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Multiloculated Extra Dural Intraspinal Giant Arachnoid Cyst- Case Report and Literature Review

Dr. Sanjay Sharma, M.S M.Ch[1]*, Dr. Ojasvi Vats[2] and Dr. Garima JR[3]

[1]*Neurosurgery Department, Muzaffarnagar Medical College (MMC), Medwin Hospital, Meerut, India.*

[2]*Junior Resident, Pacific Medical College, Udaipur, India.*

[3]*L.L.R.M Medical College, Meerut, India.*

***Correspondence:**
Dr. Sanjay Sharma, Neurosurgery Department, Muzaffarnagar Medical College (MMC), Medwin Hospital, Meerut, India.

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Keywords
Arachnoid cyst, Spinal arachnoid cyst, Extradural arachnoid cyst, Intraspinal cyst, Giant arachnoid cyst.

Background
An extradural intraspinal cyst, also known as a spinal extradural arachnoid cyst (SEAC), is a rare fluid-filled sac that develops outside the spinal cord dura mater but within the spinal canal. The Causes of spinal extradural arachnoid cysts remain unclear. Extradural spinal arachnoid cysts (SEACs) are uncommon, representing approximately 1% of all primary spinal mass lesions. They appear to be extradural outpouchings of arachnoid that communicate with the intraspinal subarachnoid space through a small defect in the dura mater [1]. Enlargement may cause progressive signs and symptoms caused by spinal cord compression. These cysts are often linked to a small defect in the dura, the outermost membrane surrounding the spinal cord, through which the arachnoid membrane (a middle layer) protrudes. They are generally thought to be either congenital (present from birth) or acquired due to factors like trauma or inflammation. Most nontraumatic spinal extradural arachnoid cysts are thought to be congenital.

They are most commonly found in the thoracic spine, particularly the mid-thoracic and thoracolumbar regions, but can occur at any level of the spine. These cysts are typically filled with cerebrospinal fluid (CSF) due to their connection with the subarachnoid space, the area surrounding the spinal cord.

Symptoms vary depending on the size and location of the cyst and the degree of spinal cord or nerve root compression. Common symptoms include back pain, radiculopathy (nerve root pain), neurogenic claudication (leg pain with walking), and in some cases, myelopathy (spinal cord dysfunction) with weakness or bowel/bladder problems.

Magnetic Resonance Imaging (MRI) is the preferred method for diagnosing SEACs, as it can clearly visualize the cyst, its relationship to the spinal cord, and any potential compression. Surgical intervention is often considered for treatment of symptomatic cysts. The specific surgical approach depends on the cyst's characteristics and may involve cyst fenestration (creating an opening) or Dural defect repair.

Case Report and Literature Review
A 13 years old child presented with pain and weakness in both lower limbs for last 3 months. he had history of repeated falls many times while walking. Neurologic examination revealed some sensory loss below bilateral knee. motor strength of 4/5 for both lower extremities. Bilateral plantar extension responses were present, and both Achilles tendons were revealed hyperreflexia. Toes and fingers of both feet were flexor position of distal phalanges and extensor position of proximal phalanges even at rest.

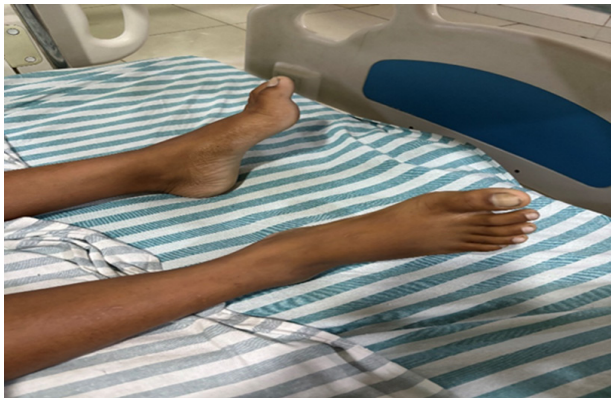

Figure 1: Position toes even at rest.
Thoracolumbar magnetic resonance imaging (MRI) of patient demonstrated a large D9-L3 Extra Dural Multiloculate cyst.

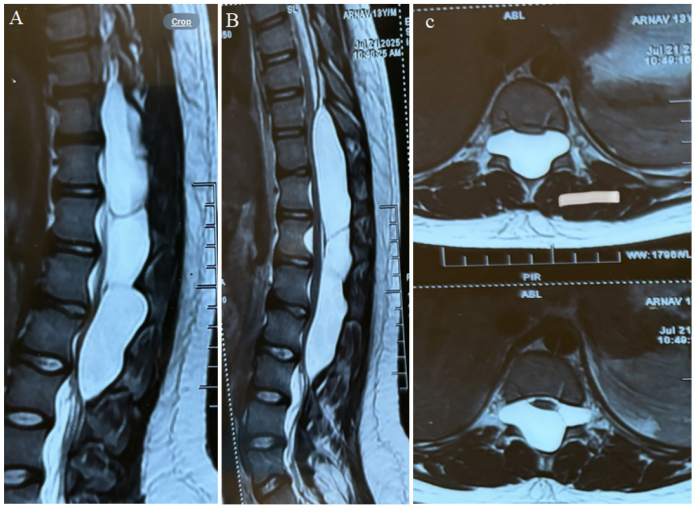

Figure 2: (A) Sagittal T2-weighted image of the thoracic spine reveals loculated arachnoid cysts separated by fibrous septa. The spinal cord is compressed and displaced to the anterior of the spinal canal.
(B) On sagittal T2-weighted image of the lumbar spine, another loculation of the arachnoid cyst is seen, located at L1-L3 levels and separated from the superior one with fibrous septa.
(C) Axial T2-weighted image from distal thoracic level shows extra dural loculated arachnoid cyst compressing the spinal cord.

Surgical management: laminectomy D9 to L3 and resection of the cyst followed by duraplasty. At surgery, the cyst was dissected carefully from the underlying dura after cyst contents were removed. Multiple septa present which were removed carefully. Communicating defects between the dura sac and cyst was identify which was closed with the help of fascia and muscle graft. Patient condition improved after surgery. The stiffness reduced and muscles power increased as toes and finger spasm relieved as shown in photo (photo no.4)

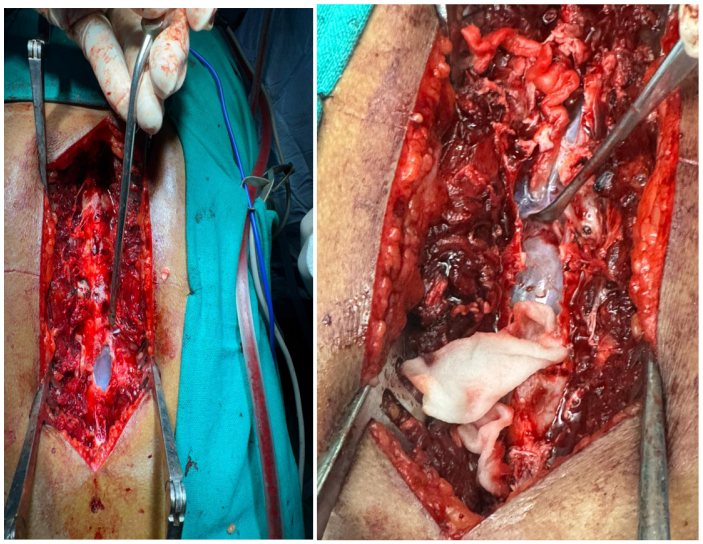

Figure 3(A): Extra dural arachnoid cyst visible. **Figure 3(B):** Showed loculation and septa of arachnoid cyst.

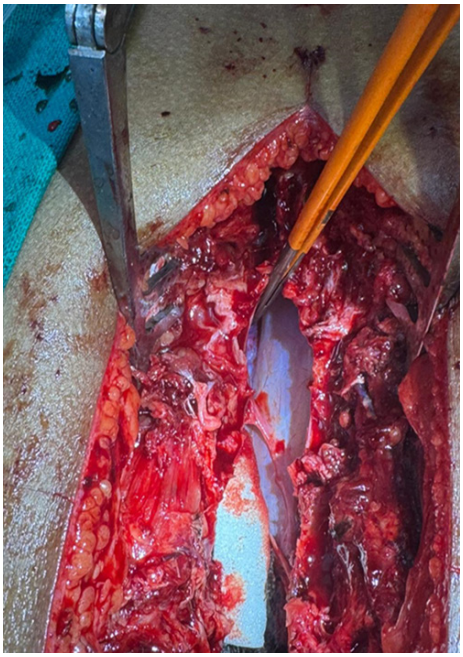

Figure 3(C): Arachnoid cyst was removed completely and septae were desected as much as posible. Duraplasty was done with muscle fascia graft.

Figure 4: Post operative after 3months-standing and walking without support.

Discussion and Review of Litreture

Spinal extradural arachnoid cysts are rare lesions of the spine and represent 1% of the expansive spinal lesions [2]. They present more frequently in young male adults (male to female ratio of 2:1) [3] and males are predominantly affected during the second decade of life [4]. The associated causes are spinal trauma, surgery,

arachnoiditis and neural tube defects [5]. Various hypotheses have been proposed to explain why spinal arachnoid cysts form: (1) proliferation or loculation of arachnoid trabeculae in the septum posticum; (2) a fault in the expansion of the arachnoid trabecula; and (3) the formation of arachnoid diverticuli in weak areas of the spinal dura mater [6,7].

Another aetiological explanation is that the condition is genetic; through the identification of heterozygotic mutations that result in loss of FOXC2 gene function (forkhead box C2 genes) in patients with familial spinal extradural arachnoid cysts. However, sporadic cysts are associated with mutations that cause loss of function of the HOXD4 gene (U homebox D4 family genes) [8]. Subsequently the active secretion of liquid through the cyst walls, passive osmosis of water and hydrostatic pressure of cerebrospinal fluid are possible mechanisms for the expansion of cystic volume [2]. The usual location of extradural cysts inside the spinal canal is the posterior aspect of the dural sac; however, they can be found anywhere inside it [6]. Symptoms can be intermittent and also exacerbated by Valsalva manoeuvres or changes in position [9]. Remissions and fluctuations of symptoms have been reported in 30% of patients as a consequence of changes of pressure in the spinal canal secondary to changes in the hydrostatic pressure of the cerebral spinal fluid [3,6]. Doita et al. [10] used cinematic magnetic resonance to study the pathomechanism of fluctuating symptoms during Valsalva manoeuvres in patients with these cysts and concluded that changes in pressure in the extradural space and in the arachnoid cyst might cause symptoms of intermittent compression of the spinal cord. Symptoms that are often asymmetric included weakness, numbness, neuropathic pain, myelopathy and incontinence, and spastic or flaccid paraparesis [11]. Very rarely, the cyst can be accompanied by herniation of the spinal cord inside it, and syringomyelia [12]. Histopathologically, these cysts comprise layers of fibrocollagenous tissue with disseminated meningothelial cells [6]. Magnetic resonance imaging has high sensitivity and specificity in diagnosing lesions that contain cerebrospinal fluid [6,13,14]. The differential diagnoses include: Tarlov or perineural cyst, ganglion cyst, synovial cyst, meningeal diverticula next to the nerve roots, meningocele, dermoid cyst and neuroma with cystic changes [15]. Nabors et al. [16]. classified them into 3 categories in 1988. Type I: extradural cyst without nerve tissue; Type IA: extradural meningeal or extradural spinal arachnoid cyst; Type IB: sacral meningocele; Type II: extradural cyst containing nerve tissue, Tarlov or perineural cyst; Type III: spinal cyst. The management can vary according to the patient's symptoms [17]. There are many surgical options reported in the literature: bone decompression, subtotal or total resection of the cyst, drainage and marsupialisation of the cyst [18]. In asymptomatic patients, conservative management with subsequent clinical observation is recommended [17]. The cornerstone of management in patients with moderate to severe neurological deterioration is complete removal of the cyst followed by obliteration of the pedicle and complete repair of the dural defect, in order to prevent the valve mechanism and recurrence of the cyst [19]. However, the site of communication of the cyst between the extradural and subarachnoid space is not always found during surgery [20]. Some other authors such as Kunz et al. [21]. Conclude that patients with mild symptoms could be treated conservatively, while patients with moderate to severe neurological deficit might benefit from surgical treatment.

Conclusions

Surgical treatment of spinal arachnoid cysts should not only provide neural decompression but also prevent cyst refilling. Simple aspiration or shunting is inadequate and not recommended. Choice of treatment should be radical cyst removal and dura repair and duraplaty with facia at the site of communicationof arachnoid cyst.

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