Isolated Intra-Thoracic Meningocele - A Rare Entity

Furqan A Nizami1*, Haroon Salaria1, Arvind Kohali2, Pooja Gadgotra1, Rubeena Naaz2 and Snober Syed1

1Department of Neurosurgery, Super-specialty Hospital, GMC, India
2Department of cardiothoracic surgery, Super-specialty Hospital, GMC, India

*Corresponding Author: Furqan A Nizami, Department of Neurosurgery, Super-specialty Hospital, GMC, Jammu and Kashmir, India.

Received: June 28, 2015; Published: September 03, 2015

Abstract

Background: The incidence of intra-thoracic meningocele is rare even in known cases of Von Recklinghausen's neurofibromatosis. Incidences of isolated intrathoracic meningocele or multiple meningoceles without neurofibromatosis are even more uncommon. The majority of intrathoracic meningoceles have been discovered by chance or after nonspecific symptoms. We report a case of isolated thoracic meningocele with no association with neurofibromatosis.

Case presentation: 40 year old male presented to the neurosurgical outpatient department with the chief complaints of pain in the chest since 3 years, but no other associated symptom. Chest radiograph showed a round homogeneous mass in the right hemi-thorax. MRI confirmed a cystic cavity adjacent to T7-T9.

Surgical intervention: Right sided anterolateral thoracotomy was done, the cystic mass was extirpated and contents of cavity sent for histopathology.

Conclusion: Isolated intra-thoracic meningocele without neurofibromatosis is a very rare entity few cases have been reported in the medical literature. Symptomatic patients need radiological evaluation and excision of the meningocele with a good predicted post-operative outcome.

Keywords: Isolated; Intrathoracic meningocele; Neurofibromatosis

Introduction

Intrathoracic meningocele is a saccular herniation of the meninges through an enlarged foramina or a defect in the vertebral column to form a CSF filled sac [1]. The meningocele can be rarely congenital or acquired the common variety, which occur as a complication of laminectomy [2]. Anterior meningocele is rare and mostly found in thoracic or sacral region, while posterior constituting 80% generally lies in lumbosacral area. These meningocele are frequently associated with general mesenchymal dysplasia such as neurofibromatosis or Marfan syndrome and is rarely present in an isolated form [3,4].

Case report

A healthy male 40 years of age was admitted to the neurosurgical department of Super-specialty hospital of Government Medical College, Jammu, with complaints of pain in the chest over 3 years. No other symptoms were associated with the pain. He had not previously undergone radiological investigations. There was no relevant previous medical history. There was no evidence of von Recklinghausen's disease nor of any cafe-au-lait spots on the skin. His skeletal development was normal. There was no family history of cutaneous neurofibromatosis or of any congenital bony defect or deformity. The patient had no history of any recurrent chest infections. There was no history of severe spinal trauma. Physical signs were normal in the chest and abdomen. A full neurological examination was carried out: muscular power was preserved in all the four limbs, reflexes were normal and there was no Babinski sign. The deep and superficial sensations and sphincter control were normal. The laboratory findings were within normal limits. The pain was in D7 and D8 dermatome. Plain X-ray of thoracic spine (Figure 1), CT scan of chest (Figure 2A, 2B) followed by MRI of thorax and spine (Figure 3A, 3B) were carried out.

Figure 1: Radiograph of the chest showing a round homogeneous mass in the right hemi-thorax at the level below right bronchus.

Figure 2A: CT scan chest
Axial cuts at the level of T7-T9 showing a round homogenous hypo dense mass on right hemi-thorax.

Figure 2B: CT scan chest
Coronal and Sagittal cuts with reconstruction showing same findings.

Surgery was planned and a pre-anesthetic assessment was carried out. The lesion approached through right anterolateral thoracotomy, pleura opened, the lungs were retracted and the lesion was identified. Its neck was opened through a longitudinal incision for extirpation to confirm the contents of cystic sac. No neural tissue found and the fluid was drained and sent for cytology and culture sensitivity checks. The neck was repaired in two layers. The Valsalva maneuver was performed to check for CSF leak, the chest drain kept in situ via a separate incision and left for 48 hours. The patient had a smooth post operative recovery and was discharged after one week after removal of stitches. Histopathology confirmed finding in favor of meningocele.

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Discussion
Intra-thoracic meningocele is a herniation of the meninges through a foramina or defect in the vertebral column. Isolated cases are rare [1-4]. The first case of intra-thoracic meningocele was reported by Pohl in 1933 [5]. It has been found in patients from 2 days to 68 years of age [6]. Most of them are discovered in fourth and fifth decade which corresponds to our case report where the patient falls in fourth decade of his life. There is no sex predilection [7-9]. Most of the cases are present on right side which corresponds to our case report, and less than 10% are bilateral [8,9].

Andrade., et al. [3] found 134 cases of intra-thoracic meningoceles reported before 1992. They stated that 69% of these thoracic meningoceles were in association with Neurofibromatosis type 1 (NF-1), whereas only 22% were isolated cases. In the same article, a total of 16 patients with thoracic meningocele between 1992 and 2003 were reported. Twelve of these 16 patients had associated NF-1 and the remaining four being isolated cases. The patient presented in this case report had no association with NF-1 or other generalized mesenchymal dysplasia and therefore is classified as being an isolated case of intra thoracic meningocele [10].

The clinical features of the meningocele depends on the size and location and its attachment to the surrounding structures which may vary from asymptomatic if the size is small or produce symptoms if it is large which may include back pain, paraparesis from insult to the spinal cord, or shortness of breath, coughing, and palpitation by compression of the lung and mediastinal structures, as was the case for the patient presented here. Patients with lateral meningocele are usually free from neurologic symptoms. Pain results from the pressure on adjacent structures or from pressure of intercostal nerves. Even progressive hydrothorax caused by rupture of meningoceles has been reported in the literature [11].

Regarding the treatment, surgery is to be avoided unless there are progressive manifestations or growth of meningocele. Some workers advise early surgery even if the patient is asymptomatic because meningoceles may grow progressively, with increasing risk [9]. For small and medium size meningoceles, the commonest approach is through laminectomy and intradural repair of cyst but in large meningoceles thoracotomy is preferred as it offers a large operating field with less chance of damage to the spinal cord [12]. The patient in the case presented here also had a large meningocele which was approached through transthoracically and the entire lesion was extirpated without injuring spinal cord. The patient recovered well post operatively as indicated through regular follow ups.

Conclusion
Intra-thoracic meningocele is extremely rare and even rarer without neurofibromatosis. 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.

Bibliography


Volume 2 Issue 1 September 2015
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