Thoraco-abdominal neuroenteric cyst: A case report and review of the literature

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Neuroenteric cysts are rare congenital anomalies, presenting in infancy or childhood. Only a few cases of posterior mediastinal neuroenteric cysts have been reported.

A 12-year-old girl presented to our hospital with a neglected mildly painful cyst in the posterior mediastinum and retroperitoneum. Magnetic resonance imaging revealed scoliosis of the thoracic spine, hemivertebrae T6 – 7, and a large hyperintense (on T2-weighted images) cystic lesion in the posterior mediastinum. The lesion had no intraspinal communication, but extended into the abdomen between the liver and kidney. Despite the long history and spatial extent of the lesion, it was excised uneventfully through a right thoracotomy. Microscopy of the cyst wall revealed partially disrupted cuboidal epithelium, muscle fibres, inflammatory cells, and, bony and cartilaginous elements.

Enterogetic duplication cysts of foregut origin with vertebral anomalies are referred to as neuroenteric cysts. This is probably only the second reported case of thoraco-abdominal neurogenic cyst originating in the posterior mediastinum.

Neuroenteric cysts are rare congenital anomalies, with only about 30 cases reported in the literature. The enterogenous type of duplication cyst, when associated with vertebral anomalies, is known as a neuroenteric cyst. These cysts present mostly in the first year of life, and the location varies from intracranial to abdominal. Posterior mediastinal neuroenteric cysts are very rare. Using the MESH keywords 'neuroenteric cyst' and 'mediastinal', we could identify only 5 such studies in the English medical literature.

We report on a 12-year-old girl with a neglected neuroenteric cyst of the posterior mediastinum and retroperitoneum, which was excised completely through a right posterolateral thoracotomy. This is probably the second case of its kind in the available literature.

Case report

A 12-year-old girl was brought to the surgery clinic at Guru Tegh Bahadur Hospital, Delhi, India, with a 3-month history of moderate dull pain in the front and back of the right lower chest. There were no gastro-intestinal, respiratory or cardiac symptoms. The patient had been investigated for anaemia and malnutrition 6 years previously. A computed tomography (CT) scan of the thorax done at that time had revealed a cystic lesion in the posterior mediastinum, and surgery had been advised; however, the patient’s parents had refused the operation for socio-economic reasons. General physical and systemic examination at the second presentation was unremarkable. The results of haematological and biochemical investigations were normal. A chest radiograph (Fig. 1) suggested scoliosis of the thoracic spine with hemivertebrae (T6 and T7). T2-weighted magnetic resonance images of the chest and abdomen (Fig. 2) revealed a hyperintense lesion in the posterior mediastinum.
An elongated cystic mass in the posterior mediastinum and abdomen. The mass was seen extending from T4 to T5 cranially and to a cleft between the right kidney and liver caudally. All tissue planes with oesophagus, liver and kidney were well maintained. The spinal cord was normal and no intraspinal communication with the cystic lesion was identified. Hemivertebrae of T6 and T7 were seen.

Complete excision of the cyst was performed, and the lesion was exposed through a right posterolateral thoracotomy. At operation, a cystic mass was seen in the posterior mediastinum abutting the oesophagus, extending caudally through a defect in the right dome of the diaphragm to a cleft between the liver and the right kidney. The entire cyst was easily separable from the surrounding structures, and it contained clear straw-coloured fluid. It was excised intact.

Histopathological examination (Figs 3a and 3b) revealed a cyst with a partially denuded cuboidal epithelial lining. Outside this were two layers of muscle fibres at right angles to each other. Elements of bone, cartilage, cholesterol clefts and inflammatory cells were identified in a few places. A final diagnosis of type II neuroenteric cyst (Wilkins and Odom classification) was made. The patient’s postoperative recovery was satisfactory, and she was well at follow-up 3 months later.

Discussion

Foregut duplication cysts are currently classified into two broad subtypes – bronchial-oesophageal cysts (formerly classified as bronchogenic and enterogenous separately) and neuroenteric cysts. The former develop as a result of sequestration of a respiratory bud during development or separation of the tracheobronchial tree from the oesophagus. They are usually lined by ciliated epithelium of the primitive oesophagus or trachea, and may contain cartilage and/or glands within the walls. Neuroenteric cysts are foregut duplication cysts that are associated with apparent vertebral anomalies. The development is related to incomplete separation of the notochord from the embryonic foregut or herniation of the endoderm of the embryonic foregut into the dorsal ectoderm. This attachment may prevent fusion of the vertebral bodies and lead to spinal anomalies.

In 1976 Wilkins and Odom classified enterogenous cysts into three types: type 1 has a single pseudostratified cuboidal or columnar epithelium with or without cilia, lying on a basement membrane; type 2 is similar to type 1 with the addition of mucous glands, serous glands, smooth muscle, fat, cartilage, bone, elastic fibres, lymphoid tissue or nerve ganglion; and type 3 is similar to type 2 with the addition of ependymal or glial tissue. This classification has been used by most authors until now, however, some have contested the presence of cartilage in type 2 cysts, suggesting that these may be more appropriately termed teratomas. In our case there were features of a classic type 2 neuroenteric cyst. Owing to the presence of bone and cartilage, some pathologists may refer to our case as a teratomatous neuroenteric cyst. Another unusual histological feature was the absence of the mucosal lining in the cyst wall. This is probably due to recurrent infections or pressure atrophy in the cyst.

Neuroenteric cysts usually present in infancy or early childhood. The location is usually in the central nervous system, often intraspinal. Ten per cent of intraspinal cases are intramedullary. These patients may present with neurological symptoms, and excision often requires a neurosurgical team. In cases where there is suspicion of a neuroenteric cyst on plain X-rays or a CT scan, magnetic resonance imaging (MRI) is essential because spinal elements in close relation to the lesion are delineated clearly only with MRI. In our patient MRI was complementary to CT to rule out communication with the spinal canal. Posterior mediastinal neuroenteric cysts have been reported very infrequently (see above). Of these patients, only 2 had abdominal involvement. One was a 24-year-old woman with a suspected recurrent pancreatic cystic neoplasm. After excision, histopathological examination of the specimen revealed embryonic foregut and features typical of a neuroenteric cyst. The second patient had a prenatally detected posterior mediastinal cyst traversing the
diaphragm and communicating with the jejunum. The lesion was removed through a thoracic incision. Posterior mediastinal cysts may grow to large sizes before detection, owing to lower resistance to growth. For the same reason, they are often asymptomatic. Symptoms may pertain to pressure effects or to infection within the cyst.

Surgical excision is indicated in all cases, in order to avoid complications. The lesion is usually removed through a thoracotomy. It is also advocated that, in case of adherence of the cyst wall to the oesophagus or spine, a partial cystectomy with ablation of the epithelium should be performed, leaving the adherent portion in situ. This is akin to the approach for other benign cysts such as choledochal cysts. We were surprised to achieve complete removal with unhindered dissection in our patient, in spite of the long duration (about 7 years) and complicated anatomy of the cyst. With the advent of thoracoscopy, such lesions might be amenable to thoracoscopic excision. Our case is the second reported case of an abdominothoracic neuroenteric cyst removed by a thoracic incision.

References