

Rapid growing mediastinal ectopic pancreas within ruptured thymic cyst treated using video-assisted thoracic surgery

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Abstract

Ectopic pancreas (EP) is typically found within other gastrointestinal organs. Its discovery in other parts of the body, especially in the mediastinum, is exceedingly rare. This paper presents a case of a 17-year-old female patient with EP in a large, rapidly growing thymic cyst. She presented to our institution with persistent chest pain. Video-assisted thoracic surgery revealed a mediastinal mass in the setting of pleural effusion. Analysis of the fluid contents of the mass and the pleural effusion demonstrated high levels of pancreatic amylase, which supported the presence of pancreatic tissue within the mass. This is the first reported case of EP in a thymic cyst with an active pancreatic exocrine function. It is also the first reported case of mediastinal EP rupture secondary to autodigestion by amylase.

KEYWORDS

exocrine pancreas tissue, mediastinal ectopic pancreas, thymic cyst, VATS

INTRODUCTION

Ectopic pancreas (EP) is defined as pancreatic tissue that grows outside of its normal location and is characterized by a lack of vascular or anatomical connections to the main pancreas.¹ Most EP tissues are found within other gastrointestinal organs, such as the stomach, duodenum, jejunum and ileum. EP is a developmental anomaly found in 2% of all autopsies. They are discovered in the gastrointestinal tract in 70%–90% of the time.² To the best of our knowledge, only 30 cases of EP in thymic cysts have been reported so far in the literature.³ This paper is the first to demonstrate that mediastinal EP may be associated with rapid growth and an increased risk for rupture, especially if the pancreatic exocrine function is preserved.

CASE REPORT

A 17-year-old female presented at another hospital with right chest pain for a week. Chest radiography (CXR) revealed right pleural effusion (Figure 1A). Comparatively, CXR performed 16 months prior had shown no abnormalities

(Figure 1B). Contrast-enhanced chest computed tomography revealed a large, cystic mass with enhancing components in the anterior mediastinum as well as passive atelectasis in the right lower lobe and pleural effusion (Figure 1C,D). Blood tests revealed a white blood cell count of 10,100/ μ l (normal range 3300–8600) and a C-reactive protein level of 6.2 mg/dl (normal range 0.0–0.1). At this time, our differential diagnoses included a ruptured teratoma, a thymoma with cystic changes and a haemorrhagic thymic cyst. We subsequently performed video-assisted thoracic surgery (VATS). Intraoperatively, we discovered a large, thymic cyst with thickened walls in the anterior mediastinum, as well as pleural effusion composed of black-coloured fluid (Figure 2A). We punctured the cyst using an 18-G needle to decrease its size and widen the thoracoscopic view (Figure 2B). The part of the cystic wall punctured was immediately sutured to prevent the leakage of inside fluid. The cyst was thoroughly dissected from the anterior mediastinum. There was little intraoperative bleeding. The operating time was 186 min. The post-operative course was unremarkable, and the patient was discharged on the fourth post-operative day.

Pathological examination of the surgical specimen showed ectopic pancreatic tissue within a hyperplastic

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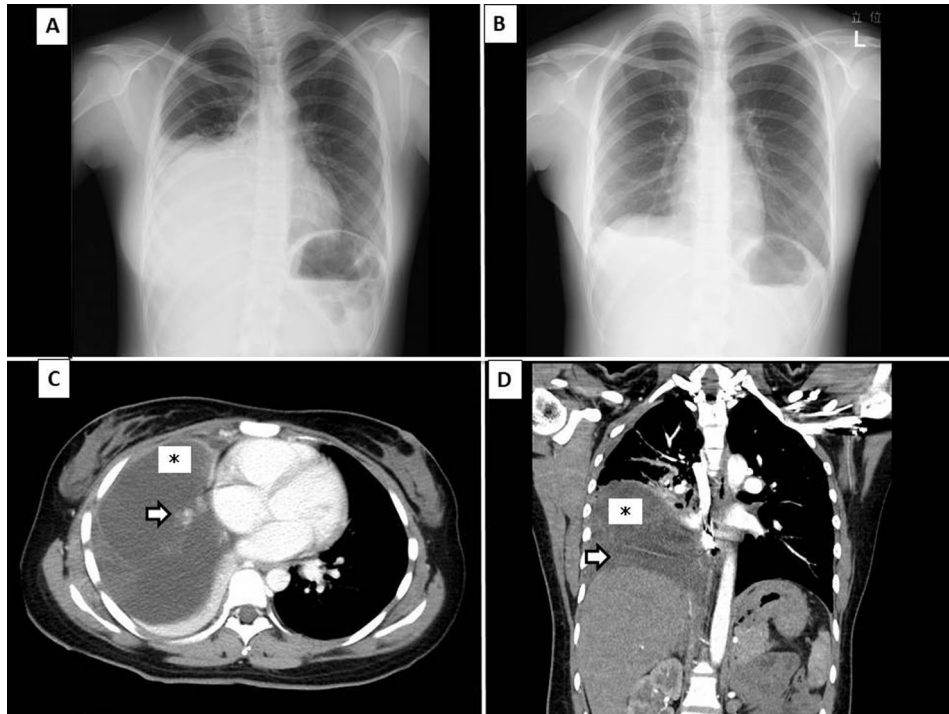


FIGURE 1 (A) The chest radiograph (CXR) taken on admission. The CXR demonstrates significant right pleural effusion. (B) The CXR taken 16 months before admission. The CXR shows no abnormalities. (C) A chest computed tomography (CT) image, axial section, taken on admission. The white arrow (\Rightarrow) points to the nodular enhancement along the cyst walls. The asterisk points to (*) the thymic cyst. (D) A chest CT image, coronal section, taken on admission. The CT image shows a large cyst with nodular wall enhancement and significant pleural effusion. The fluid in the mediastinal cyst and the pleural effusion show similar hypodensity. The asterisk points to (*) the thymic cyst. The white arrow (\Rightarrow) points to the pleural effusion

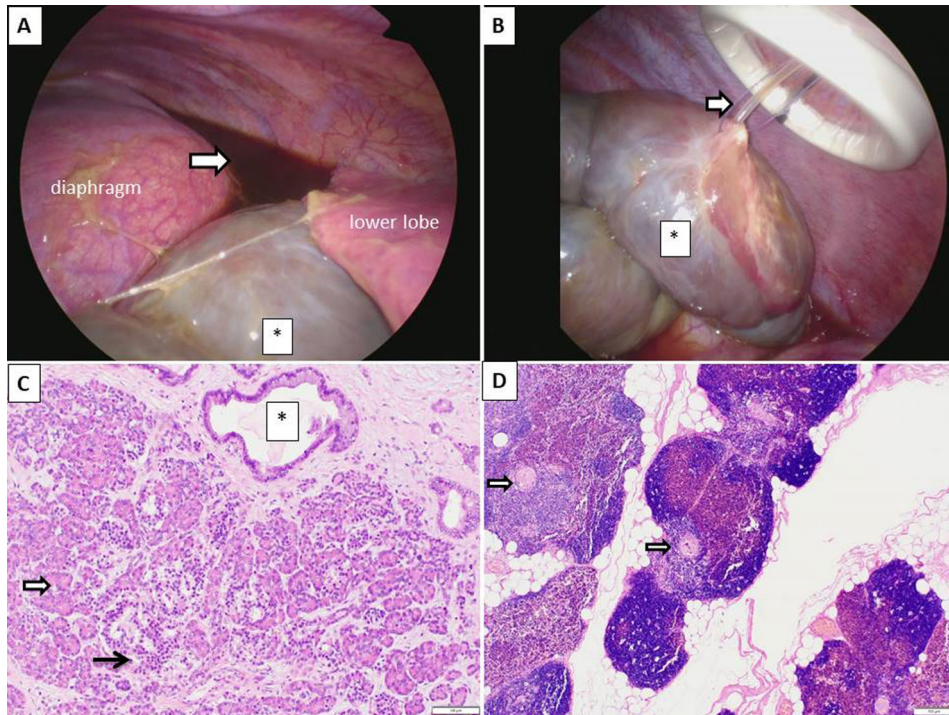


FIGURE 2 (A) An intraoperative photograph of the right thoracic cavity. Pleural effusion with black-coloured fluid is noted. The same black-coloured fluid drained from within the cyst. The white arrow (\Rightarrow) points to the pleural effusion. The asterisk (*) points to the thymic cyst. (B) An intraoperative photograph of the right thoracic cavity. An 18-G needle was used to aspirate some of the cystic fluid. The white arrow (\Rightarrow) points to the 18-G needle and the suturing stitch. The asterisk (*) points to the thymic cyst. (C) Pathological findings demonstrate ectopic pancreas tissue within the thymic cyst. The white arrow (\Rightarrow) points to an acinar cell. The black arrow (\rightarrow) points to islets of Langerhans (scale bar: 100 μ m). (D) Pathological findings demonstrate thymus hyperplasia with Hassall's corpuscles. The white arrows (\Rightarrow) point to Hassall's corpuscle (scale bar: 200 μ m)

thymus cyst (Figure 2C,D). Biochemical analysis of the cystic fluid revealed a pancreatic amylase level of 11,364 IU/L (normal range in blood, 18–53).

DISCUSSION

Mediastinal EP usually presents with symptoms such as chest pain, dyspnoea and bleeding. The severity of these symptoms was initially thought to be dependent on the site and size of the mass, as well as the degree of pressure applied by the mass onto the neighbouring organs.⁴ In this particular case, the patient presented with chest pain secondary to a ruptured thymic cyst. We theorized that the cyst wall ruptured because of the amylase produced by the EP. Our intraoperative findings supported this because the contents of the cyst and the pleural effusion were similar on visual inspection and pathological examination. This is the first reported case of mediastinal EP found in a ruptured thymic cyst. Previous reports on autodigestion by EP secretions have only been observed in teratomas.⁵

Moreover, this study was able to document the growth rate of this mediastinal tumour. Comparing both CXR images showed that the mass grew by approximately 10 cm within 16 months. This growth rate is seldom observed in EP found within the gastrointestinal tract. It is possible that the negative intrathoracic pressure allowed the cyst to expand into the thoracic cavity, whereas cyst growth in the gastrointestinal tract is usually limited by the surrounding abdominal organs.

During VATS, we discovered that the mass comprised the majority of the chest cavity. We aspirated some fluid from the mass to decrease its size and provide us with a wider operating field. The puncture site was sutured to prevent further spillage of the cystic contents into the chest cavity. Furthermore, while most reported that mediastinal EP excisions were performed via thoracotomy, we chose to perform VATS as it was less invasive.

Mediastinal EP is extremely rare, and documenting such cases is essential. Physicians must have a high index of suspicion, especially in the case of rapid growth and possible rupture.

CONFLICT OF INTEREST

None declared.

ETHICS STATEMENT

Appropriate written informed consent was obtained for publication of this case report and accompanying images.

AUTHOR CONTRIBUTIONS

Kazuki Sugata and Koichiro Kajiura drafted this manuscript. Kazuki Sugata, Koichiro Kajiura, Haruki Taniguchi and Taizo Fukumoto treated the patient. Tomoya Kuda performed the primary treatment. Akiko Matsuzaki demonstrated pathological findings.

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