Resection of anterior mediastinal ectopic pancreas by right thoracoscopy: A case report

HONGLIN ZHAO*, ZIHE ZHANG*, XIN LI, JINGHAO LIU, HONGBING ZHANG, GANG CHEN and JUN CHEN

Department of Lung Cancer Surgery, Tianjin Key Laboratory of Lung Cancer Metastasis and Tumor Microenvironment, Tianjin Lung Cancer Institute, Tianjin Medical University General Hospital, Heping, Tianjin 300052, P.R. China

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Abstract. Ectopic pancreas is uncommon in the anterior mediastinum. Herein, a 32-year-old woman presented to our institution for investigation of an abnormal mediastinal shadow on chest computed tomography. The patient underwent complete surgical resection of the anterior mediastinal mass by right thoracoscopy, and the postoperative pathology examination confirmed the diagnosis of ectopic pancreas. There were no clinical signs of pancreatitis. No recurrence or metastasis was observed during a follow-up period of 3 years. English language medical literature was also searched in order to identify other case reports describing this rare condition and identified 17 papers describing 20 cases of anterior mediastinal ectopic pancreas, which were all confirmed at surgery. All clinical characteristics in these cases were reviewed.

Introduction

Heterotopia of pancreatic tissue is a developmental anomaly found in approximately 2% of all autopsies, but pancreatic tissue within the thorax and mediastinum is much less common (1). Because both the respiratory and digestive tracts are derived from primitive foregut, congenital cysts, whether bronchogenic, enteric, or duplication cysts, are considered to have a common enteric origin and can be regarded as malformations produced during 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. Only a few cases have been reported in the English language literature, all of them arising from the anterior mediastinum.

Correspondence to: Professor Gang Chen or Dr Jun Chen, Department of Lung Cancer Surgery, Tianjin Key Laboratory of Lung Cancer Metastasis and Tumor Microenvironment, Tianjin Lung Cancer Institute, Tianjin Medical University General Hospital, 154 Anshan Road, Heping, Tianjin 300052, P.R. China
E-mail: chengang450@163.com
E-mail: huntercj2004@yahoo.com

*Contributed equally

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Case presentation

A 32-year-old woman was admitted for evaluation of an anterior mediastinal mass, which was incidentally identified on a chest X-ray. The mass was positioned above the aortic arch. Pre-contrast chest computed tomography (CT) showed a 3.6x3.9 cm cystic-solid mass situated mainly on the left side of the anterior mediastinum. The wall of the cystic lesion showed moderate contrast enhancement, while the solid part of the lesion did not show marked enhancement (Fig. 1). Mindful that the patient was a young woman, we decided, after a full preoperative workup, to perform surgery via thoracoscopy and to excise the tumor. The tumor was completely resected via a right thoracoscopy so as to protect the superior vena cava and the left innominate vein. We found that the tumor was firmly adherent to the pericardium and the left pleura and was connected by a tenuous bridge of tissue to the thymus. Grossly, the mass measured 7x4x1.5 cm and was part solid and part cystic. Postoperative recovery was uneventful and the patient was discharged on postoperative day 7. After 3 years of follow-up, there was no evidence of recurrence (Fig. 1).

Histological examination of the mass showed that the cystic wall was composed of fibrous connective tissue lined with columnar epithelium, and immunohistochemical staining was positive for both CK19 and CK20. Pancreatic acini and islet cells were present, and immunohistochemical staining indicated positivity for alpha-ACT and islet cell insulin (Fig. 2). Therefore, the final pathology diagnosis was ectopic pancreas in the mediastinum with cystogenesis.

Discussion

Ectopic pancreas is the presence of pancreatic tissue outside of the normal pancreas that has no vascular or anatomical connections to the original organ but contains acinar cells and ducts and islets of Langerhans. Ectopic pancreas is usually found in the stomach, duodenum, jejunum, or ileum; it may also be found in the gallbladder, esophagus, common bile duct, spleen, and mesentery. However, ectopic pancreas in the mediastinum is a very rare. While the exact genesis of ectopic pancreas is not known, its position in the anterior mediastinum

Table 1. Clinical features of mediastinal ectopic pancreas described in the literature.

Year	Symptom	Sex	Age	Tumor size (CM)	Gross feature	Position	Incision	Following-up (M)
1957 (2)	Dyspnea	Female	16	12	Cystic	/	Median sternotomy	/
1977 (3)	No symptom	Female	57	7x6.5	Cystic	Left	Left thoracotomy	/
1996 (4)	Heart murmur	Female	16	12	Cystic	/	Median sternotomy	/
2001 (5)	Cough, chest pain	Female	45	11x8	Cystic	Left	Median sternotomy	24
2004 (6)	Dyspnea	Male	44	10x8x7.5	Cystic	Left	Median sternotomy	/
2005 (7)	No symptom	Male	39	10x8	Cystic	Left	Median sternotomy	96
2006 (8)	Fever, neck swelling, dyspnea	Male	40	8x6x6	Cystic	Left	Median sternotomy	/
2007 (9)	Chest pain	Female	17	12.5x12.0x4.5	Cystic	Right	Right thoracotomy	/
2007 (9)	Chest pain, short of breath	Female	24	10x8x4	Cystic-solid	Right	Right thoracotomy	/
2009 (10)	No symptom	Female	32	16x13x8	Cystic-solid	Bilateral	Median sternotomy	3
2010 (11)	Chest pain, cough, dyspnea	Male	51	7.5x7.0x5.0	Cystic	Right	Median sternotomy	/
2010 (11)	Left shoulder pain, dyspnea, hemoptysis	Male	42	/	Cystic-solid	Left	Left thoracotomy	28
2012 (12)	Chest pain	Female	66	7.8x4.6x2.7 and 10.8x9.1x3.7 (two pieces)	Cystic-solid	No mention	Median sternotomy	Death (15)
2012 (13)	Hemoptysis, chest pain	Male	32	/	Cystic	Right	Median sternotomy	24
2012 (14)	Chest pain, mild cough, blood-tinged sputum	Female	31	7x3x4	Cystic-solid	Left	Left thoracoscopy	/
2013 (15)	Cervical mass	Female	22	2.4x3.8	Cystic	Cervical	Cervical approach	48
2014 (16)	Dyspnea, stridor	Male	18	16x12x9	Cystic	Left	Median sternotomy	/
2014 (17)	Chest pain, cough, fever	Male	15	7x4.5	Cystic-solid	Right	Median sternotomy	96
2014 (17)	Throat discomfort, neck swelling	Female	16	6	Cystic-solid	Median	Median sternotomy	6
2015 (18)	Cough, chest pain, shortness of breath	Male	17	7.6x7x5.8	Solid	Left	Median sternotomy	/

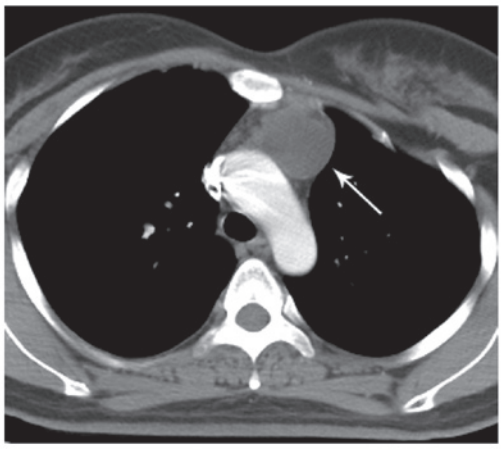

Figure 1. Chest CT. The preoperative scan shows a cystic-solid mass measuring 3.6x3.9 cm and situated mainly on the left side of the anterior mediastinum; a follow-up scan at 3 years after surgery shows no tumor recurrence. CT, computed tomography.

argues against a gastroenteric origin because lesions of gastroenteric origin normally occur in the posterior mediastinum.

We performed a PubMed search of the English language literature for papers relating to anterior mediastinal ectopic pancreas and found 17 such papers reporting a total of 20 cases (2-18), all of which were confirmed at surgery (Table I). Most of these reports were from Asian countries, with others from Europe and North America. Women slightly outnumbered men, and the average age was 32 years. The common presenting symptoms were chest pain, dyspnea, and cough. Other symptoms were neck swelling, heart murmur, and hemoptysis. There were very few patients who were entirely asymptomatic. The nature of the presenting symptoms was closely correlated with the size and location of the tumor, and the largest reported tumor diameter was 16 cm (10,16). In most of the cases, the chest CT showed a cystic or cystic-solid mass, with only one pure solid mass reported (18). There was often moderate enhancement of the cyst wall on the post-contrast

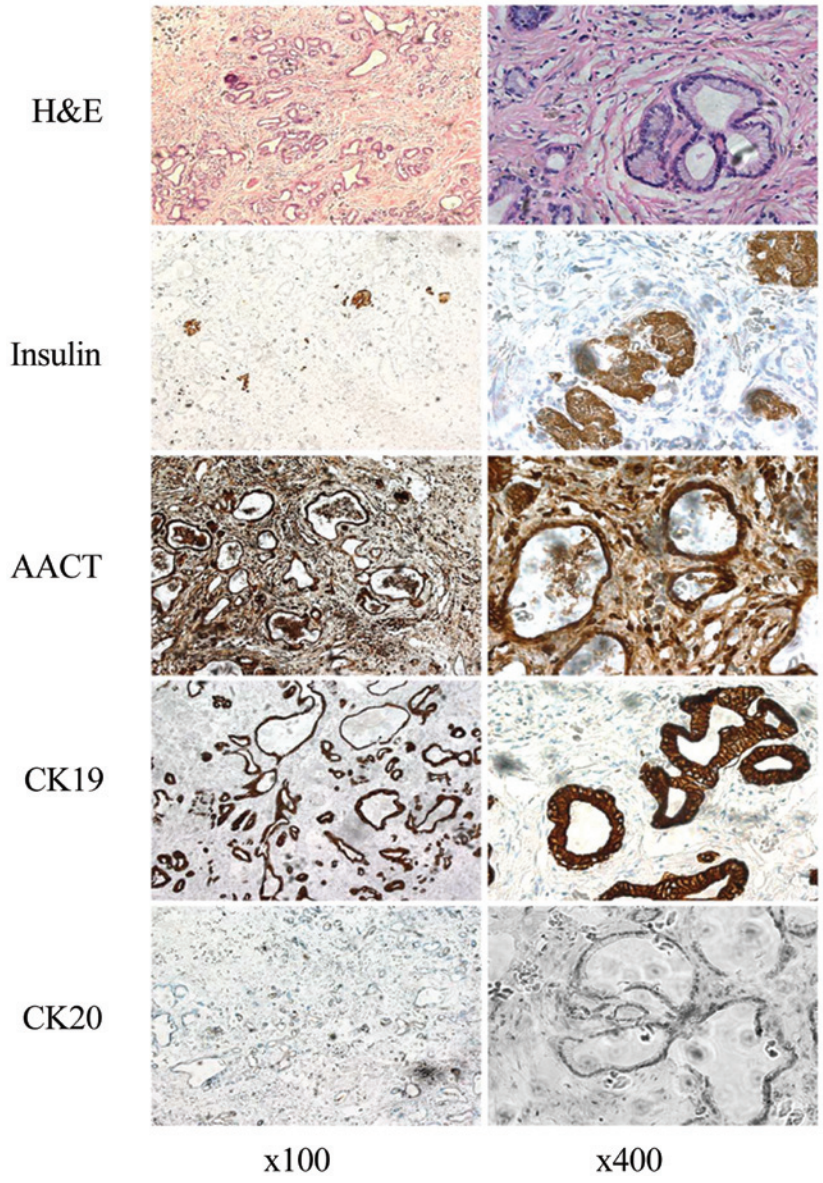

Figure 2. Hematoxylin/eosin staining and immunohistochemistry. Microscopy shows a normal acinar structure in the solid portion of the mass and pancreatic acini and islet cells in the wall of the cystic portion. Immunostaining is positive for islet cell insulin and cystic alpha-ACT, CK19, and CK20.

CT, and some cases showed enhancement of the solid part of the lesion. Arriving at a precise diagnosis preoperatively was difficult, and provisional diagnoses included thymoma, lymphoma, mediastinal thyroid, and abscess, depending on the CT appearance (19). Although several of these patients underwent preoperative biopsy, almost all of the cases were definitively diagnosed only after complete surgical resection, which is feasible without a definitive diagnosis when the mass is considered resectable (20).

Anterior mediastinal ectopic pancreas is most often benign, and surgery is curative in most cases. However, there is a report of an aggressive adenocarcinoma arising from ectopic pancreas in the anterior mediastinum, which metastasized to the anterior sternum 6 months after resection of the primary tumor (12). The disease progressed rapidly, and the patient died within 15 months after the initial presentation, despite tumor debulking and postoperative adjuvant treatment. Because of the risk of malignant transformation of ectopic pancreas in the anterior mediastinum, surgical treatment should be planned without delay, and awareness of the possibility of the rare occurrence of adenocarcinoma arising from ectopic pancreas in the anterior mediastinum can help prevent misdiagnosis.

The majority of patients with ectopic pancreas in the anterior mediastinum have had curative resection by median sternotomy, or by thoracotomy where the main tumor has penetrated into one side of the chest. Besides our patient, thoracoscopy has been used for complete resection in one other case, in South Korea (14). The surgical approach to resection of anterior mediastinal masses has traditionally been based on studies of thymectomy, and advances in minimally invasive surgery have challenged the dictum of median sternotomy as the approach of choice. When compared with sternotomy, both video-assisted thoracoscopic surgery (VATS) and robot-assisted thoracoscopic surgery (RATS) have been associated with better estimation of the relationship of the tumor with adjacent structures, decreased intraoperative blood loss, decreased length of hospital stay, and reduced postoperative pain (21). The present study selected chose thoracoscopy in

our case after careful consideration of the patient's age and the size and location of the tumor.

In summary, the possibility of ectopic pancreas should be considered when a patient presents with a cystic or cystic-solid mass in the anterior mediastinum. Ectopic pancreas in the anterior mediastinum is rare and infrequently reported, and it is difficult to diagnose without biopsy. Our experience confirms that prompt surgical resection is beneficial for affected patients. As medical technology progresses, new surgical techniques may bring further advances, and this too should be fully considered.

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Availability of data and materials

The datasets used and/or analyzed during the present study are available from the corresponding author on reasonable request.

Authors' contributions

HZhao and JC wrote this manuscript and analyzed all of the data. XL, ZZ, JL and HZhang provided medical care for the patients and collected the data. GC performed the operation. GC and JC revised the article. All authors read and approved the final manuscript.

Ethics approval and consent to participate

The present study was approved by the Ethical Review Committee of Tianjin Medical University General Hospital. Written informed consent was obtained from the patient for the publication of this case report and the accompanying images. A copy of the consent form is available for review by the Editor-in-Chief of this journal.

Patient consent for publication

Written informed consent was obtained from the patient for the publication of this case report and the accompanying images.

Competing interests

The authors declare that they have no competing interests.

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