Introduction

Various types of cystic lesions are confronted in the spinal canal and are classified in respect of their relationship to the adjacent structures and nature of the cyst content. Amongst them, cerebrospinal fluid (CSF)-filled meningeal cysts constitute for the majority of the cases [11, 18] and are called as “arachnoid cysts,” “diverticula,” [5, 9] or “pouches” [9]. They may be located in the intradural, extradural, or perineural spaces. Depending on the location, size, and mechanism of origin, the clinical course varies between asymptomatic, incidentally-diagnosed cases [17] to severe myelopathy [11]. When they communicate freely with subarachnoid space, fluctuating symptoms related to changes in CSF pressure may occur [8, 9]. Although arachnoid cysts can be observed anywhere along the length of the spinal canal, middle, and lower thoracic regions constitute for the most frequently involved areas. When progressive neurological findings exist, surgical treatment is warranted. Surgical techniques include excision, fenestration, or placement of a cysto - subarachnoid shunt [6, 11], as well as some less invasive techniques that have been proposed recently [16].

We present a case with a giant cervicothoracic extradural arachnoid cyst and its surgical management. Several features of the cyst, such as its size, location, and clinical features make the case extremely unusual.

Case Report

A 57-year-old male was referred with symptoms of difficulty in walking, weakness, numbness, and thinning of arms and legs, and impotency, which had all begun two years before. The patient had suffered a serious traffic accident when he was 10 years old, and has had a contracture of his right hand since then. Neurological examination revealed spastic quadripareisis predominantly on the left side. His radiological examination showed widening of the cervical spinal canal and left neural foramina due to a cerebrospinal fluid - filled extradural cyst that extended from C2 to T2 level. The cyst was located left anterolaterally, compressing the spinal cord. Through a C4–T2 laminotomy, the cyst was excised totally and the dural defect was repaired. Several features of the reported case, such as cyst size, location, and clinical features make it extremely unusual. The case is discussed in light of the relevant literature.

Keywords: Arachnoid cysts · Magnetic resonance imaging · Spinal cord compression · Spine · Surgical treatment
Muscle tonus of bilateral lower extremities was increased. The anal sphincter tonus was weak, and the patient reported an intermittent sphincter control defect.

Cervical radiographs showed widening of the left neural foramina and the spinal canal. Magnetic resonance imaging (MRI) revealed an extradural cystic mass extending from C2 to T2 vertebral bodies, located left anterolaterally, compressing the spinal cord (Figs. 1a, b, c and 2). It had the same CSF intensity on T1 and T2-weighted images and showed no contrast enhancement. The lesion had caused a concavity in the related vertebral bodies and widening of the neural foramina on the left side. Electrophysiological tests reported chronic severe neurogenic involvement and a delay in the somato-sensory evoked potential responses in C6 - T1 innervated muscles.

A C4 – T2 laminotomy was performed. An extradural cystic lesion, full of CSF and having a thin and transparent membrane, was observed compressing the dural sac. The lesion had caused widening of C5 – C7 foramina and there was an active flow of CSF to the cyst through the defects located at the exiting points of these three roots. The cyst wall was excised and the dural defects were repaired using muscle pieces and sutures. Despite some degree of the spinal cord and dural sac expansion, the epidural and subarachnoid spaces left enlarged, since the canal was widened. The epidural space was left unfilled for the fear of compression. The samples taken from the cyst wall were reported as fibrohyalinized meningeal tissues.

Postoperative course was uneventful. Although the spasticity gradually improved during two years of follow-up, muscle strength and other neurological findings did not change.

Discussion

Spinal arachnoid cyst is a rare entity. There are various classifications based on etiology, histopathology, or localization of the cysts. Many authors classify spinal meningeal cysts as both intradural and extradural.

Extradural cysts include arachnoid cysts, synovial cysts, ganglia, cysts of ligamentum flavum, and cysts originating from the intervertebral discs. Intradural cysts include arachnoid cysts, enterogenous (endodermal, neuroenteric) cysts, and ependymal cysts [3, 6, 7, 10, 12 – 15, 18]. The pathogenesis of spinal arachnoid cyst is not clear and probably not homogenous. Congenital [13], traumatic [1], or inflammatory [15] mechanisms were proposed. It was documented that congenital asymptomatic cysts could enlarge due to trauma and become symptomatic [3]. The size of arachnoid cysts may vary in relation to underlying mechanisms. For example, the arachnoid cysts related to arachnoiditis are usually localized and small [6, 18]. Pulsatile CSF dynamics [9], osmotic gradient between the subarachnoid space and cyst [7], and the valve-like mechanism between the cyst and subarachnoid space [10] may play an important role in the enlargement of spinal extradural arachnoid cysts. Doita et al. [4] demonstrated that increased intra-abdominal and intrathoracic pressure may lead to size changes of the spinal arachnoid cyst.

Fig. 1 T2-weighted median (a), slightly left paramedian (b), and left paramedian (c) sagittal magnetic resonance images of the cervical region shows a cystic mass extending from C2 to T2 vertebral bodies, located left anterolaterally, enlarging left neural foramina (arrows). (Arrowheads spinal cord)
A review demonstrated that arachnoid cysts involving the cervical region are extremely rare, that they never exceed three vertebral bodies in length, and that none extended into the thoracic region [6], except the case reported by Safriel et al. [14]. That case was an intradural arachnoid cyst situated anteriorly to the spinal canal extending between C5 and T3 levels. The presented cyst was extradural, located at the cervicothoracic region, anterolateral to the spinal cord, and involved eight vertebral bodies extending from C2 to T2 level. To the authors' knowledge, there is no extradural arachnoid cyst reported in the literature matching that size. The patient had no disposing factor other than trauma. He had suffered a serious traffic accident when he was a child and consequently had a contracture in his left upper extremity. Although there was an excessively long duration (45 years) between trauma and appearance of new symptoms attributable to the cyst, the concordance of the former neurological deficit and location of the cyst convinced us that the etiology of the cyst might have been the childhood trauma. During the surgery, we did not observe any laminar signs that indicated a sustained compression fracture on the posterior elements. Thus, stretching and partial avulsion of the roots may have caused arachnoid and dural tearing during the childhood trauma, causing subsequent collection of the CSF at the epidural space, communicating freely both subarachnoid space and dural sleeves of the roots. Indeed, the occurrence of multiple dural defects, through which the CSF exits, is very rare. This point makes an extra implication on the case and further supports the avulsion theory as the cause of multiple dural tearings, since the neurological deficit due to childhood trauma is concordant with multiple root involvement. Long-standing progressive enlargement of the cyst explains radiological signs such as eroded vertebral bodies and enlarged intervertebral foramina.

Conclusion

The pathogenesis, etiology, and treatment of the spinal arachnoid cyst have not been well established because of its rarity. Symptomatic cases require surgical intervention. For long segment involvements, hemilaminectomy or laminotomy, as was performed in the current case is preferable to laminectomy in order to prevent postoperative deformity. In surgery, dural defect should be closed after excision. Neurological recovery depends on the size of the cyst and degree and duration of the spinal cord compression. A long-standing spastic myelopathy is unlikely to have a significant improvement [2], as we observed in the current case. Except spasticity, the patient had no improvement on his neurological findings. Thus, in delayed cases, surgery may be taken and should be offered as a prophylactic intervention to prevent further impairment, rather than a curative one.

In conclusion, the case presented here is a very unusual occurrence of this rare entity. Although probably traumatic, it had extremely delayed clinical presentation, its location was unusual, and as an extradural cervicothoracic arachnoid cyst, it was the largest amongst those reported.

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