Ectopic mediastinal pancreas

Ektopik mediastinal pankreas

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Ectopic pancreatic tissue rarely occurs within the mediastinum. A 23-year-old male patient was admitted with dyspnea. Computed tomography revealed a 6x8 cm cystic mass within the anterior mediastinum in the thymic localization. The patient underwent thoracoscopy and surgery with the preliminary diagnosis of a thymic cyst. Histopathological examination was reported as ectopic pancreatic tissue. No recurrence or metastasis was observed during a nine-month follow-up period. We present this case which was reported as mediastinal ectopic pancreatic tissue following surgical resection due to its rare occurrence.

Key words: Anterior mediastinum; ectopic pancreas; surgery.

Ectopic pancreatic tissue is a congenital anomaly found in approximately 2% of autopsies, and most (70-90%) are located within the gastrointestinal tract. Development of this tissue within the mediastinum is very rare. In fact, to our knowledge, only 12 known cases exist in the literature, and these were usually asymptomatic. Diagnosis of ectopic pancreatic tissue is diagnosed via a pathological examination performed after surgery. In this report, we describe a rare case of mediastinal ectopic pancreatic tissue.

CASE REPORT

A 23-year-old man was admitted to the thoracic surgery department of our hospital with a complaint of dyspnea. His physical examination, complete blood count, routine biochemistry tests, and pulmonary function tests were normal, but echocardiography identified mild pulmonary stenosis (maximum gradient: 25 mmHg). Computed tomography (CT) of the chest showed a cystic mass of 6x8 cm in the anterior mediastinum with thymic localization (Figure 1a). Right video-assisted thoracoscopic surgery (VATS) was performed to resect the mass and verify the preoperative possible diagnosis of a thymic cyst. We preferred this procedure because the heart and aorta are localized within the left hemithorax, so the operation space on the right side is greater. The border between the left innominate vein (LIV) and the mass could not be identified (Figure 1b). During the dissection, LIV was ruptured, so we performed a partial median sternotomy. After controlling the bleeding with a primary repair, 50 ml of yellowish-green fluid was aspirated, and the cystic lesion was totally resected (Figure 1c). Following the primary repair on the ruptured portion of the LIV, there was obstruction of its lumen, and sufficient blood flow could not be reestablished. Thus, the obstructed venous segment was excised, and a 6 mm
A macroscopic examination revealed a grayish brown mass that measured as 6x6x2.3 cm. The cyst’s inner surface was yellowish, and the wall thickness was 1.2 cm at its widest point. Microscopically, there was thymic tissue adjacent to the cyst, but there was no evidence of teratoma. We saw only adipose tissue in and around the thymic tissue. The cyst wall was lined with cuboidal-columnar epithelium, and at certain focal areas, a pseudostratified, ciliary, columnar epithelium was present (Figure 2a). In addition to the fibrotic tissue on the cyst wall, other structures typical of pancreatic tissue, including ducts, acini, and Langerhans islet cells, were also observed (Figure 2b). The acinar cells of the exocrine pancreas were composed of polygonal cells with apparent eosinophilic apical cytoplasmic processes, and these were accompanied by ductal structures. The endocrine component had a paler cytoplasm, central nuclei, and islets with changing cell numbers (Figure 2c). Immunocytochemically, the pancreatic islet cells had a positive reaction with synaptophysin and chromogranin A (Figure 2d), and the pathological results revealed ectopic pancreatic tissue. The patient did not develop any complications after surgery. He is currently in the ninth postoperative follow-up month, and no recurrence has been seen.

DISCUSSION

Ectopic pancreatic tissue is a congenital anomaly which is defined by no anatomical or vascular connection to normal pancreatic tissue or isolated pancreatic tissue at an abnormal localization. This anomaly is found in approximately 2% of all autopsies. Pancreatic tissue within the thoracic cavity and mediastinum is distinctly uncommon and is generally seen as a component of gastrointestinal duplication cysts, intralobar pulmonary sequestrations, or mature teratomas. However, we found no chondroid tissue or any other tissue which brought teratoma to mind in our case. In addition, there was no tissue from any other organ in this specimen. Pancreatic tissue is generally localized to anterior or middle mediastinum and is frequently in the form of a cystic structure. The size of the lesion can range from 4-20 cm. In our case, the ectopic pancreatic tissue was cystic.

Figure 1. (a) Thoracic computed tomography showing a cystic lesion in the anterior mediastinum. (b) Right video-assisted thoracoscopic surgery revealed that the lesion and the left innominate vein were adjacent to each other. (c) The mass after resection. (d) The mediastinum after grafting with polytetrafluoroethylene.
in nature with anterior mediastinal localization. After 50 ml of fluid was aspirated from the mass, it measured 6×6×2.3 cm.

Mediastinal ectopic pancreatic tissue is usually seen in young patients like ours and occurs at nearly the same proportion in males and females. These lesions do not usually have specific symptoms, but chest pain, headaches, hypoglycemia, and hyperglycemia have been observed. Our patient was admitted to the hospital complaining of dyspnea.

The histogenesis behind the development of ectopic pancreatic tissue within the mediastinum is not clear, but two hypotheses have been put forward. Some contend that the pluripotent epithelial cells of the ventral primary foregut go through an abnormal differentiation. The resulting pancreatic tissue then localizes itself within the mediastinum. Others believe that the cells migrating from the pancreatic bud localize to different areas. As with our patient, the study by Al-Salam and Al Ashari demonstrated the presence of a pseudostratified epithelium within the cystic component, signifying that no migration of cells from the pancreas to that localization had taken place. Instead, the pancreatic tissue was differentiated there. Chen et al. underlined the fact that cysts develop due to increased secretion because of the presence of different epithelial structures, such as the pseudostratified ciliary epithelium in our case. However, the development of cysts cannot be explained by this mechanism alone.

Especially in symptomatic cases, the normal treatment option is surgery, and a diagnosis is established via a histopathological examination after the surgical resection of the mass. In our patient, the operation was started with VATS to resect the mass, and its cystic nature was then revealed. It was tightly joined to the LIV, and the development of the LIV rupture during the dissection led to the median sternotomy and total resection of the mass.

Histopathologically, the cyst is normally lined with cuboidal-columnar epithelium and/or focal pseudostratified ciliary columnar epithelium. There is fibrosis on the cyst wall, and the exocrine component consists of acini made up of polygonal cells with eosinophilic apical cytoplasm. In addition, the endocrine component includes islets with varying

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Figure 2. (a) The cyst wall lined with pseudostratified ciliary epithelium along with pancreatic ducts and acini (H-E x 400). (b) Pancreatic tissue on the cyst wall adjacent to the thymic tissue (H-E x 40). (c) Polygonal cells of the exocrine acini together with cells of the endocrine component represented as islet structures (H-E x 400). (d) The pancreatic islet cells had reacted positively with chromogranin A (x 100).
numbers of cells, and ductal structures can also be found. Perez et al.,
Cagirici et al., and Tamura et al., reported that the distribution of acini and
islets caused the structure to be irregular. The histological structure of our case was similar in that
the structure of the acini and the islet was irregular. Immunohistochemically, markers like synaptophysin
and chromogranin A are useful when identifying the endocrine component.

Tamura et al. followed-up a patient with mediastinal ectopic pancreatic tissue for eight years after surgery and
with no recurrences or metastases that are often found with other cases involving ectopic pancreatic tissue.
Our patient also did not develop any complications after surgery, and no recurrence or metastasis was detected
during a follow-up period of nine months.

In conclusion, in young patients who do not have any specific symptoms and who have large mediastinal
cystic masses in addition to other differential diagnoses, mediastinal ectopic pancreatic tissue should certainly be kept in mind.

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