

## A Rare Case of an Isolated Intra-Thoracic Meningocele: Case Report and Review of the Literature

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**Abstract****Case Report**

A spinal meningocele is a saccular protrusion of the meninges through a dilated intervertebral foramen or a bony defect of the vertebral column. Intrathoracic meningocele is a rare condition. Only a few cases were related in the literature. It is usually associated with neurofibromatosis type 1. Isolated intrathoracic meningoceles without neurofibromatosis is a very rare entity few cases have been reported in the medical literature. Regardless of the treatment plan cross-sectional imaging techniques such as CT and MRI are essential not only for the diagnosis but also to determine the relationship to the surrounding structures and the exclusion of other neuromas and any skeletal deformities. Surgical excision is the treatment of choice in symptomatic patients.

**Keywords:** Meningocele, Thoracic, Isolated.

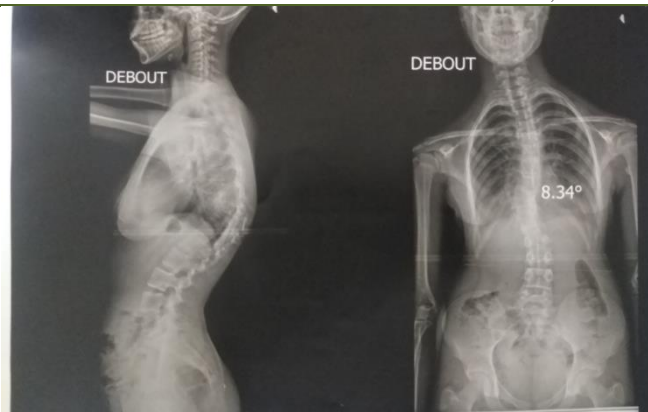
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### INTRODUCTION

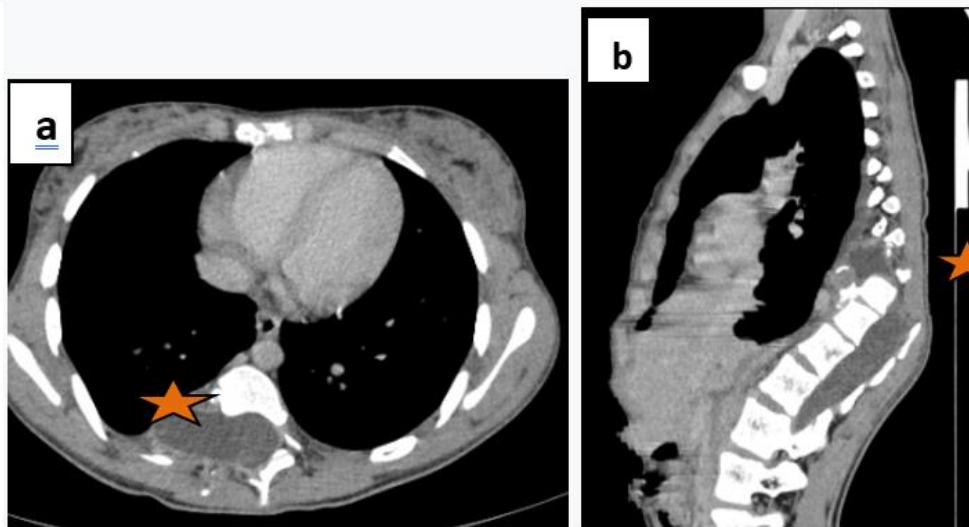
Spinal meningocele is a rare disorder defined as a protrusion of the spinal meninges through a defect in the vertebral column or foramina [16]. Lateral meningocele syndrome is a rare disorder characterized by the widespread presence of protrusions of the arachnoid and the dura matter extending laterally through inter- or intravertebral foramina into the paraspinal, retroperitoneal, or intrathoracic region. It is a hereditary disorder that primarily affects the connective tissues. This disorder manifests itself with formations of cysts at different levels of the central nervous system along with meningeal diverticula protruding through the intervertebral spaces and filled by cerebrospinal fluid (CSF). Other clinical findings associated with the lateral meningocele syndrome include specific facial anomalies, cryptorchidism, hypotonia and muscle atrophy, scoliosis, restricted joint movements, pectus deformities, and abdominal hernias. Lateral meningoceles may be unilateral or bilateral and may exist as solitary or multiple. The pathogenesis of lateral meningoceles is complex because symptom onset depends on the location of the cyst and the meningeal protrusions [11].

### CASE REPORT

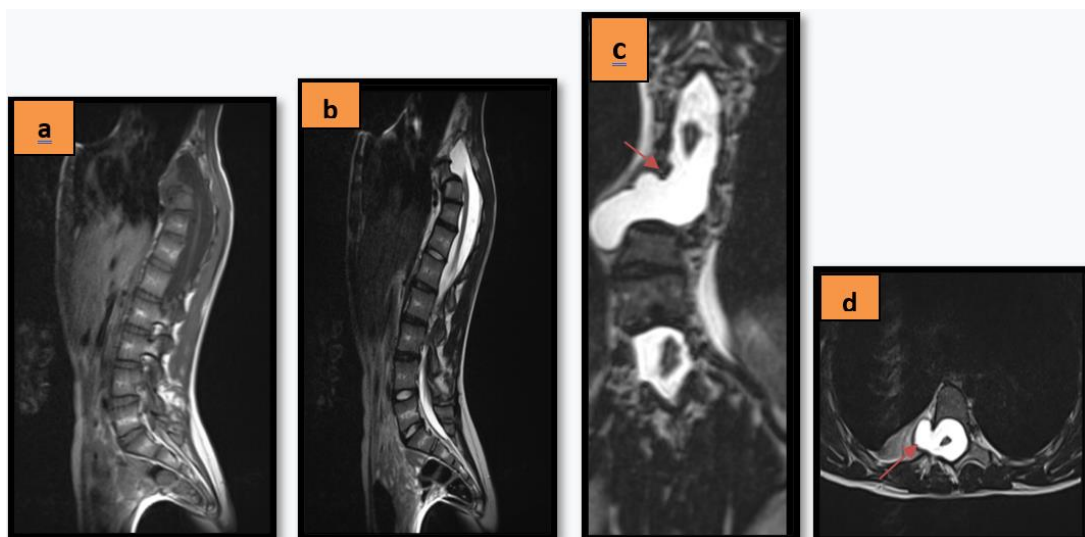
A 13-year-old girl presented with complaints of back mass with functional impotence and claudication of both lower limbs. She had attained normal milestones. No history suggestive of hydrocephalus, urologic, or gastrointestinal problems. General physical examination was unremarkable except for the scoliosis. Neurologic examination revealed impaired sensation over the D9 right dermatome. Radiographic evaluation revealed a dorsal scoliosis with right convexity (Fig. 1). CT revealed Straightness of the dorsal spine from D1 to D8, with acute angulation at D9-D10 and posterior recession at D10, resulting in significant reduction of the medullary canal opposite, with upstream widening. There is an associated right foraminal defect at D8-D9, giving rise to a liquid content of similar density to that of the CSF, continuing with the ductal content, enhanced peripherally by the PDC (meningeal enhancement), measuring 23x50 mm, protruding endothoracically (Fig. 2).Magnetic resonance imaging of the entire spine showed no evidence of Arnold-Chiari malformation, diastematomyelia, tethered cord, or lipoma, dorsal scoliosis significantly reducing the medullary canal at D9-D10-D11 associated with a right foraminal meningocele at D8-D9 (Fig. 3).



**Fig. 1:** X Ray of postero anterior and lateral dorso-lumbo-sacral spine showed scoliosis with right convexity



**Fig. 2:** Transversal (a) and sagittal (b) thorax CT scan with contrast showed a big cystic mass on the left hemithorax with continuous at the spinal canal



**Fig. 3:** Serial sagittal T1W image (a) and sagittal (b), coronal (c) and axial (d) magnetic resonance imaging T2W and of dorso-lumbo-sacral spine show right foraminal meningocele at D8-D9 with foraminal enlargement →

## DISCUSSION

The meningocele is a benign and rare lesion defined as a herniation of the meninges through a

vertebral column defect. More than 80% of spinal meningoceles are located in the lumbo sacral area. The first intra-thoracic location was described in 1933 by Pohl [16].

Lateral meningocele is a rare presentation of spinal dysraphism characterized by extensions of the dura and arachnoid through an enlarged neural foramen [6].

In the thorax, it can be unilateral or bilateral and solitary or multiple. Lateral meningoceles are more frequent because of the relative weakness of the paravertebral muscles and the high pressure gradient between the cerebro-spinal fluid and the thorax [6].

The more accepted mechanism for these lesions is the dural dysplasia along with enlargement of the neural foramen. In these patients, the pleural traction through the negative intrathoracic pressure during inspiration, together with pulsation of the aorta and cerebrospinal fluid pressure, provokes invagination of the subarachnoid space through the foramen [9].

These may occur idiopathically or may be associated with other conditions such as dysraphism or genetic conditions (e.g., neurofibromatosis type 1, Marfan syndrome, Ehlers–Danlos syndrome) [2], but rarely as an isolated lesion [16]. The difference between isolated meningocele and meningocele associated with neurofibromatosis-1 is the family history of neurofibromatosis and the presence of café-au-lait spots, cutaneous neurofibromas, and axillary frecklings and Lisch nodules on the iris, and it's associated with osseous lesion including progressive thoracic scoliosis, and vertebral anomalies [16].

The clinical manifestations of meningocele closely relate with its size and its relationship with surrounding structures. The patient may be asymptomatic or can present with back pain or paraparesis. In contrast, in the setting of a small meningocele, no symptoms can be recorded, and the lesion may be incidentally diagnosed on a routine chest radiograph [6].

Most patients with intrathoracic meningocele are asymptomatic, and incidentally diagnosed on a plain chest x-ray. A few patients present with back pain, cough, or dyspnea due to compression of the lung and the mediastinal structures. The symptoms depend on the size and the location of the meningocele within the thorax [6].

Radiological examination reveals kyphoscoliosis and scalloping of vertebral bodies in approximately 66% of patients with intrathoracic meningocele. Costal changes occur in about half the patients. Enlargement of the inter-vertebral foramina is also very frequent [5].

CT scan and/or MRI are necessary to uphold the diagnosis. On CT scan, the meningocele appears as a well-defined, homogeneous, hypodense, paravertebral mass. Plain radiographs can be helpful by showing an enlargement of the spinal canal, posterior vertebral body

scalloping, pedicle erosion or a widening of the interpedicular distance [6].

MRI, when available, is the best examination for this kind of cystic lesions. It usually shows an intradural, extramedullary space-occupying lesion with T1 and T2 weighted signal intensities identical to that of cerebrospinal fluid (6). It also allows better delineation of the details of multiple lesions, including paravertebral expansion and dural ectasia with scalloping of pedicles, laminae, vertebral bodies and widening of the spinal canal [6].

Since intrathoracic meningocele appears only occasionally, any well substantiated Criteria for its treatment are wanting. The majority of the patients were in the fifth and sixth decades [1].

The treatment is a challenge and depends on the size and the complications of meningocele. When intrathoracic meningocele rapidly increases and complaints of pain, neurological or pulmonary signs, and compression of the trachea or esophagus are present, surgical treatment should be considered. It consists on a resection of the meningocele and repair of the dural defect through laminectomy. However, in the case when meningocele was combined with kyphoscoliosis, it is not easy to suture the meningocele through a posterior laminectomy.

In addition, it can bring about instability of the vertebrae after the removal of the meningocele, posterior spinal arthrodesis as well as anterior spinal arthrodesis might be needed [16]. In case of small meningoceles or asymptomatic patients, a regularly follow-up by chest x ray, Computed Tomography (CT) scans or MRI is indicated [16].

## CONCLUSION

The spinal meningocele is a rare condition. The occurrence of congenital scoliosis with intrathoracic spinal cord herniation is rare. Most patients with intrathoracic meningocele are asymptomatic and incidentally diagnosed after a plain chest x-ray. MRI best depicts the lesions. Surgeons and radiologists should be aware of this rare clinical entity.

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