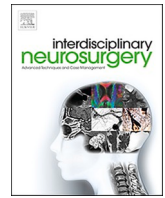




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Intraspinal lymphangioma of the lumbar spine confused with spinal synovial Cyst: A case report

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ABSTRACT

Introduction: Lymphangiomas are benign tumors that result from the aberrant proliferation of lymphatic vessels. Although they are common in the head and neck, they can occur anywhere. Intraspinal lymphangiomas, however, are extremely rare, with only a few relevant cases have been reported in the literature. Herein, we present a case of intraspinal lymphangioma confused with a spinal synovial cyst in an 80-year-old male.

Case Presentation: An 80-year-old male patient presented with severe lower back pain of 3-month duration radiating to the lower limbs. An MRI of the lumbar spine was done and showed a large right intra-spinal extradural cystic structure causing severe compression on the nerve roots. The imaging was suggestive of a spinal synovial cyst, and the patient was referred to orthopedic surgery for synovial cyst excision. The total resection of the mass was done successfully. Histopathologic examination revealed spinal lymphangioma. Postoperative evaluation showed stable vital signs, normal physical exam findings, and marked improvement of the patient's lower back and radicular pain. He was followed up for 3 months without any reported complications or adverse events.

Discussion: Due to the wide spectrum of pre-operative differential diagnoses for lymphangiomas, a definite pre-operative diagnosis is extremely difficult; thus, histopathologic assessment is the sole specific method for their diagnosis.

Conclusion: This case demonstrates how spinal synovial cysts are an important differential for lymphangiomas in patients presenting with intraspinal extradural lesions.

1. Introduction

Lymphangioma is a benign tumor that results from the haphazard proliferation of lymphatic vessels [1]. Although soft tissue lymphangiomas are not uncommon, intraspinal bony lymphangiomas associated with spinal compression are extremely rare [2]. We describe a patient who presented with progressive radicular back pain and initial suspicion of lumbar stenosis along with a spinal synovial cyst causing severe spinal canal stenosis with nerve root compression that, on further evaluation, turned out to be an intraspinal extradural lymphangioma.

2. Case presentation

We present an 80-year-old male who presented to the emergency department with severe lower back pain that had been present for three months; the pain was constant and radiating to lower limbs. No exacerbating or relieving factors were found, and it was not associated with any other complaints. Past medical history revealed long-standing hypertension that was treated with valsartan and hydrochlorothiazide. Six months prior to presentation, a total right knee replacement had been performed.

Upon admission, vital signs were normal. A physical exam revealed lower back tenderness along with left knee tenderness and a scar over

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the right knee consistent with the previous surgery. The straight-leg test was positive bilaterally. Upon testing of the extremities, muscle bulk was found to be normal with intact sensation and normal tone. Importantly, the patient had an intact anal wink, no fecal or urinary retention or incontinence, normal reflexes, and no saddle anesthesia. Power testing revealed an overall score of 4/5 in both upper and lower extremities, and distal pulses were bilaterally palpable.

Based on our patient’s presentation, an MRI of the lumbar spine was done and showed arthropathy of bilateral facet joints at the L5/S1 level with a large right intra-spinal extradural cystic structure causing severe compression on the nerve roots (Fig. 1 A and B). The imaging was suggestive of a spinal synovial cyst, and the patient was referred to orthopedic surgery for lumbar spine decompression of the L5-S1 vertebra, along with synovial cyst excision. The patient then received the surgical procedure, and total excision of the specimen was successfully done. The operative course was nearly similar to that usually experienced during excision of the synovial cysts. Macroscopically, the specimen was composed of a cystic mass measuring 3.5 × 2.5 × 1.5 cm (Fig. 2). It was then sent to the pathology laboratory for microscopic diagnosis, which showed histological evidence consistent with lymphangioma (Fig. 3 A & B).

Postoperative evaluation showed stable vital signs, normal physical exam findings, and marked improvement of the patient’s lower back and radicular pain. The patient was given postoperative intravenous antibiotics and analgesia and then discharged safely with satisfactory outcomes. An MRI three months later revealed no signs of recurrence or mass effect.

3. Discussion

Lymphangioma is a benign soft tissue tumor that results from the slow proliferation of lymphatic vessels; it most commonly involves the head and neck area but can occur anywhere [1]. Lymphangiomas seldom involve the spinal column, and when they do, they usually occur as a solitary acquired lesion [2]. Based on our literature review, we were only able to find a few relevant reports that manifested as solitary

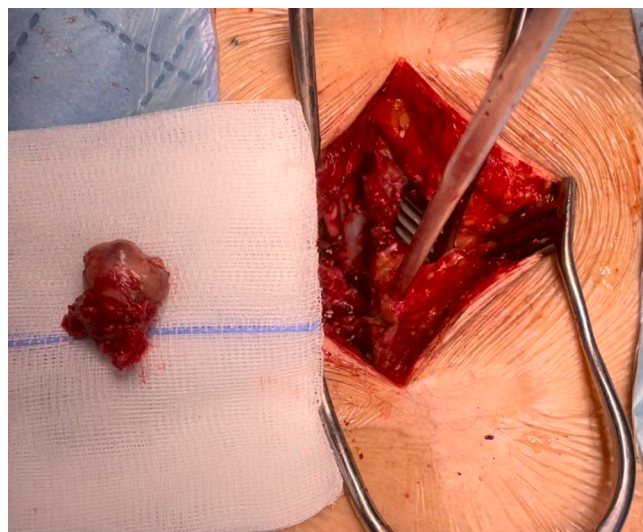


Fig. 2. Macroscopic view of the resected specimen composed of a cystic mass measuring 3.5 × 2.5 × 1.5 cm, it was initially suspected to be synovial cystic mass, but pathological evaluation revealed it to be a lymphangioma.

lymphangiomas causing severe spinal canal stenosis with nerve root compression [1–6].

The pathophysiology of lymphangiomas differs in the congenital form when compared to the acquired. Congenital lymphangioma results from malformations in the lymphatic tissue and presents most commonly in the first two years of life in the head and neck [1]. Whereas the acquired form is caused by obstruction of the lymphatic vessels, leading to mass formation that rarely resolves spontaneously [5]. The tumor can bleed internally, leading to expansion in its volume [7]; this could have contributed to the progressive worsening of our patient’s spinal compressive symptoms.

On MRI, lymphangiomas can be mistaken for spinal cystic lesions; this is because both can demonstrate low signal intensity on T1 and high

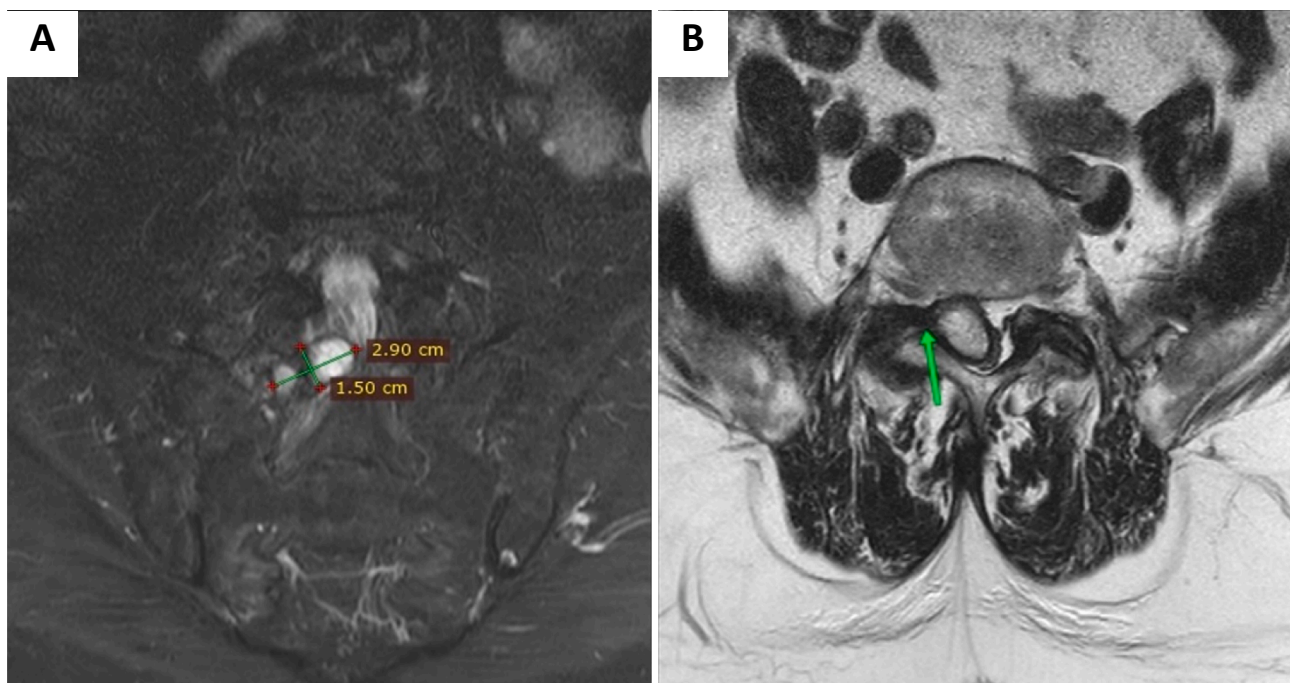


Fig. 1. A & B: is a magnetic resonance imaging showing arthropathy of bilateral facet joints at L5/S1 with a large right intra-spinal extradural cystic structure (presenting low T1 signal, High T2/STIR signal) causing severe compression on thecal sac. There is impression of a connection (arrow) of the cystic structure with right facet joint suggesting spinal synovial cyst.

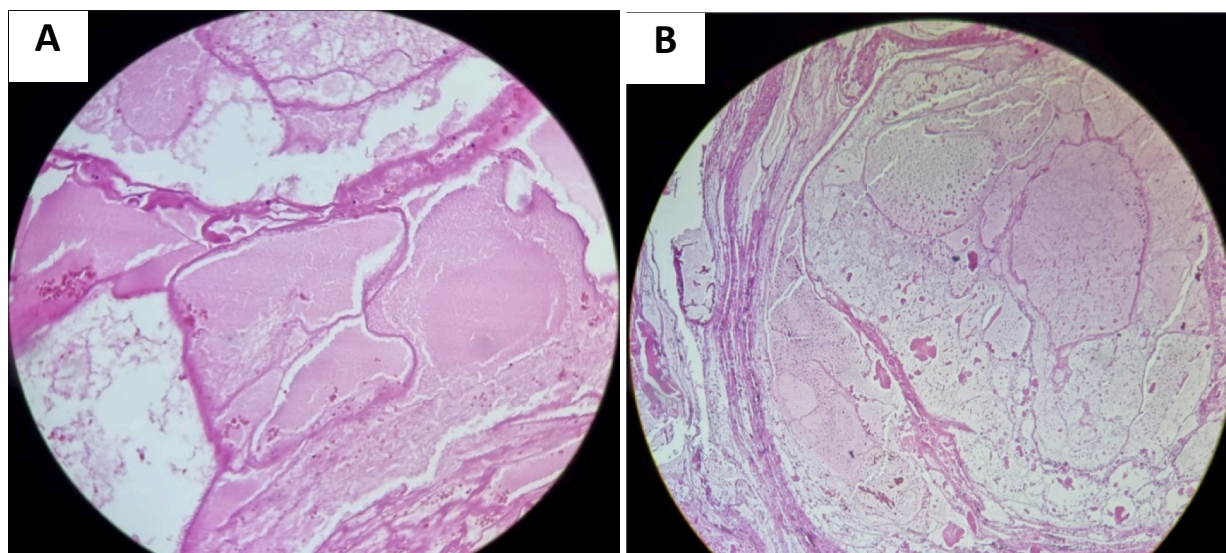


Fig. 3. A & B: Microscopic view of the resected specimen showing histological evidence suggestive of lymphangioma, along with evidence of tissue undergoing coagulative necrosis.

signal intensity on T2/STIR. This is clearly demonstrated in our case, as our preoperative evaluation led us to a preliminary diagnosis of a spinal cystic lesion instead of a lymphangioma, which was then proven otherwise via pathological diagnosis.

The differential diagnosis of lymphangiomas is broad and includes hemangiomas, intraspinal arachnoid cysts, metastatic tumors, lipomas, embryonal tumors, and other rare intraspinal tumors such as heman-giopericytomas [6]. Due to the broad differential, establishing a definitive preoperative diagnosis based on imaging and clinical presentation is difficult, making histological examination the gold standard test for diagnosing lymphangiomas [6].

Surgical resection remains the best treatment option for lymphangiomas causing spinal cord and/or nerve root compression with resultant focal neurological deficits [2]. Inoperable tumors can be treated with a wide variety of options, including embolization, sclerotherapy, chemotherapy, radiation, and recombinant interferon. Local recurrence of the tumor can also occur; this is especially true if surgical resection and/or aspiration fail or are incomplete [2].

Kerolus M.G. et al. presented a case of an intradural, extramedullary cavernous lymphangioma in the thoracic spinal cord in an 83-year-old female patient [1]. She presented with progressive ataxia, band like sensation in the middle and lower thoracic dermatomes and bilateral lower extremity weakness. MRI was done and revealed hyperintensity on T2 and enhancement of an intradural cystic mass with notable compression of the spinal cord. Complete surgical resection was not possible because the tumor was adhering to the pia and microvasculature of the thoracic spinal cord. The mass later recurred and was treated with diversion of the cystic fluid into the peritoneum.

Chu M. et al. reported a case of lymphangioma in the epidural space of the cervicothoracic spine in a 61-year-old female patient [2]. She presented with pain and numbness of her left upper limb, MRI was done and revealed a soft tissue mass in the left epidural space at C6 to T1 level. The patient underwent an osteoplastic laminectomy.

Lee C.S. et al. described a case of cervical epidural lymphangioma in a 43-year-old woman who presented with neck pain and right arm numbness for 2 months [3]. MRI demonstrated an epidural cystic mass with extension to the right C6-C7 neural foramen and fluid–fluid levels on T2 weighted image. The tumor was completely excised microscopically with histology confirming the diagnosis of cavernous lymphangioma.

Fattahi A. et al. revealed a case of lymphangioma in the thoracic spine with epidural compression in a 61-year-old man [4]. He presented

with progressive partial paralysis of the lower limbs and sphincter disturbance for 5 months. MRI revealed an enhancing posterior epidural mass at the T7-T8 level with significant cord compression. The patient underwent a T7-T8 laminectomy.

Kim H.S. et al. described a case of cystic intraspinal epidural lymphangioma of the cauda equina in a 63-year-old female at the level S1-S2 [5]. The mass was removed by percutaneous full endoscopic treatment using epidural anesthesia.

Jiang Y.G. et al. reported two cases [6]. The first case was a 47-year-old man with one month history of bilateral lower limb weakness and constipation with decreased knee and ankle reflexes but an otherwise normal physical exam. MR showed two intraspinal tumors at the level of S2/S3 projecting into the sacrum. Intraoperative evaluation revealed a cystic structure adhering to the cauda equine that was removed and sent for pathology, which then revealed a cystic lymphangioma. The second case described a 12-year-old boy complaining of backpain, lower limb weakness, and constipation. History was unyielding and physical examination showed limitation of lumbar spine movement and tenderness along the spinous processes of L1-L5 vertebra. MRI showed an irregular intraspinal lesion that pressed on the spinal cord at the lumbar spine. Intraoperative evaluation revealed a saccular extradural intraspinal mass that was completely excised, and sent for pathology, which showed spongiform-type lymphangioma (Table 1).

4. Conclusion

Our patient presented with an extremely rare case of intraspinal extra-dural lumbar lymphangioma that was confused with a spinal synovial cyst. The lesion caused severe radicular and back pain that warranted surgical intervention with complete resection of the mass. This led to the complete resolution of the patient's symptoms. This case demonstrates how spinal synovial cysts are an important differential for lymphangiomas in patients presenting with intraspinal extradural lesions.

5. Decelerations

Patient consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Table 1

Summary of The Reported Cases with Intraspinial Lymphangioma, in Comparison with The Patient Presented in This Work. (M: Male, F: Female).

Report	Age (y)/ Gender	Site	Presentation	Imaging	Treatment	Outcome
Our case	80/M	Lumbar spine.	Severe lower back pain.	Large right intra-spinal extradural cystic structure causing severe compression on the nerve roots.	Lumbar spine decompression of the L5-S1 vertebra, along with synovial cyst excision.	Full recovery.
[1]	83/F	Thoracic spine.	Ataxia, band like sensation in thoracic dermatomes, bilateral lower extremity weakness.	Hyperintensity on T2 and enhancement of an intradural cystic mass.	Complete surgical resection was not possible; mass recurred and was treated with diversion of the cystic fluid into the peritoneum.	Progressive neurological decline 3 months postoperatively; MRI showed tumor recurrence. Cysto-peritoneal shunt was done and patient improved slowly over time.
[2]	61/F	Cervico-thoracic spine.	Pain and numbness in left upper limb.	Soft tissue mass in the left epidural space at C6 to T1 level.	Surgical resection and an osteoplastic laminectomy.	Persistent mild pain in left upper limb, no recurrence on imaging.
[3]	43/F	Cervical Spine.	Neck pain, right arm numbness.	Epidural cystic mass with extension to the right C6-C7 neural foramen.	Complete microscopic excision.	Full recovery, no recurrence.
[4]	61/M	Thoracic Spine.	Progressive partial paralysis of the lower limbs and sphincter disturbance.	Enhancing posterior epidural mass at the T7-T8 level.	T7-T8 laminectomy and surgical resection of mass.	Full recovery, no recurrence.
[5]	63/F	Cauda equina.	Autism, dyspnea ecchymosis, gingival bleeding.	2 cm well enhanced epidural mass at right S1 epidural space.	Excision with percutaneous endoscopic procedure.	Full recovery, no recurrence.
[6]	47/M	Cauda equina/ Sacrum.	bilateral lower limb weakness, constipation, decreased knee and ankle reflexes.	Intraspinial tumors at the level of S2/S3.	Complete excision.	Full recovery, no recurrence.
[6]	12/M	Lumbar Spine.	Backpain, lower limb weakness, constipation.	Irregular intraspinal lesion that pressed on the spinal cord at the lumbar spine.	Complete excision.	Full recovery, no recurrence.

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Authorship

All authors attest that they meet the current ICMJE criteria for authorship.

Declaration of Competing Interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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