

Cervico-Thoracic Neurenteric Cyst - A Case Report

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Neurenteric cysts are uncommon tumors of developmental origin. They are asymptomatic in infants, but can present with cord compression in the adolescent and young patient. A large cervico - thoracic neurenteric cyst extending to superior mediastinum in a child aged one and half years is discussed. The mass showed histological features of neuroenteric cyst with teratomatous elements.

Case report

A one year and four months old girl presented with history of fever, cough and cold at the age of seven months, which lasted for 15 days. During that period, parents noticed deviation of the neck towards the left, which was progressive for a few days and then remained static. General and neurological examination revealed no significant findings except torticollis to left, sluggish tendon reflexes.



1a



1 a and b: Sagittal T2 and T1 weighted images of the cyst loculations and thoracic components are documented.



1) C) : T2 weighted coronal Image demonstrating the posterior mediastinal component of the cyst.

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Frontal, lateral radiographs of the cervical spine and supine radiograph of the chest showed a large soft tissue density mass occupying the upper two thirds of the right hemi thorax and a butterfly vertebra at C5 level.

MR of the cervical and dorsal spine showed a large multi loculated cystic lesion in the pre and para vertebral regions extending from C5-D8 level measuring 8.4 x 4.3 x 6.7 cm. The cyst was compressing mediastinal structures

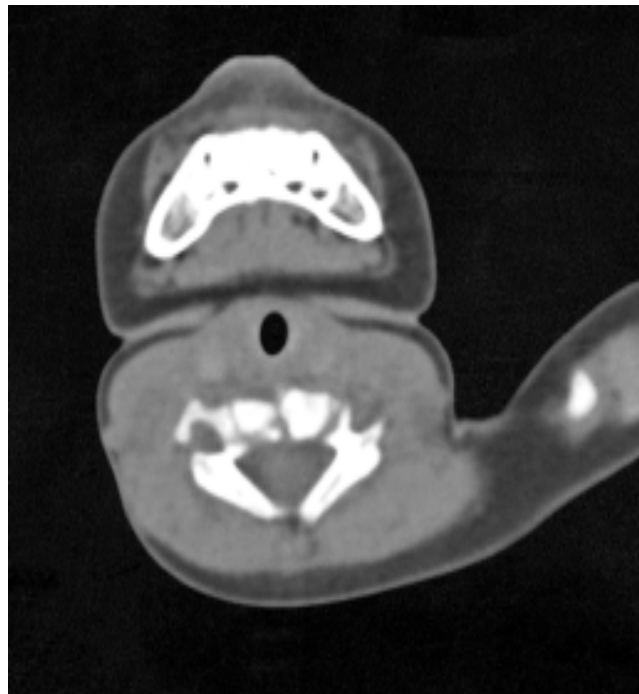
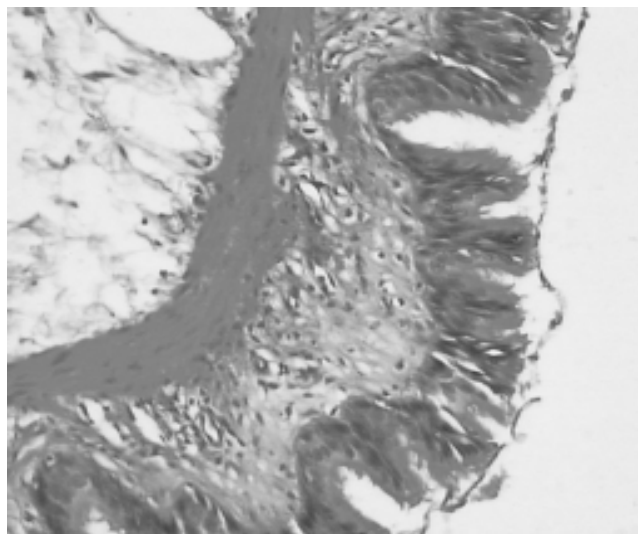


Fig 2 a) Axial CT image of C5 vertebral body showing butterfly vertebra
b) Sagittal reconstruction of the CT thorax showing the calcification within the cyst.

and the right main bronchus was elevated over the mass. The cyst extended intra spinally into the subarachnoid space through a foramen in the C5 vertebral body and was seen to be in contact with the cord. The cyst was hypo-intense on T1W1 and hyper intense on T2W1. Small intramedullary hyper intensity was noted in the cord at C4-C5 level. The cyst wall showed mild enhancement. There was a 1cm x 0.4cm tubular calcification in the cyst at D1 vertebral level on the right postero-lateral aspect of trachea.



3) Paraffin section of neuroenteric cyst showing respiratory epithelium with underlying muscularis mucosa (hematoxylin and Eosin x 400)

The child was operated in two stages by Neurosurgical and Cardiac surgery teams and the cyst was completely excised. A loculated cyst in the prevertebral region of the neck, containing thick cheesy material, calcified sub mucosal wall was noted at surgery. In the thoracic area, the cyst was retro pleural on right side extending to left side across the mid line, cyst contents were cloudy. The cyst communicated with the anterior subarachnoid space at C5 level through an opening in the body of C5 vertebra. There was thick submucosal calcification within this cyst.

Histopathological studies of cervical component revealed a cyst lined by ciliated, tall columnar epithelium leading to transitional to squamous epithelium. The submucosal area had muscularis mucosae of variable thickness with a nodular island of nerve twigs and large dystrophic neurons, submucosal calcification was detected. Thoracic component showed cysts lined by respiratory epithelium (endodermal derivative). Squamous epithelium (ectodermal) and focal bone, marrow and cartilage islands representing mesodermal components.

Overall features were compatible with neuroenteric cyst type III of Wilkin and Odome classification.

Discussion

Neurenteric cysts also called endodermal cysts, Enterogenous cysts, enteric cysts, are rare congenital cysts of the spine lined with endoderm. They result from failure of separation of ectoderm from the endoderm during the third week of embryonic life with persistent canal of Kovalevsky. The nature of the eventual abnormality depends on the extent to which this adhesion subsequently disappears. Persistence of the entire tract results in the extreme form of combined anterior and posterior spina bifida with dorsal enteric fistula. Persistence of only a part of the tract produces the isolated intraspinal cyst. The most common location is the cervico-dorsal region; mediastinal cysts are reported with a mainly right paravertebral location and ventral to the spinal cord (1). They are associated with vertebral anomalies like lipoma diastomatomyelia gut cysts, bowel duplication. Neurenteric cysts are also considered to be a form of occult spinal dysraphism, as are lipomas, lipomeningomyeloceles, dermoids epidermoid, terminal syringohydromyelia and myelocystocele. The age of presentation ranges from 5 wks to 52 years (3).

These cysts infrequently cause symptoms, usually the large cysts are asymptomatic. Adolescents and young adults present with intermittent progressive radicular pain worsened by elevating the intraspinal pressure leading to myelopathy and long tract signs. These cysts are usually smooth, unilocular, most commonly located at the lower cervical or upper thoracic region rarely in the lumbar region (4). The cysts uncommonly occur intracranially where they are located in the pons, cerebello pontine angle cistern., or rarely in the foramen magnum (5).

Neurenteric cysts are intradural extramedullary lesions situated ventral or ventrolateral to the cord, rarely dorsal or dorsolateral to the cord or within the cleft of diastematomyelia (6). Intramedullary component is seen in 10% of cases.

Plain radiographs may reveal widening of the spinal canal at the site of the cyst along with associated skeletal anomalies and scoliosis. CT scan shows the associated congenital anomalies of the spine like spina bifida, butterfly vertebra, lack of segmentation, partial fusion, scoliosis together with the cyst. In some cases only enlarged spinal canal can be seen. MRI demonstrates the exact site /

relationship of the cyst to the spinal cord, site of intraspinal attachment of the cyst. The cysts are iso-intense to CSF on T1WI and T2WI, if the cyst contents are clear like CSF. If the cyst contents are proteinaceous the cyst contents will be hyperintense on T1WI. The cyst in our patient was having CSF intensity in all pulse sequences.

Though enterogenous cysts are typically endodermal lined cysts, histological variants were described and classified by Wilkins & Odome.

Histological classification of enterogenous cyst (modified after Wilkins and Odome)

Type I Single pseudostratified, cuboidal or columnar epithelium with or without cilia lying on a basement membrane.

Type II Similar to type I with addition of mucous glands, serous glands, smooth muscle, fat, cartilage, bone, elastic fibres, lymphoid tissue or nerve ganglion.

Type III Similar to type II with addition of ependymal or glial tissue.

The lesion in the present discussion was mainly extra spinal with small intra spinal component, presenting incidentally. The lesion was multilocular and there was mild enhancement of the walls. Histologically, there were epithelial, neuronal, mesodermal (cartilage elements). This cyst belongs to type II or "teratomatous", according to the modified Wilkins and Odome classification (7).

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