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Short Illustrated Review

Spinal extradural arachnoid cyst

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Summary

Spinal extradural arachnoid cysts are rare expanding lesions in the spinal canal. They usually present with progressive signs and symptoms caused by spinal cord compression if they enlarge. A comprehensive review about spinal extradural arachnoid cyst is made including the author's own case of a 59-year-old woman with a 6-month history of progressive back pain radiating to both legs. Key points concerning the possible pathogenesis including symptomatology, diagnosis, and the implications for treatment are highlighted. Surgical treatment is curative and this rare clinical entity should be considered in the differential diagnosis of spinal extradural lesions.

Keywords: Spinal extradural arachnoid cyst; arachnoid cyst; spinal cord compression; surgical treatment; MRI.

Introduction

Extradural arachnoid cysts of the spine are thought to be rare findings and uncommon causes of myelopathy secondary to spinal cord compression. These cysts are reported to develop in any location, although they are most commonly found in the thoracic spine [3, 8, 26]. They are more common in males, and their peak incidence is in the patient's second decade of life [8, 21, 27]. They are usually found posterior to the spinal cord but have been described in the posterolateral and anterior positions as well [3]. The origin of extradural arachnoid cysts and their enlargement to form compressive lesions has been the subject of much conjecture.

The causes of spinal extradural arachnoid cysts remain unclear. It is reported that they are extradural outpouchings of arachnoid that communicate with the intraspinal subarachnoid space through a small defect in the dura [19]. They give rise to fluctuating symptoms as a consequence of internal changes in pressure resulting from changes in the hydrostatic pressure of the cerebrospinal fluid caused by physical exertion, coughing, sneezing, and straining [3, 8, 26].

So far, no systematic reviews have been published about spinal extradural arachnoid cysts, and comprehensive reviews, covering all relevant aspects, are needed to better understand these cysts. The present paper aims at being the major part of such a comprehensive review and we address the possible pathogenesis including symptomatology, diagnosis, and the implications for treatment.

Literature review

We searched for relevant literature on PubMed. The following search terms were used: spinal extradural arachnoid cyst. The search was limited to studies on humans, published in English or German and in the period from 1963 to March 2005. This resulted in 59 titles; among them, we found 3 clinical papers and 17 case reports (Table 1). References of retrieved articles and of relevant overview articles were checked to identify additional studies.

Case reports

Ersahin *et al.* presented a 16-year-old boy with a spinal extradural arachnoid cyst [6]. An extradural arachnoid diverticulum extending from T10 to L1 was excised totally by hemilaminectomy. Surgery caused prompt improvement of the neurological deficit. Liu *et al.* reported a 24-year-old man with an extradural arachnoid cyst from T12 to L4 [17]. The patient presented with low back pain, and underwent recapping laminectomy from T12 to L2 and cyst extirpation. Three months

Table 1. *A summary of previously reported cases with spinal extradural arachnoid cysts*

Case	Authors & year	Sex, age (yrs)	No. of cases	Level	Symptoms	Symptom duration	Operation	Outcome
1	Kim *et al.* [13], 1986	M/16	1	T7–9	fatigability of lower extremities	9 mos	laminectomy excision of the cyst	complete recovery
2	Winer *et al.* [29], 1989	M/77	1	T10–L1	pain in the right heel	10 yrs	laminectomy excision of the cyst	considerable improvement in the right heel pain
3	Congia *et al.* [4], 1992	M/15	1	T10–12	spastic paraparesis	2 yrs	laminectomy excision of the cyst	incomplete recovery
4	Ersahin *et al.* [6], 1999	M/16	1	T10–L1	back pain radiating leg pain	1 mos	hemilaminectomy excision of the cyst	complete recovery
5	Rimmelin *et al.* [27], 1997		2					
		M/24		T12–L3	pain & dysesthesia in the leg	1 yr	surgery not performed	–
		M/14		T2–9	walking difficulty & spastic paraparesis	not described	laminectomy excision of the cyst	incomplete recovery
6	Myles *et al.* [21], 1999	F/9	1	T1–S1: multiple, 6 cysts	progressive gait disturbance	2 wks	T1–L2 laminoplasty excision of the cyst cysts within lumbar & sacral canal: surgery not performed	incomplete recovery
7	Prevo *et al.* [25], 1999	F/15	1	T11–L1	spastic paraparesis	3 yrs	laminectomy excision of the cyst	complete recovery
8	Kim *et al.* [12], 1999	M/36	1	S2–4	coccygeal pain dysuria	several months	oibliterating fistulous channel	complete recovery
9	Martin [18], 2000	M/31	1	T8–9	back pain sciatica	6 mos	laminectomy excision of the cyst	complete recovery
10	Ido *et al.* [11], 2002	F/24	1	L1–2	back pain leg pain	3 mos	transforaminal approach & excision of the cyst	complete recovery
11	Muthukumar [20], 2002	M/25	1	S2–3	low back pain perineal pain	2 yrs	S1–4 laminectomy	complete recovery
12	Doita *et al.* [5], 2003	F/31	1	T11–L1	thoracic back pain & leg stiffness	3 mos	T11–L1 laminoplasty excision of the cyst	complete recovery
13	Liu *et al.* [17], 2004	M/24	1	T12–L4	low back pain	1 yrs	T12–L2 laminoplasty excision of the cyst	complete recovery
14	Chang *et al.* [2], 2004		2					
		F/13		T12–L2	lower extremities claudication	6 mos	limited L1 laminectomy	complete recovery
		F/12		2 cysts L1–2	spastic paraparesis	3 mos	limited L1 laminectomy	complete recovery
15	Neo *et al.* [23], 2004	F/29	1	T11–L3	leg muscle weakness	9 yrs	T12–L1 fenestration selective closure of the dural defect using clips	complete recovery
16	Nakagawa *et al.* [22], 2004	M/12	1	lower thoracic to sacral	myelopathy	4 yrs	closing dural defect after partial resection of the cyst	complete recovery
17	Novak *et al.* [24], 2005	F/15	1	T3–6	progressive paraparesis	2 yrs	laminectomy resection of the cyst	complete recovery
18	Choi *et al.*, 2005 (illustrative case)	F/59	1	T11–12	back pain radiating leg pain	6 mos	laminectomy	complete recovery

postoperatively, low back pain resolved completely. Chang *et al.* [2] reported on the experience of two children with spinal cord compression caused by three extradural arachnoid cysts. In both patients, plain radiographs demonstrated widening of the interpedicular distance, which suggested progressive widening of the spinal bony canal. They concluded radical cyst removal and dura defect closure were the surgical intervention of choice in patients with symptomatic extradural arachnoid cyst. Myles *et al.* [21] reported on the treatment of multiple extradural spinal arachnoid cysts in a 9-year-old child affecting the thoracic, lumbar, and sacral spine. A T1–L2 laminoplasty was performed and cysts at the thoracic spine were excised. The cysts within the lumbar and sacral canal were not surgically approached at that time because the patient had presented with myelopathy. They placed emphasis on an underlying defect in the dura of the spinal canal that which predisposed to the formation of cysts. Winer *et al.* [29] reported a case, who presented as a very slowly progressive conus lesion in a 77-year-old man. After surgical excision of the cyst, the patient showed a considerable symptomatic improvement. Muthukumar [20] gave a report on a 25-year-old man with a sacral extradural arachnoid cyst causing low back and perineal pain. He underwent sacral laminectomy with opening of the arachnoid cyst and ligation of the fistulous tract and there was complete clinical recovery postoperatively. He stressed that this entity should be considered in the differential diagnosis of low back and perineal pain, and that surgical treatment was curative. Prevo *et al.* [25] presented a 15-year-old girl with a slowly progressive spastic paraparesis, and concluded that an extended extradural spinal arachnoid cyst could be an unusual cause of progressive spastic paraparesis. Kim *et al.* [12] reported a case of sacral extradural arachnoid cyst associated with concomitant lumbar intradural arachnoid cyst in a 35-year-old male. The patient presented with a history of severe sacrococcygeal pain, constipation, and dysuria for several months. A favorable outcome could be achieved by decompression of the cyst, obliteration of the fistulous channel between the cyst and the thecal sac, and fenestration of the arachnoid cyst into the subarachnoid space. Martin [18] reported a case of herniation of the thoracic spinal cord into the mouth of an extradural arachnoid cyst in a 31-year-old man, who was treated successfully by surgical release of the neck of the hernia. Ido *et al.* [11] reported on the effectiveness of a transforaminal surgical procedure for spinal extradural arachnoid cyst. They resected a large extradural cystic lesion close to the L1 nerve root sleeve following closure of the ostium through a transforaminal approach. Novak *et al.* [24] described the case of a fifteen-year-old girl with progressive paraparesis of the lower limbs which was caused by an intraspinal extradural dorsal arachnoid cyst from T3 to T6. Diagnosis was established by MRI and MRI myelography demonstraing CSF-like content of the cyst. The patient was successfully treated with ligation of the pedicle of the cyst by laminoplasty.

Clinical studies

Krings *et al.* [14] presented a retrospective clinical investigation on the diagnostic and therapeutic management of spinal arachnoid cysts. Of seven patients with symptomatic spinal arachnoid cyst, five patients were identified with extradural arachnoid cysts. Postoperative outcome was favourable in those patients without preoperative cord damage. They stressed that surgical removal was significant in terms of neural decompression and prevention of refilling of the cyst, which could be best accomplished by complete resection of the cyst and closure of the communication between cyst and subarachnoid space. Kunz *et al.* [16] gave an overview on the treatment of the surgically-treated eight patients with lumbosacral extradural arachnoid cysts. Only three of the patients experienced postoperative relief of pain, but none was symptom free. The results of conservative treatment were nearly the same as those of operative treatment. They concluded that patients with slight and not clearly related uncharacteristic symptoms should be excluded from surgery, whereas patients with a short pain history and a clear neurological deficit might profit most from surgery. Kulkarni *et al.* [15] presented an analysis of the management of 7 cases of extradural arachnoid cyst. There were two cysts in the thoracic region, four in the thoracolumbar region and one in the cervicothoracic region. Radical resection of the walls led to a lasting cure from extradural arachnoid cyst and there was no recurrence of the cyst during the period of follow-up. They reported the site of communication of the cyst contents with subarachnoid cerebrospinal fluid (CSF) could not be identified during surgery.

Focus on imaging

Most radiological papers reported magnetic resonance imaging (MRI) had great sensitivity to and specificity for extradural arachnoid cyst and seemed to be the diagnostic procedure of choice [4, 13, 27]. With all sequences, the signal within the lesion is iso-intense to CSF. The varying degrees of cord atrophy and the extent of myelomalacia can be assessed by MRI, and the MRI may, therefore, be helpful in predicting neurological outcomes. They also pointed out that both myelography and computed tomography (CT) myelography had the major advantage in terms of demonstrating the location of the communication in the dura with the intraspinal subarachnoid space and the determination of communication between different cysts. Recently, kinematic MRI studies have been performed to define the physiological or pathological changes that occur in the subarachnoid space. Doita *et al.* [5] reported on the use of kinematic MRI to clarify the pathomechanism of fluctuating symptoms caused by an extradural arachnoid cyst during straining. They concluded that the pressure changes that occurred in the extradural space as well as in the arachnoid cyst might cause spinal cord compression, and this should be considered as an alternative explanation for intermittent exacerbation of symptoms in patients with extradural arachnoid cyst. Nakagawa *et al.* [22] reported on the usefulness of constructive interference in steady state (CISS) imaging for the diagnosis and treatment of a large extradural spinal arachnoid cyst.

They used 3D constructive interference in steady state (CISS) MRI to clearly demonstrate the pedicle of the cyst.

Focus on new treatment

Neo *et al.* [23] reported a giant spinal extradural arachnoid cyst treated by selective closure of the dural defect, using kinematic MRI to detect the communication site. Kinematic MRI showed a pulsating flow voiding in 29-year-old woman with an extradural arachnoid cyst from T11 to L3, suggesting the location of the communication site. Fenestration of the T12–L1 region was performed and a small dural rent was found on the left side of level L1. This was closed using small clips. They concluded selective closure of the dural defect based on kinematic MRI would be useful for treating extradural arachnoid cysts.

Illustrative case

A 59-year-old woman presented with a 6-month history of progressive back pain radiating to both legs along the posterior aspect of the thigh. The leg pain got worse three weeks before admission. The patient reported numbness in both thighs posteriorly, and denied any bowel or bladder symptoms. On clinical examination, the motor strength and deep tendon reflexes in the legs were normal. Her sensation showed hypesthesia below the level of T10, bilaterally.

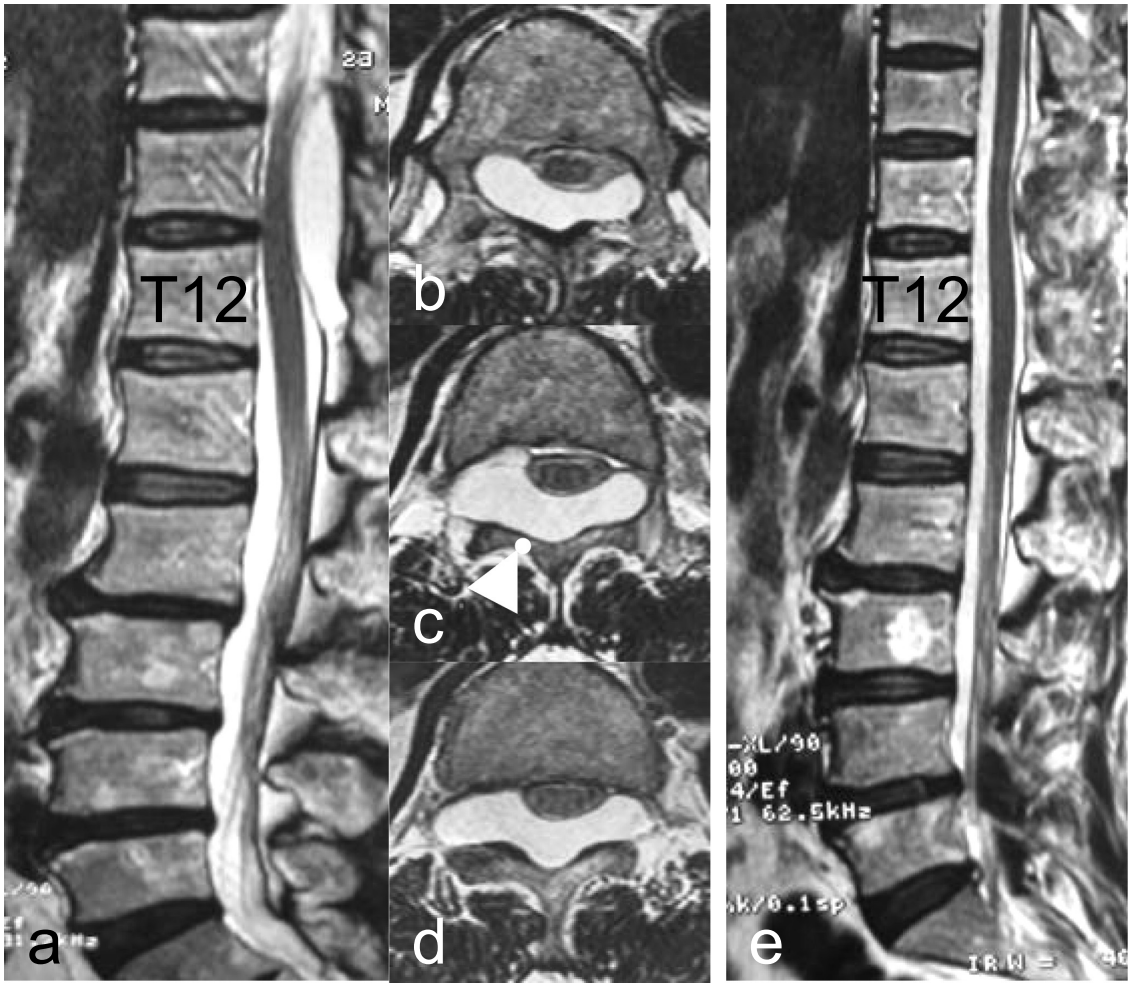

Fig. 1. Spinal MRI demonstrates a cystic lesion with a well-demarcated and lobulated shape at the T11–T12 level. The cystic lesion is iso-intense compared with CSF hyperintense on T2-weighted images (a). Axial MRI on T2-weighted images, which are displayed in sequence from rostral (b) to caudal (d), reveals that the cyst is mainly in the posterior extradural space with extension into the neural foramina bilaterally. There is also thinning of the laminae in the upper part of the cystic lesion (arrow head). Sagittal T2-weighted MRI obtained 7 days postoperatively demonstrates that the cyst has disappeared completely (e)

MRI demonstrated a cystic lesion with a well-demarcated and lobulated shape in the posterior extradural space at the T11–T12 level with extension into the neural foramina bilaterally (Fig. 1). There was also thinning of the laminae in the upper part of the cystic lesion (Fig. 1c). The cystic lesion was iso-intense compared with CSF on T1 & T2-weighted images. The lesion showed no enhancement after gadolinium was administered.

The patient underwent a total laminectomy centered on T11, including the majority of T12 and the inferior portion of T10. The laminae at the T11 were noted to be markedly thin and flattened. After removing the laminae at the involved level, a large, thin-walled cyst containing CSF was encountered. The walls of the cyst appeared to be formed by a thickened arachnoid membrane, and was easily dissected from the dura, which had been pushed forward by the cyst. The cyst was found to arise from a small neck protruding through a defect in the right lateral aspect of the dura at the level T11. The neck was obliterated and the dura was repaired. The spinal cord appeared to be completely decompressed after the cystic mass was removed.

Pathological examination of the cyst wall revealed that it was composed of fibrocollagenous layers with scattered meningothelial cells. There was a single-cell lining (Fig. 2). A diagnosis of arachnoid cyst was made.

Postoperatively, the patient experienced marked relief of pain and did not have hypesthesia below the level of T10. MRI obtained 7 days after surgery demonstrated that the cyst had disappeared completely (Fig. 1e). One year after surgery, the patient was neurologically normal without any deficits.

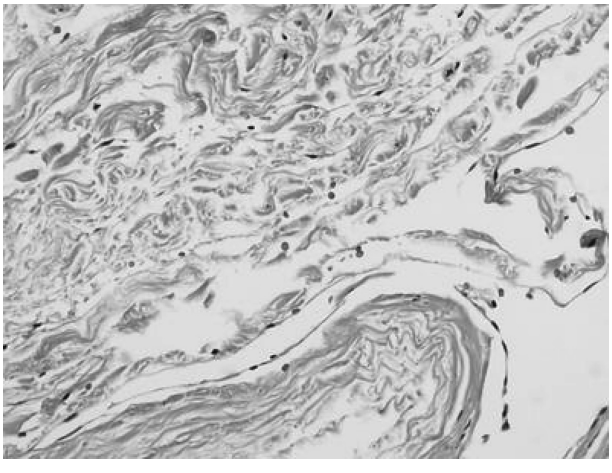

Fig. 2. Photomicrograph of the cyst wall demonstrates fibrocollagenous layers with a single-cell lining. H & E, original magnification 200

Discussion

Spinal extradural arachnoid cysts are most common in the mid to low thoracic spine and usually located in the posterior aspect of the thoracic or lumbar spine displacing the spinal cord anteriorly [2, 3, 26].

Pathogenesis

The causes of arachnoid cysts are unclear. They are known to be associated with trauma, surgery, arachnoiditis, and neural tube defects [10, 21, 26]. In many cases, there is no underlying cause and most of non-traumatic spinal extradural arachnoid cysts are thought to be congenital [20, 21]. Therefore, they are thought to arise from a congenital dural defect which allows the arachnoid membrane to herniate through the adjacent dura mater [7, 19, 27]. In the illustrative case, the patient had no history of trauma, surgery, or evidence of arachnoiditis demonstrated on imaging studies. The authors think the bone changes (thinning of the laminae) observed on MRI and at the time of surgery suggest a long-standing lesion with a congenital origin.

How the spinal cord is affected by such cysts is unclear, but several theories have been proposed on the pathogenetic mechanism [8, 28]: 1) a one-way valve causes intermittent increased pressure within the extradural cyst, thereby compressing the spinal cord; 2) a hyperosmolar collection of fluid within the cyst causes free water to enter, resulting in an enlargement of the cyst, thereby causing compression; 3) the cyst wall secretes fluid and it expands gradually due to lack of communication with the subarachnoid space. A number of cases reported in the literature have speculated that the valve-like mechanism with intermittent surges in CSF pressure explains the expansion of the cyst and spinal cord compression [8, 19, 26, 28].

Symptoms are generally related to compression of the spinal cord or nerve roots [5]. The most common presenting symptoms and signs are pain and progressive spastic or flaccid paraparesis, which are often asymmetrical. The symptoms are fluctuating with remissions and exacerbation. The intermittent exacerbation of symptoms has been explained by most authors as occurring because the inflated cyst causes some degree of spinal cord compression, when CSF pressure is temporarily raised and fluid enters the cyst on straining and coughing [8, 19, 28].

Valsalva manoeuvres which occur during daily activities might increase the volume of the cyst [20]. Increase in pressure and volume of the cyst during erect posture

and during Valsalva manoeuvres might produce the traction on the nerve roots and spinal cord by the arachnoid cyst, and might be responsible for the sensory and motor disturbances that are often seen in these patients.

Diagnosis

Different imaging techniques can be used for diagnosing arachnoid cyst. MRI has great sensitivity to and specificity for CSF containing lesions and is reported to be helpful in assessing the extent of the lesion [4, 13, 27]. With all sequences, the signal within the lesion is iso-intense to CSF. In the illustrative case, MRI was helpful in diagnosing the lesion, revealing iso-intensity to CSF in both T1- and T2-weighted sequences. A thin wall, septations within the cyst can be visualised. With intravenous contrast enhancement, an exact differentiation between cystic tumours, synovial cysts and arachnoid cysts can be made.

Plain x-ray films have not proved useful in the diagnosis of arachnoid cyst [1, 14]. Only indirect signs attributable to the mass effect of large cysts such as an enlarged spinal canal, bony erosions of the spine, slender pedicles, widened foramina or an increased interpedicular distance can be visualised [13]. Both myelography and computed tomography (CT) myelography may demonstrate the location of the communication in the dura with the intraspinal subarachnoid space and the determination of communication between different cysts [4, 13]. Because of changes in posture, arachnoid cyst can be missed on myelographic studies unless the patient is examined in the prone position. Additionally, delayed CT myelography can show differential emptying of cysts and allow the precise location of the dural defects to be defined [13, 14]. MRI is helpful in assessing the extent of the lesion; however, it is much less useful in delineating the neck of the cyst or determining where it lies with respect to the dura [21].

Kinematic MRI is a new technique that is able to visualise fluid and tissue movement. Doita *et al.* [5] tried to demonstrate the mechanism of spinal cord compression by a spinal extradural arachnoid cyst. This technique can be used for detecting a dural defect, which makes the cyst communicate with the dural sac, showing a pulsating turbulent flow voiding.

Treatment

Treatment of extradural arachnoid cysts is complete surgical removal with an excellent prognosis regardless of degree of cyst size [14, 15, 17, 20, 24, 25]. Novak *et al.* [24] achived a complete recovery for a patient with an extradural arachnoid cyst from T3 to T6 following en block resection of the cyst and ligation of the pedicle of the cyst by laminoplasty. If the cyst is adherent to the spinal cord, marsupialization has been reported to successfully achieve relief from symptoms [8]. Evacuation or aspiration of the cyst will result in only temporary improvement of symptoms [9].

The authors think a dural defect or stalk can not always be found during surgery. Kulkarni *et al.* reported that the site of communication of the cyst contents with subarachnoid CSF could not be found during the surgery in their series [15]. Upon surgery, an individually tailored approach with a careful microsurgical operating technique without impairment of the neural tissue seems to be the approach of choice.

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Comment

The authors present an interesting and well written short review of a rare but important entity of spinal space occupying lesions. A comprehensive review of the literature concerning spinal extradural arachnoid cysts focussing on special imaging techniques and new treatment options, is provided.

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