Despite the advances in prenatal diagnosis and operative techniques, spinal dysraphism remains one of the most devastating problems of pediatric neurosurgery. We hereby describe an unusual case of myelomeningocele presenting as a retromediastinal mass. To the best of our knowledge, this is the only case published to date.

Case Report

Presentation and Examination. This 11-year-old boy suffering from paroxysmal nocturnal dyspnea and alterations in effort capacity was evaluated. His systemic and neurological examinations were completely normal except for kyphoscoliosis. Routine anterior–posterior and lateral chest x-ray films demonstrated a posterior mediastinal mass. Magnetic resonance imaging demonstrated a cystic mass measuring $8 \times 7 \times 6$ cm, which seemed to be connected with the spinal subarachnoid space through the fusion anomaly at the vertebral corpus below the carina level. The outer margin of the lesions comprised dura mater filled with fluid resembling CSF in density. Neuronal elements penetrated into the cyst from the spinal cord as well (Fig. 1). A thoracic computerized tomography scan demonstrated the fusion anomaly at T4–6 (Fig. 2). The radiological interpretation was that the boy suffered from anterior thoracic myelomeningocele.

Following the radiological findings, the patient was reexamined carefully and his history was retaken in detail. He had never suffered from any neurological deterioration or systemic symptoms, which could be a clue for the presence of hydrocephalus, or from any urological or motor problems. He has been continent since 17 months of age and has been walking since the 14th month. He was able to talk after the 19th month. He had no social problems during his primary school life, and his school success was within normal limits. Kyphoscoliosis was not present at birth; it became remarkable after the 2nd year of life and has progressed ever since. Unfortunately, the boy underwent no medical consultations for this pathological entity until his respiratory problems came to attention. Additional MR imaging was performed to rule out the pathological features that may accompany myelomeningocele, such as hydrocephalus and Chiari malformation. Surprisingly, no other pathological features were seen in those examinations (Fig. 3).

**Abbreviations used in this paper:** CSF = cerebrospinal fluid; MR = magnetic resonance; NF = neurofibromatosis; VB = vertebral body.
Operation. The patient was transferred to our department for surgery. We performed a right anterolateral thoracotomy, reaching the cystic mass located adjacent to the T-4 VB with a transpleural approach. After dissection of the adhesions to the surrounding tissues, the huge cystic mass was seen. With the aid of a pneumatic drill, the right margin of the abnormal T4–6 VBs were removed and the pedicles of the sac was exposed (Fig. 4). After the sac was opened, the CSF was aspirated and the structures that were thought to consist of neural placode were seen. Following the reduction of the placode to the spinal canal, the dura was closed primarily and fibrin glue was applied. The defect between the T-4 and T-6 VBs was reconstructed with a distraction cage and the boy’s own rib, which had been removed during the thoracotomy. The operation ended with a chest tube in the operation site. The early postoperative period was uneventful.

Postoperative Course. The boy experienced complete relief from dyspnea immediately after surgery. On the 3rd day postoperative day, his drainage tube was removed. He was discharged on the 10th postoperative day without any neurological deficit or complication. His early postoperative MR images demonstrated total removal of the mass. The 1-year follow-up period was uneventful, and he will be monitored for a possible increase in kyphoscoliosis.

Discussion

Despite the advances in prenatal diagnosis, as well as maternal supplements of folic acid before conception, neural tube defects are still among the most common problems in pediatric neurosurgery. Although these malformations are believed to occur from the failure of the neural tube to close during embryogenesis, many controversies exist regarding embryological mechanisms.3

Among spinal dysraphic lesions, meningoceles and neurenteric cysts are the most common in the anterior thoracic region. Meningoceles are protrusions of the dura mater and accompanying arachnoid membrane through a defect in the spinal column, with the spinal cord remaining in the confines of the spinal canal.3 These lesions usually occur in the dorsal part of the body, but in the literature examples of anteriorly seated meningoceles do exist, which are mostly in the sacral region. The presence of the thoracic meningoceles is usually associated with NF (that is, in 68.8% of cases). Only 22.4% of intrathoracic meningoceles occur as an isolated pathological feature. The more accepted mechanism for these lesions is the dural dysplasia in patients with NF and enlargement of the intervertebral foramen. In these patients, the pleural traction through the negative intrathoracic force during inspiration, together with the pulsation of CSF pressure, provokes invagination of the subarachnoid space through the intervertebral foramen.3 In our case, there

Fig. 1. Preoperative sagittal MR images demonstrating protrusion of the myelomeningocele sac from the defective vertebral arches.

Fig. 2. Computerized tomography scan demonstrating the fusion anomaly at the thoracic level.
were no signs of NF, and the neuronal elements and the sac were settled anteriorly, not laterally, through a hemivertebral segment.

Spinal neurenteric cysts are rare, infrequently reported congenital abnormalities believed to be derived from an abnormal connection between the primitive endoderm and ectoderm during the 3rd week of life. Intraspinal neurenteric cysts represent 0.3 to 0.5% of all spinal tumors. As with the other types of dysraphic lesions, no consensus exists on their pathogenesis. Theories include the following: 1) a primary adhesion of endoderm anterior to the notochord; 2) incomplete excalation of the notochord; 3) persistence of the neurenteric canal or formation of an accessory neurenteric canal with a split notochord; and 4) displacement of endodermal cells. Neurenteric cysts are not confined to the spinal column, they may be found within the brain, mediastinum, abdomen, or pelvis, as well as in subcutaneous locations. They are most commonly found, however, in the lower cervical and upper thoracic region. Their location may be intraspinal, extraspinal (most commonly in the pos-

Fig. 3. Axial (left) and coronal (right) MR images demonstrating the normalcy of the rest of the central nervous system.

Fig. 4. Intraoperative photograph demonstrating the exposure of the huge sac and its pedicle.
terior mediastinum), or both, entering the spinal canal through a VB defect. In the majority of cases, an attachment to the alimentary tract is identified. Cyst contents can be clear, mucinous, whitish, or turbid liquids. Histologically, the most common finding is a pseudostratified or stratified cuboidal or columnar epithelium, mucin-secreting epithelial cells, and goblet cells lying on a basement membrane. In our case, there was no attachment to the alimentary tract. The fluid aspirated from the cyst was verified to be CSF by biochemical analyses. The cyst wall consisted of squamous cells and histological analysis showed no mucin-secreting cells.

In the light of MR images and operative findings, we think that the lesion described and its neuronal elements were a part of myelomeningocele deformity because the settlement and appearance of the neuronal elements and the abnormal structures were as seen in ordinary posterior myelomeningoceles, except for the deformed VBs. The neural elements thought to be neural placodes are clearly seen in sagittal MR images.

The many theories regarding the pathogenesis of spinal dysraphism include simple nonclosure, overgrowth and nonclosure, reopening, overgrowth and reopening, and primary mesodermal insufficiency. Primary mesodermal insufficiency seems to best describe our case. As reported by Urui and Oi, Tanimura and coworkers found an abnormal development of VB and arch as well as proliferation and dilation of the capillary vessels in the spinal cord and bleeding region in their model of spina bifida vitamin A–induced mice. They thought that the primary lesion was the abnormal differentiation of mesenchymal tissues, such as vertebral and vessel formation, and that the spina bifida was secondary. We do not know if the deformity in the bone was primary or if it was secondary to the malformed neuronal or dural/arachnoidal elements.

This case is extraordinary, not only for its rarity but also for its biological behaviors. Myelomeningoceles are anomalies usually associated with devastating problems such as hydrocephalus, multiple brain anomalies, Chiari malformation, genitourinary malformations, ophthalmological problems, central nervous system infections, and alterations in intellectual development; yet the entire evaluation of our patient revealed none of these anomalies. The only problems he complained of were dyspnea, decreased effort capacity, and kyphoscoliosis.

We do not know whether the alterations in our patient’s effort capacity was attributable to the growing retromediastinal mass of the myelomeningocele sac, leading to compression of the pulmonary tissue itself, or to his kyphoscoliotic structure, leading to pulmonary restriction. In both instances, the choice of treatment was surgery; therefore, the operation was performed to cure both pathological entities and to prevent the patient not only from experiencing future neurological complications such as hydrocephalus but also from experiencing any other respiratory problems.

Conclusions

To the best of our knowledge, this is the only case of myelomeningocele in the literature presenting as a retromediastinal mass. The case also represents one of the rare examples of myelomeningocele with no neurological deficits. Anterior thoracic myelomeningoceles should be included in the differential diagnosis of mediastinal mass lesions, even if patients do not present with neurological deficits.

References
