Scholarly Research Exchange

Volume 2009 • Article ID 452627 • doi:10.3814/2009/452627

Case Report

Idiopathic Intradural Extramedullary Arachnoid Cyst and Associated Syringomyelia

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Received 11 December 2008; Revised 6 May 2009; Accepted 11 May 2009

We report a case and discuss clinical features and radiological differential diagnosis of dorsal idiopathic extramedullary arachnoid cyst associated to a caudal syringomyelic cavitation. The aim of this article is to review the current literature on a rare topic as idiopathic intradural extramedullary arachnoid cyst with associated syringomyelia. We emphasize the fact that correct diagnosis and adequate treatment, which we believe is microsurgical fenestration of the cyst into the subarachnoid space, may lead to disappearance of syringomyelia and complete resolution of preoperative spinal cord compression symptoms.

1. Introduction

Idiopathic intradural extramedullary arachnoid cysts are a rare cause of spinal cord compression, and they are even more rarely described in association to a syringomyelic cavity. We report the case of an arachnoid cyst associated to a syringomyelic cavitation, in absence of trauma, dural defects, or other predisposing factors.

2. Case Report

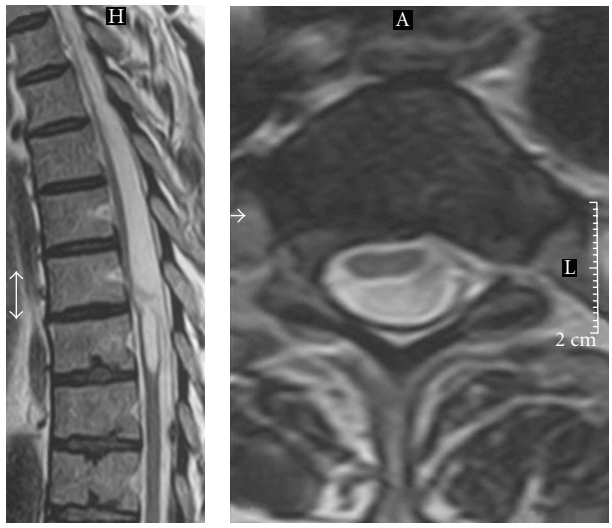
A 54-years-old man presented to our Institution for gait disturbances since the last 3 years with a slowly worsening clinical course. At admission, he could only walk with a walker, and complained of paresthesias in both feet, and ankles, associated to urinary incontinence. He reported no antecedent history of surgery, trauma, or infections. Neurological examination revealed a marked spastic paraparesis with increased deep tendon reflexes in lower limbs, subclonic Achilles tendon reflexes, and bilateral Babinski sign. A level of hypoesthesia at about 2 cm under the mammillary line was detected. MRI of the spine showed a cystic intradural extramedullary lesion extending from D3 to D7 (Figures 1(a) and 1(b)) with anterior dislocation of the spinal cord and an associated syryngomielic dilatation at D8. No contrast enhancement after Gadolinium administration was observed. Sensory and motor evoked potentials revealed a corticospinal pathway dysfunction.

The patient underwent a D6–D8 laminectomy, with microsurgical fenestration of the cyst into the subarachnoid space; intraoperatively, cerebrospinal fluid flow from the cyst to the spinal subarachnoid spaces was observed. Careful dissection of any arachnoidal adhesions tethering the spinal cord was also performed.

The patient experienced a marked postoperative improvement in neurological performances, with autonomous deambulation. An MRI exam of the thoracic spine 6 days postoperatively showed a good decompression of spinal cord with initial reduction in syrinx size. An MRI fifteen months later documented total disappearance of the syringomyelic cavitation (Figures 2(a) and 2(b)). After a 4, 5 years follow-up period, the patient is autonomous in all his activities.

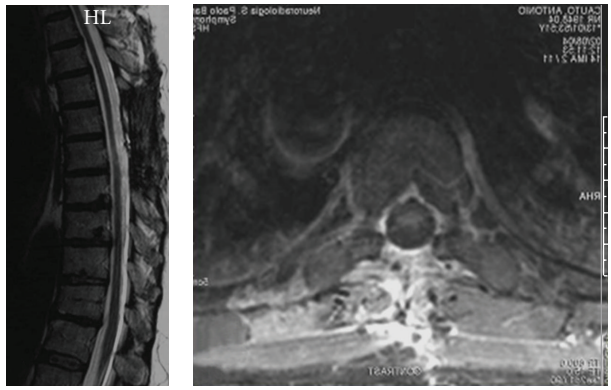
3. Discussion

Intradural extramedullary arachnoidal cysts are a rare cause of spinal cord or radicular compression and can be either idiopathic or the result of various events. As for their location, the majority of intradural spinal arachnoid cysts occur in the thoracic region (80%); 15% of cases are described in the cervical region and 5% in the lumbar region. Craniocaudal extension of these cysts may involve up to 17



(a) (b)

Figure 1: Preoperative T2 weighted MR images, sagittal (a), and axial (b) sections: the spinal cord is displaced anteriorly by the arachnoidal cyst. An associated caudal syringomyelic cavity at D8 is evident.



(a) (b)

Figure 2: Postoperative MRI, performed 15 months after surgery, sagittal (a), and axial (b) sections: the spinal cord is totally decompressed and there is no evidence of arachnoidal cyst or syringomyelic cavity.

vertebral levels [1]. Most are dorsal to the spinal cord, and a minor percentage, as in our case, is ventrally located [1–4]. Intradural extramedullary arachnoid cysts are less common than extradural ones [2, 4] and are twice as frequent in males as in females [5].

3.1. Patogenesis. Spinal arachnoid cysts can be congenital in origin or secondary to trauma, infection, subarachnoid haemorrhage, meningeal infection, and other insults that cause inflammation and subarachnoid adhesions [1]. Trauma, arachnoiditis, and other inflammatory reactions have been implicated as causal factors involved in their formation [6]; all sharing the common pathogenetic mechanism of adhesive arachnoiditis [1].

Arachnoid cysts can be idiopathic too, and various theories have been proposed to explain their formation. Perret et al. [7] postulated an origin from the septum posticum of Schwalbe, which divides the dorsal subarachnoid space in the midline from the cervical to lumbar region; this theory may account for cysts, but it does not explain those in a ventral location. Teng and Rudner [8] proposed that normal variations of intraspinal CSF pressure lead to cyst formation through dilatation of low-resistance areas within the arachnoid; subsequent cyst enlargement is probably to be related to a one-way valve mechanism at the neck of the diverticulum. Fortuna et al. suggested [9] that all types of congenital arachnoid diverticula result from hypertrophy, proliferation, and dilatation of arachnoid granulations; then if the dilatation is confined by a resistant dura, an intradural cyst would result, whereas a weakened or deficient dura would allow herniation of the arachnoid into the epidural space to form an extradural cyst.

3.2. Clinical Features. Clinical presentation depends on cyst location, as anterior cysts are more likely to cause weakness and myelopathy, while dorsal cysts present more commonly with neuropathic pain and numbness; local compression of spinal tracts by the cyst may explain the difference in symptoms. Alternatively, ventral lesions may produce vascular compression in the distribution of the anterior spinal artery and thus cause weakness or myelopathy.

The development of symptoms is usually slow, and we believe that marked recovery of neurological deficits after surgery is possible even if the patients symptoms date from a long time as in our case. Radiological differential diagnosis of spinal cord cystic lesions includes neoplasms as cystic ependymoma, parasitic foci, radicular cysts, and arachnoid cysts. MRI is the diagnostic procedure of choice for both diagnosis with preoperative surgical planning and postoperative follow-up, as it adequately delineates their location in relation to the spinal cord and their craniocaudal extension [10]. Myelography and myelo-CT have demonstrated to be useful too, but a higher sensitivity of MRI as compared with myelo-CT in the detection of intradural arachnoid cysts has been reported because the latter has yielded false-negative results in some cases [11].

3.3. Associated Syringomyelia. Several anomalies have reported to be associated to arachnoid cysts, as neural tube defects in children [12] and various vertebral anomalies in adults, as Klippel-Feil anomaly [6] or kyphoscoliosis [1], or rarely spinal cord herniation [13]. In some rare cases [4, 14, 15] as the one we report, a syryngomielic cavitation can be associated to an idiopathic arachnoid cyst.

Syrinx formation is usually related to posttraumatic conditions, hydrodynamic changes associated with arachnoiditis, or developmental abnormalities and spinal cord

compression resulting in spinal cord ischemia (myelomalacia). Spinal cord neoplasms are often (25%–60%) associated with a syringeal cavity too, and they are usually intramedullary tumours; extramedullary lesions rarely cause syrinx formation, and when they do, association with a more cranial rather than caudal syrinx is usually reported [8]. As for syringomyelia in association with idiopathic arachnoid cyst, the proposed mechanism to explain their formation is that of an alteration in CSF flow within the spinal subarachnoid space, leading to the formation of syrinx cavities [15, 16]. The main predisposing condition has been individuated in a dural defect: arachnoid herniation may occur through the defect into the extradural space, and CSF would flow freely in and out. In time, as the spinal cord develops adhesions to the edges of the dural defect, CSF flow becomes impeded and constant CSF pulsations force the cord further through the defect. According to some authors syringomyelia is more likely to be associated with ventral (50%) than dorsal cysts (27%) [4].

In our patient the syrinx was dorsally located and caudal to the level of the arachnoid cyst; we think that the cyst induced hydrodynamic changes, and syrinx formation was related to an altered interstitial CSF backflow, rather than to a direct effect of spinal cord compression, with a one-way valve mechanism. To support that, in our experience cyst fenestration led to a restore of normal CSF circulation and subsequent syrinx resolution.

3.4. Treatment. The surgical procedure of choice is complete excision of the cyst whenever possible; simple aspiration of the cyst does not represent a definitive treatment, as this usually results in recurrence. The most common treatment option is represented by cyst fenestration [15].

4. Conclusions

We report the rare case of a patient harbouring an idiopathic intradural extramedullary arachnoid cyst, associated to a caudal syringomyelia causing spinal cord compression and progressive myelopathy. Microsurgical fenestration of the cyst into the subarachnoid space led to resolution of the syrinx and regression of symptoms.

References

[1] H. Bassiouni, A. Hunold, S. Asgari, U. Hübschen, H.-J. König, and D. Stolke, "Spinal intradural juxtamedullary cysts in the adult: surgical management and outcome," *Neurosurgery*, vol. 55, no. 6, pp. 1352–1360, 2004.

[2] R. Dulou, E. Blondet, G. Dutertre, J.-M. Delmas, F. De Soultrait, and P. Pernot, "Spinal cord compression by arachnoid cysts," *Neurochirurgie*, vol. 52, no. 4, pp. 381–386, 2006.

[3] H.-J. Lee and D.-Y. Cho, "Symptomatic spinal intradural arachnoid cysts in the pediatric age group: description of three new cases and review of the literature," *Pediatric Neurosurgery*, vol. 35, no. 4, pp. 181–187, 2001.

[4] M. Y. Wang, A. D. O. Levi, and B. A. Green, "Intradural spinal arachnoid cysts in adults," *Surgical Neurology*, vol. 60, no. 1, pp. 49–55, 2003.

[5] M. Bruneau, T. Duprez, D. Rommel, and C. Raftopoulos, "Surgical treatment of a syringomyelia associated with an idiopathic arachnoid malformation disclosed by preoperative MRI," *Surgical Neurology*, vol. 62, no. 6, pp. 552–555, 2004.

[6] R. K. Osenbach, J. C. Godersky, V. C. Traynelis, and R. D. Schelper, "Intradural extramedullary cysts of the spinal canal: clinical presentation, radiographic diagnosis, and surgical management," *Neurosurgery*, vol. 30, no. 1, pp. 35–42, 1992.

[7] G. Perret, D. Green, and J. Keller, "Diagnosis and treatment of intradural arachnoid cysts of the thoracic spine," *Radiology*, vol. 79, pp. 425–429, 1962.

[8] P. Teng and N. Rudner, "Multiple arachnoid diverticula," *Archives of Neurology*, vol. 2, no. 3, pp. 348–356, 1960.

[9] A. Fortuna, E. La Torre, and P. Ciappetta, "Arachnoid diverticula: a unitary approach to spinal cysts communicating with the subarachnoid space," *Acta Neurochirurgica*, vol. 39, no. 3-4, pp. 259–268, 1977.

[10] D. P. Beall, D. J. Googe, R. L. Emery, et al., "Extramedullary intradural spinal tumors: a pictorial review," *Current Problems in Diagnostic Radiology*, vol. 36, no. 5, pp. 185–198, 2007.

[11] J. L. Dietemann, F. M. M. de la Palavesa, B. Kastler, J. M. Wafter, and E. Buchheit, "Thoracic intradural arachnoid cyst: possible pitfalls with myelo-CT and MR," *Neuroradiology*, vol. 33, no. 1, pp. 90–91, 1991.

[12] C. H. Rabb, J. G. McComb, C. Raffel, and J. G. Kennedy, "Spinal arachnoid cysts in the pediatric age group: an association with neural tube defects," *Journal of Neurosurgery*, vol. 77, no. 3, pp. 369–372, 1992.

[13] I. J. Barrenechea, J. B. Lesser, A. L. Gidekel, L. Turjanski, and N. I. Perin, "Diagnosis and treatment of spinal cord herniation: a combined experience," *Journal of Neurosurgery: Spine*, vol. 5, no. 4, pp. 294–302, 2006.

[14] A. Takeuchi, K. Miyamoto, S. Sugiyama, M. Saitou, H. Hosoe, and K. Shimizu, "Spinal arachnoid cysts associated with syringomyelia: report of two cases and a review of the literature," *Journal of Spinal Disorders and Techniques*, vol. 16, no. 2, pp. 207–211, 2003.

[15] L. T. Holly and U. Batzdorf, "Syringomyelia associated with intradural arachnoid cysts," *Journal of Neurosurgery: Spine*, vol. 5, no. 2, pp. 111–116, 2006.

[16] P. W. A. Willems, W. M. van den Bergh, and W. P. Vandertop, "An arachnoid cyst presenting as an intramedullary tumour," *Journal of Neurology Neurosurgery and Psychiatry*, vol. 68, no. 4, pp. 508–510, 2000.