dynamics and results in cyst enlargement.

Most SACs have been diagnosed incidentally in the middle ages. The common symptoms of SAC located in the sacral region are sphincter dysfunction, low backache, or radiculopathy, with sexual impotence being a rare presentation.

Magnetic resonance imaging is the most common imaging modality of choice for diagnosis, which clearly delineating the location, extension, relation to dura and nerve root, and the presence of communication, with lobulation being single or multiple, and the presence of other congenital anomalies.

Differential diagnosis includes synovial cyst, Tarlov cyst, meningocele, dermoid cyst, and meningeal diverticula along spinal nerve roots, and cystic schwannoma or meningioma changes. Goyal et al. observed that SACs of the sacral region were called with different nomenclatures, producing ambiguity including sacral meningoceles, arachnoid pouches, arachnoid diverticula, and meningeal cysts.

Nabors et al. have classified spinal meningeal cysts into three types: type I, extradural cysts with nerve root fibers; type II, extradural cyst without nerve root fibers; and type III, intradural cysts. Type I can be further subclassified: type IA is called extradural arachnoid cyst whereas type IB is called sacral meningocele. Type III is also called as intradural arachnoid cyst or Tarlov cyst.

Asymptomatic or incidentally detected or smaller SACs can be managed conservatively. Surgical aspiration of the cyst will result in only temporary improvement. Surgical excision of the cyst is considered as the treatment of choice in symptomatic SAC cases. Patients should be regularly followed up after surgical management.

**CONCLUSION**

Pediatric cases especially presenting with sphincter disturbance should also be investigated for SAC. Although rare, it represents a benign condition, which can be corrected by early surgery, providing gratifying...
result with recovery of bladder control, as occurred in the present case. Pediatricians and neurosurgeons should be careful of such an entity. A high index of clinical suspicion is required and neuroimaging can prove to be a definitive aid in diagnosis and surgical planning. Early surgery is a safe and effective method for relieving symptoms.

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**Conflicts of interest**
There are no conflicts of interest.

**References**
1. Liu JK, Cole CD, Kan P, Schmidt MH. Spinal extradural arachnoid cysts: Clinical, radiological, and surgical features. Neurosurg Focus 2007;22:E6.
2. Rohrer DC, Burchiel KJ, Gruber DP. Intraspinal extradural meningeal cyst demonstrating ball-valve mechanism of formation. Case report. J Neurosurg 1993;78:122-5.
3. Choi JY, Kim SH, Lee WS, Sung KH. Spinal extradural arachnoid cyst. Acta Neurochir (Wien) 2006;148:579-85.
4. Muthukumar N. Sacral extradural arachnoid cyst: A rare cause of low back and perineal pain. Eur Spine J 2002;11:162-6.
5. Goyal RN, Russell NA, Benoit BG, Belanger JM. Intraspinal cysts: A classification and literature review. Spine 1987;12:209-13.
6. Nabors MW, Pait TG, Byrd EB, Karim NO, Davis DO, Kobrine AI, et al. Updated assessment and current classification of spinal meningeal cysts. J Neurosurg 1988;68:366-77.
7. Thakar S, Kiran NAS, Hegde AS. A sacral arachnoid cyst causing holocord syringomyelia. J Neurosurg Pediatr 2011;8:299-302.
8. Kalkarni AG, Goel A, Thiruppathy SP, Desai K. Extradural arachnoid cysts: A study of seven cases. Br J Neurosurg 2004;18:484-8.
9. Bond AE, Zada G, Bowen I, McComb JG, Krieger MD. Spinal arachnoid cysts in the pediatric population: Report of 31 cases and a review of the literature. J Neurosurg Pediatr 2012;9:432-41.