A giant gastroenteric cyst associated with pectus excavatum and compression of the thoracic duct: A case report

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Gastroenteric cysts are unusual developmental cysts encountered in the posterior mediastinum of patients. They are usually lined by alimentary (squamous or enteric) epithelium and contain gastric mucosa and, rarely, pancreatic tissue. Gastroenteric cysts are usually associated with vertebral abnormalities and less often with complete situs inversus.¹ We present a case of a gastroenteric cyst with pectus excavatum and compression of the thoracic duct.

Clinical Summary

A 6-month-old girl was admitted to the pediatric emergency unit with fever, cough, and dyspnea. On her past medical history, the presence of such a cyst in the right hemithorax had been demonstrated at the intrauterine 32nd week by means of ultrasonography (Figure 1, A), and the cyst had been aspirated to diminish its pressurizing effect. The patient was free of symptoms until 6 months of age. Physical examination on presentation revealed cyanosis, tachypnea, intercostal retractions, severe pectus excavatum, and diminished breath sounds and fine rales in the right hemithorax. The hemoglobin level of the patient was 10.5 g/dL, the leukocyte count was 19,900/mm³, the thrombocyte count was 840,000/mm³, the erythrocyte sedimentation rate was 12 mm/h, and the C-reactive protein level was 20.5 mg/L. Arterial blood gas analyses revealed moderate hypoxemia and decreased oxygen saturation. Blood biochemical measurements were within normal limits. Posteroanterior and lateral chest roentgenograms showed homogenous opacity localized in the right hemithorax and pectus excavatum (Figure 1, B and C). Thorax computed tomography revealed a cystic mass almost filling the right hemithorax (Figure 1, D). Through a right thoracotomy, a huge cyst, approximately 6 × 12 cm in size, was identified in the posterior mediastinum adjacent to the esophagus. In addition, the thoracic duct was deemed to be atrophic because of the compression of the cyst and

the proximal end. The distal ends of the thoracic duct were dilated, and therefore the cyst was thought to be a cystic hygroma. The cyst was totally excised (Figure 2, A). On histologic examination, a gastric mucosa was seen within the cyst (Figure 2, B). Nine months after surgical intervention, the patient remained well.

Discussion

Mediastinal cystic lesions constitute approximately 18% of all mediastinal lesions in children. Congenital foregut cysts of the mediastinum are extremely rare lesions.² Sirivella and colleagues³ reported 4 enterogenous cysts among 20 patients with foregut cysts of the mediastinum. Eighty percent of the patients with foregut cysts were symptomatic in varying degrees of severity. Symptoms included dysphasia, pyrexia, cough, hematemesis, weight loss, retrosternal chest pain, and recurrent pneumonia.³ Our patient was symptomatic as well. Gastroenteric cysts are associated with vertebral abnormalities, such as scoliosis, anterior spina bifida, hemivertebrae, butterfly vertebrae, or vertebral fusions. We did not observe vertebral abnormalities; however, the patient had pectus excavatum.

Chest radiography, computed tomography, and magnetic resonance imaging are enough for the demonstration of a gastroenteric cyst. In addition, bronchoscopy and esophagoscopy might be helpful for the diagnosis. Barium swallow effectively pinpoints an extrinsic compression of the esophagus causing dysphasia.¹⁻³ Congenital foregut cysts might be diagnosed on the basis of prenatal ultrasonography, as in our patient.⁴

Surgical excision of these cysts is indicated. Optimal timing for operation is dictated by the severity of symptoms.³ When an operation is not possible, bronchoscopic or thoracoscopic needle drainage might be alternative treatment procedures. However, these practices can cause severe complications, and in addition, the risk of malignant transformation will still remain.⁵ We preferred right thoracotomy for complete surgical removal of the cyst.

In this article we reported the first case, to our knowledge, of a patient presenting with a congenital gastroenteric cyst associated with pectus excavatum and compression of the thoracic duct. Although the cyst was aspirated during intrauterine life in an effort to decrease the compression of the cystic fluid, this procedure seemed to be unsuccessful because it resulted in atrophy after long-term compression of the thoracic duct. Therefore such cysts, whatever their size, should be removed as soon as possible before symptoms are evident.

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Figure 1. *A*, Prenatal ultrasonogram showing a cystic lesion in the right hemithorax. *B*, Posteroanterior chest radiograph. *C*, Pectus excavatum and the cyst were evident on the lateral chest radiograph. *D*, Chest computed tomographic scan showing a giant cystic mass

Figure 2. *A*, Macroscopic appearance of the totally removed giant cyst (6 × 12 cm). *B*, Microscopic appearance of the cyst wall showing all layers of the gastrointestinal mucosa. (Hematoxylin and eosin staining, original magnification 40×).

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