HIGH THORACIC CONGENITAL KYPHOSIS ASSOCIATED WITH A HIGH MEDIASTINAL NEUROENTERIC CYST (A Case Report)

SURAT A., M.D, ACAROĞLU E., M.D, YAZICI M., M.D, LEBLEBİCİOĞLU G., M.D.

A case of high thoracic kyphosis, that was found to be associated with a neuroenteric cyst located at the posterior mediastinum is presented.

Total excision of the cystic lesion immediately anterior to the vertebral column could be accomplished by a right thoracotomy and anterior as well as posterior in-situ fusions were performed for the rapidly progressing deformity.

Congenital kyphotic deformities of the spinal column located at the high thoracic area are known to be rapidly progressing lesions with dismal prognoses. The natural history of these deformities are often associated with a high rate of spinal compression and neurologic involvement. Posterior fusions alone are sufficient for neither halting the progression of the deformities, nor decompression of the spinal cord that is usually pressed to the posterior sides of the vertebral bodies. Hence anterior and posterior fusions should be the treatment of choice and should be contemplated as early as possible.

Neuroenteric cysts are a subgroup of split notocord syndromes, formed by the adhesions between the neural tube and the gut. They may be in continuity with the gut or the subarachnoid space or rarely both, and may as well form an enteric fistula. The most common locations are the posterior mediastinum and the retroperitoneal region.

Case Report:

A two and one half year old girl was brought to the our department with her parents observation of a rapidly progressing hump at the base of her neck. The deformity was noticed three months ago. The patient had begun walking by the age of thirteen months but was not fully continent as yet.

She had a small hump at her high thoracic area, no neurologic defect could be detected. Radiologic examination revealed a high thoracic konjenital kyphosis measuring 57 degrees from T1 to T5 and lower thoracic hypo-kyphosis. Suspecting of a dysraphic spine Magnetic Resonans Imaging (MRI) of the region and a control visit three months later were scheduled.

MRI revealed a huge cystic mass located at the posterior mediastinum in the vicinity of the vertebral deformity that was thought to be a neuroenteric cyst and the patient's deformity has progressed to 68 degrees during the three months. Neither myelographic examination nor the three months. Neither myelographic examination nor the upper GI series demonstrated any leakage of dye into the cystic cavity.

The decision to operate was mainly based on the rapid progression of the vertebral deformity. A right thoracotomy through the third right rib, elevating the scapula, revealed a retropleural cystic mass filled with a clear and very viscous fluid. The cyst could be thoroughly excised and an anterior fusion using the excised rib as a strut graft was performed after discectomies. Her post-operative recovery period was eventless and a posterior fusion was performed four weeks later. The patient was discharged from the hospital wearing a Minerva type cast. The cast was removed at three months follow-up visit and despite some displacement of the anterior strut graft a bony fusion was achieved.

Histologic examination of the mass revealed a cyst cavity lined up with primitive neural like elements and the diagnosis of a neuroenteric cyst was confirmed.

DISCUSSION

Congenital kyphosis is a well known clinical entity and if located at the high thoracic region carries a very high risk of cord compression and neurologic involvement. Results obtained by braces have been very unsatisfactory and early surgical fusion of the deformity is usually necesary. Posterior approaches per-se have very high rates of pseudoarthroses and are nearly always insufficient for spinal decompression. Both ante-

Hacettepe University School of Medicine Dept. of Orthopaedics and Traumatology

rior and posterior fusions and if nececesary anterior decompression should be the treatment of choice.

Congenital deformities of the vertebral column are frequently associated with dysraphic choanges of the spinal cord, so MRI examination has long been a standard preoperative procedure. As can be seen from our case it has proven helpful in delineating other possible abnormalities, that may prove very difficult to deal with if not prepared.

Whether the association of the cyst and the vertebral congenital deformity is purely incidental or if there is a cause-effect relation between the two is very hard to decide. Neuroenteric cyts are thought to be caused by adhesions between the neural tube and the endoderm that will form the fore-gut and these adhesions might probably have caused formation defects at the anterior portions of the somites.

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