Isolated giant intrathoracic meningocele associated with vertebral corpus deformity

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Abstract
Published reports of intrathoracic meningocele with vertebral corpus defects in the absence of neurofibromatosis are very rare. We report a 9-year-old male with intrathoracic meningocele. We believe that vertebral corpus defects may play a certain role in the etiology of intrathoracic meningocele.

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1. Introduction

Intrathoracic meningocele is defined as a cystic sac whose wall is formed by spinal meninges and containing cerebrospinal fluid, protruding into the thoracic cavity through an enlarged intervertebral foramen [1]. These lesions are commonly associated with neurofibromatosis [2], and the incidence of isolated intrathoracic meningocele without neurofibromatosis is very rare. Their etiology remains controversial [3]. In the absence of neurofibromatosis, the diagnosis of meningocele can be troublesome. The differential diagnosis of posterior mediastinal mass also includes bronchogenic cyst, lymphoma, neurogenic tumors, teratoma, seminoma and germinal cell tumors.

We present a case of isolated giant intrathoracic meningocele with an unusual etiology, associated with multiple vertebral corpus deformity and thoracic skeletal deformity.

2. Case report

A 9-year-old male presented to our clinic with gradually worsening skeletal deformity and shortness of breath. His past medical history included a chest infection in early childhood, and an initial thoracic computed tomography (CT) taken at the age of 6 months showed fusion defect of thoracic vertebrae at levels T7–9, where the anterior aspect of the corpora were absent. On physical examination, the patient had marked kyphoscoliosis and chest wall abnormality of the left hemithorax, and his biological age was approximately 6 years old. His physical examination was further unremarkable; and specifically, there was no evidence of any gross neurological deficit. His chest X-ray confirmed marked vertebral deformity and mediastinal widening. His helical CT showed a significant bony deformity in the 7th–9th thoracic vertebrae, associated with fusion defects on the anterior aspect of these vertebrae (Fig. 1). An axial CT demonstrated a well-defined, 8 × 7 × 5 cm giant cystic lesion at the level of carina in the right paravertebral space, communicating with the spinal canal and significant chest wall abnormality of the left hemithorax (Fig. 2a and b). An MRI scan showed a homogenous cystic mass in the right hemithorax, communicating with the spinal canal through the anterior aspect of the 7th to 9th thoracic vertebrae. The intensity of this cystic mass was similar to the cerebrospinal fluid on T2-weighted images (Fig. 2c). Although the spinal cord was seen at the level of the vertebral body defect, no prominent spinal cord segment was found within...
the meningocele sac. An echocardiogram excluded any congenital intracardiac pathology. The appearances were consistent with the diagnosis of an isolated intrathoracic meningocele with significant skeletal deformity. Although the skeletal defects were considerable, as there were no neurological findings present, conservative management and follow-up was thought to be the best option for this patient at the present time. However, as his kyphoscoliosis is expected to get worse as the child grows, he may ultimately need an orthopedic operative correction.

3. Discussion

Isolated intrathoracic meningocele is a rare condition. Anterior or lateral intrathoracic meningocele, which are mostly found in the superior part of the thoracic vertebral column, may be seen at any age [4]. The majority of the cases are seen in the right hemithorax [5].

Although the developmental mechanism of thoracic meningocele is unknown, several causes suggested as an etiological factor include congenital herniation of the subarachnoid space, elongation of the nerve root sleeve, dural dysplasia, and cystic degeneration of neurofibroma. It is also reported that they may arise in association with spinal dysplasia, spinal trauma or operation [6].

There are a number of options available as a definitive diagnostic tool in this condition, such as CT scan, MRI scan and myelography. However, we have found that helical CT
can be very useful for identifying vertebral deformities in association with this condition.

In our case, due to the fact that the intrathoracic meningocele and vertebral corpus defects were identified together in anatomical relationship to one another, we think that this vertebral corpus defect is an etiological cause of the intrathoracic meningocele.

As our patient was asymptomatic, in consultation with orthopedic surgeons, we initiated conservative management and follow up at present time. Surgery should be contemplated when it becomes evident during careful follow-up that the meningocele sac gradually increases in size and produces neurological and/or cardiopulmonary complications, or when his kyphoscoliosis requires operative correction.

References