EXTRA-MEDULLARY INTRA-DURAL ARACHNOID CYST of THORACIC SPINE WITH SPINAL ARACHNOID WEB: A RARE CASE

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Abstract:

Intradural extramedullary arachnoid cyst in the spine is extremely uncommon, with only 5% occurring in this site. These cysts rarely cause compression of the spinal cord. The etiology of these cysts is uncertain, however, the majority are congenital. Other possible causes include - trauma, infection, inflammation, postoperative complication, or lumbar puncture. Diagnosis can be establishedusing MRI. If the patient is asymptomatic, conservative therapy is advised. Complete excision isrecommended for patients with clinical symptoms and severe neurological damage.

The spinal arachnoid web is an abnormal thickening of thebands of intradural arachnoid tissue that extend from the pial surface of the dorsal aspect of the spinal cord. It can originate from a malformed or ruptured arachnoid cyst. These rare arachnoid webs can cause compressive myelopathy, particularly in the thoracic spine. Inflammation following trauma, prior surgery,haemorrhage,or infection can lead to an arachnoid web.

We report a case ofa 28 year-old-femalepresenting with a thoracic intradural extramedullary - arachnoid cyst along with spinal arachnoid web having complaints of gait disturbance and who responded well to surgical intervention. Complete surgical excision was done with a good outcome.

Keywords: Arachnoid cyst; rare cyst; Cyst excision; Intradural; MRI; Outcome

Introduction:

Spinal arachnoid cysts are uncommon accounting for 1% of all primary spinal mass lesions. They can be extradural or intradural in location. These arachnoid cysts are characterized by a wide range of clinical manifestation ranging from asymptomatic to severe myelopathy or radiculopathy[2]. They rarely cause spiral cord compression.[1,2,3,5,7]Diagnoses is based on combined radiological imaging, intra- operative findings and histopathological findings.

We present a case of 28-year-old female who presented with severe myelopathy caused by aextra medullary intraduralthoracic spinal arachnoid cyst along with spinal arachnoid web.Dorsal spine MRI revealed long segmental cystic lesion of CSF at dorsal extra medullary space extending from D_1 to D_7 vertebral level causing moderate to severe compression of cord along with arachnoid web originating from arachnoid cyst, confirmed intraoperatively. Only a handful of reports ofextra medullary intradural thoracic spinal arachnoid cyst along with arachnoid webhave been reported so far.

Case description:

A 28-year-old female patient came to the neurosurgery OPD with complaints of inability to walk. Shecomplained of gradual weakness in both feet for the past 6 months, which worsened gradually with her movements restricted to and using wheel chair only. Patient had lower segment caesarean section (LSCS) one year back. She had no difficulty while urinating ordefecating. There were no bumps on her back, or any history of fever, weight loss, trauma, or any similar illness in her family. Physical examination at presentation revealed Glasgow Coma Scale (GCS) Eye (E) 4 Motor (M) 5 Verbal (V) 6 with blood pressure at 110/70 mmHg, heart rate 72 per minute, and respiratory rate 20 per minute. Neurological examination revealed spastic paraparesis with power grade 1/5 in most muscle groups of the lower extremities. Muscle stretch reflexes were brisk with sustained ankle clonus (grade 5) and extensor plantar response (Babinski positive).Sensory examination revealed loss of pain, touch, vibration and proprioception sensations below C_7/D_1. No spinal deformity was present

on external examination. Reflexes of the upper limb, strength of hand grip and power were normal bilaterally with The atrophy or wasting of muscle groups.

Dorsal spine MRI revealed "focal segmental prominence of the posterior CSF space extending from the inferior end plate of D_1 to D_7 causing moderate to severe cord compression with ventral displacement and cord signal change.Arachnoid web was confirmed intra operatively. Electromyography was not done.The arachnoid cyst was exposed by total laminectomy from D_1 to D_7 level. Dura was incised and large sized intradural arachnoid cyst along with arachnoid web was located dorsal to spinal cord. The cyst contained clear fluid Suggestive of CSF. The cyst ruptured during surgery, fluid was drained and excision of the cyst was done in pieces. The dura was repaired with duroplasty. There was no complication following repair of dural defect and no further leakage of CSF found. Removal of cyst resulted in decompression and immediate cord re-expansion.She made excellent post-operative improvement and was ambulating without support on 2nd day of surgery. Power in lower limb muscles was 4/5 on day 2 of surgery. Histological evidence revealed multiple pearly grey white soft tissue pieces measuring altogether 0.8 X 0.7 X 0.5cm. Microscopic section revealed fibro collagenous tissue focally lined by arachnoid cells. The sub epithelium shows mild chronic inflammation and foci of calcification. The histopathology report was consistent with the clinical diagnosis of arachnoid cyst.The patient was ambulating without support at the time of discharge.

Discussion

Terminology and Classification Arachnoid cyst isfirst detected by Schlesinger in 1893 and first reported by Spiller et al in 1903. Based on different pathogenic conception, several terms such as "Arachnoid Diverticulum", Leptomeningeal Cyst", "localized adhesive arachnoiditis", and "spinal serious meningitis" have been used as another name for Spinal Arachnoid Cyst. Nabor et al classified spinal cyst into three types, namely type 1: extradural meningeal cyst without spinal nerve fiber (anterior and lateral meningococcus); type 2:

extradural meningeal cyst containing spinal nerve fiber; type 3: intradural arachnoid cyst [3, 9]. Based on the classification by Nabor, this patient's case falls into the spinal cyst type 3.

The pathogenesis of how spinal arachnoid cyst occurs is not entirely understood and is deeply multifactorial. Several theories mentioned that this arachnoid cyst is a congenital defect of the dura mater, and is always related to the subarachnoid space via small defects in the dura and herniation of the arachnoid [1,9]. Several cases may be the result of trauma, infections, inflammations, post-surgery complication, and lumbar puncture [1]. There was were no other predisposing factors. Therefore, the arachnoid cyst in this patient is deemed to be congenital or idiopathic.

Funao et al stated that the average age of patients with the spinal arachnoid cyst is at the onset of 39 (with an age range of 22 – 60 years old) [9]. Fam et al argued the onset of arachnoid cyst is at the age of 53 (with an age range from 34 – 91 years old) [6]. This patient was one of the rare cases in which the disease occurs at the age of 28. Intradural arachnoid cyst in the spine is a very uncommon cyst and rarely becomes the cause of spinal cord compression which gives off myelopathy symptoms [1,2].

Based on reports, only 1 out of 290 intraspinal cysts produces symptoms [2]. Arachnoid cyst is one of the cyst types which causes spinal cord compression. Arachnoid cysts take place more frequently in extradural than intradural locations [8]. Clinical symptoms that emerge in patients with intradural extramedullary arachnoid cyst are pain (64%), including back pain (46%), extremity radiculopathy pain (9%), and neck pain (9%). Limb weaknesses can happen, and autonomic function dysfunction may arise, among which is urinary tract disorder. Final reported 50% of patients will suffer from the urinary tract and intestinal disorder [9].

Other emerging clinical symptoms that may appear are hypoesthesia, incontinence, paraparesis, or tetraparesis [3].

Wang et al have reported that intradural cyst located in the anterior region will present features consisting of extremities weakness and myelopathy, while cyst located in the posterior region will provide symptoms such as hypoesthesia and neuropathic pain [9].

Clinical symptoms found in this patient consisted of immense pain, especially after activities, that radiated from the neck down to the upper back, further spreading to both arms and hands. Weaknesses were found on both hands and feet which happened after the pain took place and grew more intense. Autonomic dysfunction found on the patient, consisting of retention uri and retention alvi, was identified after sensory and motor complaints. 3.5. Imaging

Extradural arachnoid cysts are much more common than intradural cysts. Based on research conducted by Nabor et al, 18 out of 22 patients were diagnosed with an extradural arachnoid cyst while only 4 of them have an intradural arachnoid cyst. Despite this, cases in children differ by a considerable margin where 58% of child patients have an intradural arachnoid cyst. This is likely due to the high occurrence of congenital malformations of the central nervous system in children [1,2,9]. The thoracic region has been reported to be the area in which intradural arachnoid cyst occurs most frequently with 78% involvement of the dorsal region. The reason why the intradural arachnoid cyst takes place more often in the dorsal region is that the dorsal column is the longest segment which also has a narrow spinal canal [9].

Thoracic cysts commonly occur on young teenagers, while thoracolumbar and lumbar cysts usually appear on adults in their 40s [1,10]. Magnetic Resonance Imaging (MRI) is useful in detecting the mass and consistency of the cerebrospinal fluid. MR, will identify hypointense on T1W1 and hyperintense on the T2W1 [11]. Myelography is suitable to detect the dural defect which makes it possible to provide an accurate diagnosis for this lesion [1,9]. In this case, the cyst is locateBased on reports, only 1 out of 290 intraspinal cysts produces symptoms [2]. Arachnoid cyst is one of the cyst types which causes spinal cord

compression. Arachnoid cysts take place more frequently in extradural than intradural locations [8]. Clinical symptoms that emerge in patients with intraduralextramedullary arachnoid cyst are pain (64%), including back pain (46%), extremity radiculopathy pain (9%), and neck pain (9%). Limb weaknesses can happen, and autonomic function dysfunction may arise, among which is urinary tract disorder. Final reported 50% of patients will suffer from the urinary tract and intestinal disorder [9]. Other emerging clinical symptoms that may appear are hypoesthesia, incontinence, paraparesis, or tetraparesis [3]. Wang et al have reported that intradural cyst located in the anterior region will present features consisting of extremities weakness and myelopathy, while cyst located in the posterior region will provide symptoms such as hypoesthesia and neuropathic pain [9]. Clinical symptoms found in this patient consisted of immense pain, weaknesses was found on both feet which happened after the pain took place and grew more intense.

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Management The selected therapy for patients with symptomatic neurological damage due to intradural arachnoid cyst in the spine is total cyst excision [1,2,6,9,10]. Aspiration and partial excision of the cyst wall are not advised and there have been reports of recurrence happening after total cyst excision [9,10]. Asymptomatic patients are only recommended observation as their conservative therapy [2]. Clinical symptoms found in this patient is the increasingly

aggravating neurologic deficit—an indicationof the need for surgery. This patient had undergone total excision on the entire arachnoid cyst wall.

Outcome Several studies have reported favorable results after undergoing total excision of arachnoid cyst: complete disappearance of pain and no patients were experiencing a recurrence of cyst and kyphosis formation [7,9,10]. Funao et al deduced that patients with prolonged symptoms and a large cyst will produce less promising postoperative results. In his study, 12 patients with clinical symptoms of more than one year and cyst size larger than 5 vertebrae showed unfavorable results after their total cyst excision [9,11]. This patient's clinical symptoms and neurologic deficit occur for 6 months (less than one year) and the cyst involved7 vertebral segments. After having to go through total cyst excision, the patient has shown good results compared to before surgery: decreased pain and improved extremity strengths.

Previous reports of Thoracic Arachnoid cysts

Author	**Average age (years)**	**Number of cases**	**Localisation**
Mohindra et al. [14]	25(4-46)	10	C_3,T_5,L_1,S_1
da Conceicao et al. [15]	28	1	T_1-T_{12}
Wenger et al. [16]	55	1	T_{5-6}
Rao et al. [17]	9	1	T_{1-5}
Novegno et al. [18]	31	1	T_{11-12}
Osenbach el al. [19]	41.5	14	C_3, T_9, L_2
Wang et al. [20]	52 (17-80)	21	C_4, T_{14}, L_3
Van Nuenen el al.[21]	65	1	T_7
Payera and Bruhlhart[22]	33	1	T_{11}-L_1
Kumar et al. [23]	40 & 75	2	T_{3-5}& T_{5-7}
Peruzzotti- Jametti et al. [24]	53	1	C_1-T_{11}
Endo et al.[12]	54	1	T_{3-8}
Albayrakhümeyra et al.[25]	48	1	T_4
Present Case	28	1	T_{1-7}

Conclusion:

An intradural extramedullary arachnoid cyst is an extremely rare cyst. It is a type of cyst that is very uncommon to cause spinal cord compression and oftentimes, it is asymptomatic. The

diagnosis of arachnoid cyst is based on the appeared clinical symptoms, physical examination, and histopathology. The therapy of choice when clinical symptoms appear, and spinal cord compression takes place which leads to the neurologic deficit is the total cyst excision. However, when the case is asymptomatic, then the therapy of choice is observation. This case utilized MRI with contrast as the imaging modality, proving to be very helpful in establishing diagnosis so that proper therapy management can be given according to the patient's condition, producing the most optimal result.

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in the cervical region, an area in which intradural arachnoid cyst rarely takes place. Cervical MRI had been conducted on this patient to help establish the diagnosis in addition to myelography.

3.6. Management

The selected therapy for patients with symptomatic neurological damage due to intradural arachnoid cyst in the spine is total cyst excision [1,2,6,9,11]. Aspiration and partial excision of the cyst wall are not advised and there have been reports of recurrence happening after total cyst excision [9,11,13]. Asymptomatic patients are only recommended observation as their conservative therapy [2]. Clinical symptoms found in this patient is the increasingly aggravating neurologic deficit—an indication of the need for surgery. This patient had undergone total excision on the entire arachnoid cyst wall.

3.7. Outcome

Several studies have reported favorable results after undergoing total excision of arachnoid cyst: complete disappearance of pain and no patients were experiencing a recurrence of cyst and kyphosis formation [7,9,11]. Funao et al deduced that patients with prolonged symptoms and a large cyst will produce less promising postoperative results. In his study, 12 patients with clinical symptoms of more than one year and cyst size larger than 5 vertebrae showed unfavorable results after their total cyst excision [9,12].

This patient's clinical symptoms and neurologic deficit occur for 4 months (less than one year) and the cyst involved only 3 vertebral segments. After having to go through total cyst excision, the patient has shown good results compared to before surgery: decreased pain and improved extremity strengths.

4. Conclusion

An intradural extramedullary arachnoid cyst is an extremely rare cyst. It is a type of cyst that is very uncommon to cause spinal cord compression and oftentimes, it is asymptomatic. The diagnosis of arachnoid cyst is based on the appeared clinical symptoms, physical examination, and histopathology. The therapy of choice when clinical symptoms appear, and spinal cord compression takes place which leads to the neurologic deficit is the total cyst excision. However, when the case is asymptomatic, then the therapy of choice is observation. This case utilized MRI with contrast as the imaging modality, proving to be very helpful in establishing diagnosis so that proper therapy management can be given according to the patient's condition, producing the most optimal result.

Compliance with ethical standards

Disclosure of conflict of interest: None

Statement of ethical approval: Institutional ethical clearance and patient informed consent obtained.

Statement of informed consent : Obtained

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