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SPINAL INTRAMEDULLARY ARACHNOID CYST: DIVERGENT OR INSURGENT LESION

Neurosurgery

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ABSTRACT

Spinal arachnoid cysts in general are not a common clinical entity. When encountered, they are usually seen in the extradural or the intradural-extramdeullary location. Intramedullary primary spinal arachnoid cysts are extremely rare and only few cases have been published in the medical literature.

We report a case of a primary thoracic intramedullary spinal arachnoid cyst in an adult male who was successfully operated at our center.

KEYWORDS

spinal arachnoid cyst; thoracic spine; intramedullary.

Introduction:

Arachnoid cysts arising in the spine are usually incidentally discovered on spinal MRIs [1]. Thoracic location is the most commonly encountered and the majority are asymptomatic and lie in an extradural or extramedullary location [1, 2]. When symptomatic, the symptoms are usually variable and non-specific hindering the diagnosis and treatment plan somehow challenging [2].

Intramedullary primary arachnoid cysts are considered as a rarity entity with poorly understood pathophysiology, and as a paucity of very few published cases in the literature most of which were seen in the Pediatrics population [3].

Our case represents one of these very rare primary intramedullary spinal arachnoid cysts arising in the lower thoracic spine of a young adult male causing progressive sensory and motor symptoms in the lower extremities.

Case Presentation:

A 21-year old male patient, previously healthy, who presented to us with a history of mild chronic radicular sensory and motor complaints in the lower limbs associated with difficulty in walking without assistance. Patient was doing relatively well until 9-months prior to admission when he started complaining of moderate low back pain, which was continuous and exacerbated progressively. The pain was then radiating to the left lower limb, associated with numbness and paresthesia in the whole limb. Four months later these left radicular symptoms subsided but he started to have right lower limb pain associated also with numbness and paresthesia in the whole limb as well as weakness in the aforementioned limb and inability to walk or stand steadily. There was no previous history of sphincter control problems (neither urethral nor anal) and no history of saddle sensory disturbances. There was no previous history of any significant trauma, infection or systematic disease. His Physical Examination showed significant right lower limb motor power deficits 3/5 in all major groups. Babinski test: upward going on the (right) planter, knee jerk revealed hyper-reflexia bilaterally. Patient showed pin brick, touch and temperature decreased sensation below T-12 dermatome. Romberg test was positive. The Patient was thoroughly evaluated and underwent Whole Spine MRI which showed an intramedullary cystic spinal lesion (17 mm* 12mm) at T10-11 level with intensity similar to CSF in all studies and no appreciable enhancement with contrast and no restrictive pattern on diffusion weighted images, which represented most likely an intramedullary arachnoid cyst (Figure.1). The routine hematological and biochemical laboratory evaluation was normal. The management options discussed with the patient who opted surgical treatment.

The cystic lesion was found posteriorly in the cord (intramedullary) filled with fluid similar to CSF. It was causing significant cord compression. The fluid content was completely evacuated and the cyst wall was grossly excised dissecting it from the cord gently (Figure.2). The patient showed dramatic clinical neurological improvement in the postoperative period retaining his normal stance and gait on the 5th postoperative day.

Surgical details

The surgical steps were as follows: with the patient in a prone position after the induction and general anesthesia, the level of the pathology was marked under fluoroscopy guidance. The region is draped in a standard sterile fashion, with a conventional midline approach centered over the appropriate segment using anatomical fluoroscopy and landmarks, then skin and the subcutaneous tissues are opened one-level above and below for adequate exposure, with sub periosteal dissection done down to the specified level to expose the spinous process and the laminae bilaterally, fluoroscopy was re-used to reconfirm the level and a laminectomy performed one level above and below. The dura was opened at the midline with a scalpel, according to the location of the tumor. The incised dura then reflected, and tacked-up with the para-spinal muscles. The cystic lesion was found posteriorly in the cord filled with CSF- similar fluid using a microsurgical technique. Meticulous hemostasis achieved before the dural closure, which was performed using 4-0 vicryl or 6-0 nylon suture with interrupted sutures or continuously locked sutures.

Discussion:

Spinal arachnoid cysts are very infrequent in all age groups and so with the pediatric population. Age and gender do not play a role in the incidence in the first two decades of life [4]. Spinal arachnoid cysts are most often located in the mid- to lower thoracic area [5-8], are found predominantly in males (The incidence predominates in males after the second decade of life) and tend to be symptomatic during the second decade of the patient's life [5,7].

In their typical presentation, spinal arachnoid cysts cause progressive signs and symptoms suggesting spinal cord compression. However, because a cyst can occur at any spinal level and in a patient of any age, no one clinical presentation is pathognomonic, and the clinical sequelae can differ drastically from patient to patient. Nevertheless, we can make certain generalizations: a spinal arachnoid cyst that compresses the spinal cord typically causes waxing and waning pain and progressive spastic or flaccid para paresis, which often are exacerbated by Valsalva maneuvers. Spinal arachnoid cysts can also present with symptoms suggestive of an isolated radiculopathy. Less typical presentations include non-cardiac chest pain, isolated gait difficulty, and isolated urinary urgency [5]. Arachnoid cysts has been

categorized into those that arise primarily with a probable congenital origin and secondary arachnoid cysts that can develop after: trauma; infection; hemorrhage or iatrogenic spinal procedures. Different classifications had been proposed; arachnoid cysts can be classified in relation to the dura mater (Table.1) [4]

Or according to the classification introduced by Nabors et al. [8], listed in Table.2.

Based on the findings of surgical examinations, radiological features and histopathological review of 22-cases, Nabors et al. proposed a classification of spinal meningeal cysts into three categories: spinal extradural meningeal cysts without spinal nerve root fibers (Type I), spinal extradural meningeal cysts with spinal nerve root fibers (Type II), and spinal intradural meningeal cysts (Type III) which represent a separate entity. Type I meningeal cysts were further classified into Type IA, the extradural arachnoid cyst and Type IB, the sacral meningocele [3].

Primary spinal intradural intramedullary arachnoid cyst is extremely uncommon and has been sparsely reported in the medical literature (total of 13 published cases in all age groups) and mostly in the pediatrics population. Only 5 such reports are available in adult population so far [9-12].

The pathophysiology of intramedullary arachnoid cysts is still not fully understood. The suggested theories for the origin of arachnoid cysts in general is still also largely debatable.

They are often attributed to congenital defects. Another possibility is that arachnoid adhesions develop secondary to inflammation, which may arise from infection (meningitis), hemorrhage, or an iatrogenic cause such as injected contrast medium or anesthetics or from the intraoperative contaminants of fibrin glue. Some cysts may be caused by trauma from lumbar puncture, anesthetic procedures, or intradural surgery. Other cysts are idiopathic. Nontraumatic spinal extradural meningeal cysts are believed to be congenital proposed causes of cyst expansion are active secretion from the internal cell lining, an osmotic spinal gradient between the subarachnoid space and the cyst, pulsatile cerebrospinal fluid (CSF) dynamics , or valve-like mechanisms [3,8,10,13,14].

The suggested origin of intramedullary arachnoid cysts is still under scientific research and only few published theories could be found. Goyal et al. [3,11] suggested that the misplaced cellular elements during embryogenesis as the possible etiology. Fortuna and Mercuri suggested trapping of arachnoid granulation at various locations including intramedullary as a pathogenic factor in cyst formation and CSF production and accumulation. [15]

For a patient with symptoms, surgical treatment offers an excellent chance of neurologic recovery. Complete microsurgical resection is the ideal treatment. The standard treatment of an isolated spinal AC is complete surgical removal of the cyst [16-19]. Surgery typically results in excellent outcomes in terms of resolution of symptoms, and is effective across a large range of cyst sizes.

Unresectable cysts should be attempted to be drained and marsupialized. Unfortunately, not all isolated spinal ACs can be fully resected, owing to their location or to intraoperative findings such as extensive adhesion of a cyst to the spinal cord. In such cases, fenestration of the cyst wall, percutaneous drainage, or shunting the cyst into the peritoneal cavity may relieve symptoms. However, what seems to be common between all the published cases of intramedullary arachnoid cysts is the excellent surgical outcome and the good prognosis when detected and treated properly [20-22].

Conclusion:

Despite the rarity and complexity of spinal intramedullary arachnoid cysts and their obscure origin, a surgeon should keep this diagnosis in mind when assessing any related presentation, due to the fact that this lesion carries an excellent prognosis when diagnosed and treated properly.

Ethics Statement:

This study was carried out in accordance with the recommendations of Royal medical Services Ethical Committee for Medical Research, ethical approval (24/12/2017).

Conflict of Interest Statement:

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

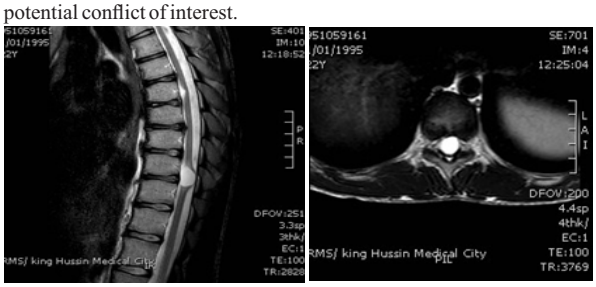

Figure.1: Sagittal and axial MRI showing small intramedullary cystic lesion.

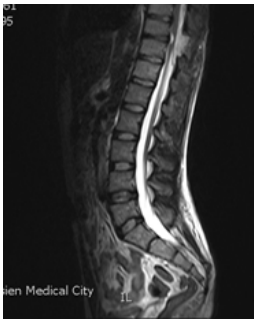

Figure.2: Sagittal post-operative MRI showing total obliteration of the cystic lesion.

TABLE.1 Arachnoid Cysts classification in relation to the Dura Mater

1.Extradural	
2. Intradural	A. Extramedullary B. Intramedullary

TABLE.2 Nabors et al. in 1988 Made the Following Classification of Arachnoid Cysts

Type I: Extradural. Without nerve root fibers Ia. Extradural arachnoid cyst Ib. Sacral meningocele or occult sacral meningocele
Type II: Extradural with nerve root fibers (includes Tarlov's perineural cyst and spinal nerve root diverticulum)
Type III: Intradural

References

1. Greenberg Handbook of Neurosurgery, 8th edition , pages 265, 1142-43.
2. Hughes G, Ugokwe K, Benzel EC. A review of spinal arachnoid cysts. Cleveland Clinic journal of medicine. 2008;75 (4):311-5.
3. Ranjan M, Yasha TC, Prasad C, Sampath S and Indira Devi B. Spinal Intramedullary Arachnoid Cyst – a Rare Case or a Distinct Rare Entity?. Austin Neurosurg Open Access. 2014;1 (4): 1018.
4. McComb JG. Chapter 17 - Pediatric Spinal Arachnoid Cysts A2 - Wester, Knut. Arachnoid Cysts: Academic Press; 2018. p. 239-51.
5. Kumar A. Chapter 19 - Spinal Arachnoid Cysts A2 - Wester, Knut. Arachnoid Cysts: Academic Press; 2018. p. 271-83.
6. Cloward RB. Congenital spinal extradural cysts: case report with review of literature. Ann Surg 1968;168:851_64.
7. Gortvai P. Extradural cysts of the spinal canal. J Neurol Neurosurg Psychiat.1963;26:223_30.Amsterdam: North-Holland; 1976. p. 137_75.
8. Nabors MW, Pait TG, Byrd EB, et al. Updated assessment and current classification of spinal meningeal cysts. J Neurosurg 1988;68: 366_77.
9. Novegno F, Umana G, Di Muro L, Fraioli B, Fraioli MF. Spinal intramedullary arachnoid cyst: case report and literature review. Spine J. 2014; 14: e9-15.
10. Gezici AR, Ergün R. Intramedullary arachnoid cyst: 3-year follows up after bilateral dorsal root entry zone myelotomy. Acta Neurochir (Wien). 2008; 150: 595-597.
11. Goyal A, Singh AK, Singh D, Gupta V, Tatke M, Sinha S, et al. Intramedullary arachnoid cyst. Case report. J Neurosurg. 2002; 96: 104-106.
12. Diyora B, Kamble H, Nayak N, Dugad P, Sharma A. Thoracic intramedullary arachnoid cyst. Neurol India. 2010; 58: 964-966.
13. Galzio RJ, Zenobii M, Lucantoni D, Cristuib-Grizzi L. Spinal intradural arachnoid cyst. Surg Neurol. 1982; 17: 388-391.
14. Gimeno A. Arachnoid, neurenteric, other cysts. Congenital malformations of the spine and spinal cord. Vinken PJ, Bruyn GW, editors. Handbook of clinical neurology. Amsterdam: North-Holland. 1978; 32: 393-448. 1978.
15. Fortuna A, Mercuri S. Intradural spinal cysts. Acta Neurochir (Wien). 1983; 68: 289-314.
16. Hughes G, Ugokwe K, Benzel EC. A review of spinal arachnoid cysts. Cleveland Clinic journal of medicine. 2008;75(4):311-5.
17. Takahashi S, Morikawa S, Egawa M, Saruhashi Y, Matsusue Y. Magnetic resonance imaging-guided percutaneous fenestration of a cervical intradural cyst. Case report. J Neurosurg 2003;99:313_15.
18. 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 Pediatrics 2012;9(4):432-41.
19. Rabb CH, McComb JG, Raffel C, Kennedy JG. Spinal arachnoid cysts in the pediatric age group: an association with neural tube defects. J Neurosurg 1992;77(3):369-72.
20. Chan RC, Thompson GB, Bratty PJ. Symptomatic anterior spinal arachnoid diverticulum. Neurosurgery 1985;16:663-5.
21. Wang MY, Levi AD, Green BA. Intradural spinal arachnoid cysts in adults. Surg Neurol 2003;60:49-55.
22. Willems PW, van den Bergh WM, Vandertop WP. An arachnoid cyst presenting as an intramedullary tumour. J Neurol Neurosurg Psychiatry 2000;68:508-10.