CASE REPORTS

Mediastinal Enteric Cyst: Unusual Clinical Presentation and Histopathology


Institut Clínic de Pneumologia i Cirurgia Toráctica, Hospital Clínic, Barcelona, Spain.

A mediastinal enteric cyst is an uncommon entity which is rare in adults and usually found incidentally. In most cases the lesion is lined by gastrointestinal mucosa, and theories as to the origin of such lesions are diverse. We report an adult case of thoracic enteric cyst that presented with cardiac tamponade and for which histopathological examination revealed the presence of pancreatic tissue. Review of the literature yielded only 1 case of mediastinal enteric cyst with pancreatic tissue.

Key words: Mediastinal enteric cyst. Cardiac Tamponade. Pancreatic tissue.

Introduction

Because both the respiratory and digestive tracts develop from the primitive foregut, cysts derived from them are considered to have a common enteric origin, whether they are specifically bronchogenic, enteric, or duplication cysts. Such lesions are malformations produced during the stage of differentiation and embryologic development of the primitive intestine. Enteric cysts in the thorax account for 20% of digestive tract duplications. Their structure consists of a smooth muscle layer and a gastrointestinal mucosal lining. Sixty percent of enteric cysts are diagnosed in patients younger than 1 year and usually present with respiratory symptoms. The incidence is lower in adults, and the symptoms are less specific. The treatment of choice is radical excision of the lesion. We report the case of an adult with an enteric cyst that presented with severe, atypical signs and symptoms. The histopathological findings were also unusual.

Case Description

The patient was a 44-year-old man denying tobacco, alcohol, or street drug use and reporting no contributory medical history. He came to the emergency department because of epigastric pain and sudden dyspnea followed by central chest pain not described as tightness. Upon arrival the patient was hypotensive with a heart rate of 105 beats per minute and breathing rate of 24 breaths per minute. He presented with sudden hemodynamic instability with hypotension (80/20 mm Hg), cold sweats, and pallor. On physical examination jugular venous distention with symmetric peripheral pulses was noted. A chest x-ray demonstrated widening of the mediastinum mainly on the left side, with blurring of the aortic knob and left pleural effusion. On suspicion of aortic aneurysm, a contrast echocardiogram was performed. The image revealed pericardial effusion with impairment of the right cavities and well-defined fluid accumulation above the heart; cardiac chamber filling was normal, and no communication with the pericardial space or the cystic lesion was evident. The computed tomography scan of the chest (Figures 1 and 2) showed an image of a 10×8×7.5 cm cyst extending from the supra-aortic branches to the upper part of the left ventricle. The cyst occupied the left side of the anterior mediastium and appeared as a mass molded around the vascular structures. After administering contrast, a vascular origin of the lesion was ruled out. Given the hemodynamic instability of the patient, pericardiocentesis
was performed, and 500 mL of dark serous fluid was extracted; subsequently, the patient improved. Gram and Ziehl-Neelsen stains and culture of the fluid were negative; biochemical analysis showed adenosine deaminase, 35 U/L; lactate dehydrogenase, 1102 mg/dL; glucose, 122 mg/dL, and proteins, 52 mg/dL. The diagnosis was anterior mediastinal cyst ruptured at the pericardial cavity with severe pericardial effusion and cardiac tamponade. Surgery was scheduled for both diagnostic and therapeutic purposes, and at 72 hours the intervention was carried out by median sternotomy. Pericardial enlargement and a soft cystic tumor containing chocolate-brown fluid and adhering to the anterosuperior and middle left mediastinum were observed. On opening the pericardium fibrinous pericarditis and a dark fluid with some clotting were noted. The lesion was excised with the phrenic nerve left intact. During the immediate postoperative recovery period the patient developed progressive hypoxemia, nonhypercapnic respiratory insufficiency (pH, 7.48; PaO₂, 55 mm Hg; PaCO₂, 34.2 mm Hg; bicarbonate concentration, 25.2 mmol/L; fraction of inspired oxygen, 50%), bibasilar crackles, and bilateral lung infiltrates. Acute respiratory distress syndrome was diagnosed, and noninvasive mechanical ventilation was initiated with good results (arterial blood gases: PaO₂, 135 mm Hg; PaCO₂, 41 mm Hg; pH, 7.46; fraction of inspired oxygen, 50%). Ventilation was withdrawn on day 5. The pathology report referred to an enteric cyst containing ectopic pancreatic tissue, a multiple-lobed glandular structure, and ducts excreting serous-type cells. Lobe interiors contained cellular clusters with endocrine differentiation reminiscent of pancreatic tissue and forming islet-like structures.

Discussion

Enteric cysts (or duplication cysts, a term first used in 1711 by Blasius and Bremer) are a group of rare congenital malformations consisting of a smooth muscle wall and gastrointestinal-like mucosal lining, a gastric mucosal lining being the most common. Enteric cysts in the chest represent 7% to 20% of all enterogenic cysts. Theories as to the origin of enteric cysts have referred to an anomaly in primitive foregut development during the embryonic stage (set forth in 1929 by Meyenburg and in 1937 by Böss). The most accepted theory for enteric cysts involves the split notochord syndrome, in which persistence of the neuroenteric band occurs. Enteric cysts are more common in children, tend to appear at the right posterior mediastinum, and present clinically as respiratory distress. Such cysts are less common in adults and diagnosis is usually incidental or is suggested by insidious respiratory symptoms with fever and chest or epigastric pain. Enteric cyst is associated with pericardial and vertebral anomalies (spina bifida, hemivertebrae, or vertebral fusion) with the latter occurring in up to 50% to 80% of cases. The triad of a posterior mediastinal mass, a right-sided location, and a vertebral defect suggests a diagnosis of enteric cyst in more than 70% of cases. One complication described in the literature is cyst ulceration and perforation, which is more frequent in those cysts with an inner lining of gastric mucosa. Patients present with hematemesis or hemoptysis, depending on whether the cyst communicates with the esophagus or an airway (as reported in 1937 by Böss, in 1938 by Seydl, and in 1951 by Pohlmann). Given the risk of complications in severe cases, treatment for such lesions is radical surgical excision. Our patient presented with a clinical picture of cardiac tamponade related to cyst perforation which we could attribute neither to an infection (all microbiological studies being negative) nor to ulceration of the cyst wall by enzymatic activity as no gastric mucosa was observed during inspection of the tissue. Pancreatic tissue in the mediastinum is rare and we have found only 3 cases of pancreatic tissue ectopia in this region described in the literature. It does, however, appear more frequently in the abdominal region, along the digestive tract. After reviewing the literature we found a single case of enteric cyst with gastric mucosa associated with pancreatic tissue. The reason why pancreatic tissue appears in the mediastinum is not clear: it may be related to heteroplasia or to pancreatic cell...
In our patient, this finding might have been such a case of heteroplasia, as stated above, or pancreatic tissue heterotopia over a digestive structure (an observation in 2% of autopsies). We have reported this case because of the unusual histology and because of the serious and exceptional nature of presentation, which has not been described in the searchable literature until now.

REFERENCES