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Congenital Thymic Cyst in the Retro Cava Position: A Case Report

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Introduction: Congenital thymic cyst (CTC) is a rare mediastinal mass and the retrocaval position of this cyst has not been reported in the medical literature so far.
Case Presentation: The present study reports on a 66-year-old female with coronary artery disease (CAD) who was scheduled for coronary artery bypass grafting (CABG) at our hospital. During mediastinal exploration and pericardiotomy for CABG, a large CTC was incidentally found in the retrocaval position, as a watery cyst from its anatomic location and content. Pathological examination revealed that it was a CTC. No cases of retrocaval position of CTC have been reported. Thus, here we report on this case and review the previously reported cases.
Conclusions: To our knowledge, this is the first case of CTC that was identified incidentally in the retrocaval position and caused intermittent obstruction of vena cava.

Keywords: Coronary Artery Disease; Cronary Artery Bypass; Cyst

1. Introduction

Thymic cysts were classified by Constantacos as congenital or acquired cystic lesions of thymus (1). Abnormal descending of thymus from the pharyngeal pouch to mediastinum in the fetal life, leads to aberrant location of the thymus in the neck and mediastinum. The most common site of these cysts is either the anterior mediastinum or the neck yet sometimes for unknown reasons, they can be found in the fetal thymus tissue sliding to ectopic site of neck or out of midline mediastinum line to plural cavity or in subclavian vein or as in our patient, in the retrocaval position. It is found that thymic cysts include 1% of mediastinal cystic lesions, and occur predominantly from the 2nd to 5th decades of life (2). Considering the embryology of the thymus, the presence of a congenital thymic cyst outside the midline of mediastinum is very rare and thymic cysts located solely at the retrocaval site have not been described. Here we report on a rare large congenital thymic cyst exclusively located at the retrocaval position.

2. Case Presentation

A 66-year-old female was admitted to our hospital in Kermanshah (Iran) in July 2013, with chest pain and congenital thymic cyst (CTC) and characteristic history of flushing and periorbital edema. Her symptom was not positional and was not exacerbated by breathing. Her medical history was remarkable and previous myocardial infarction events were reported. Routine blood tests were normal, and no tumor markers were found. Chest radiography was normal and routine echocardiography for heart revealed no abnormally. Her medical and family histories, clinical status and blood and urine tests, were unremarkable. Postoperative retrospective analysis of signs and symptoms revealed facial edema caused by intermittent obstruction of superior vena cava (SVC) by a cyst. Exploration of mediastinum was performed through a median sternotomy, resulting in a postoperative diagnosis of a mediastinal cyst. A soft and fluctuant cyst was present in the middle mediastinum in residual thymus tissue around the SVC and retro subclavian vein, and was easily dissected from these organs. No pedicle was attached to the inferior aspect of the cyst. The cyst had a thin wall and contained watery fluid. Judging from its anatomic location and contents, the cyst was diagnosed as a congenital cyst. The cyst was resected after dissection of the inferior surface in the left lateral aspect of SVC and retro subclavian vein in mediastinum (Figure 1). As of the content of the cyst, the number of cells was few and the cell count revealed no lymphocytes. The fluid did not contain of triglyceride. Microscopically, the cyst wall consisted of thin fibrous connective tissue with monolayer cells on its luminal surface. Thus, the cyst was histologically compatible with the congenital thymic cyst (Figures 2 - 4). The patient's postoperative course was complicated by a transudative left pleural effusion. It emerged on the first postoperative day, decreased gradually on the fifth postoperative day and dissolved spontaneously one week after the operation. The patient was discharged on the 12th postoperative day and had no symptoms eight months later. The surgical specimen was a solitary cyst

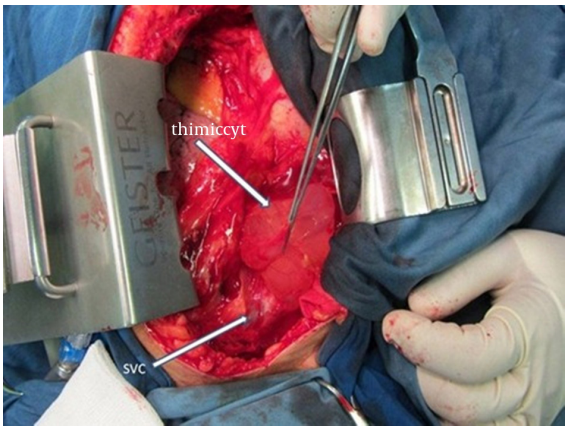

Figure 1. Intraoperative View of the Cyst at the Retrocaval Position

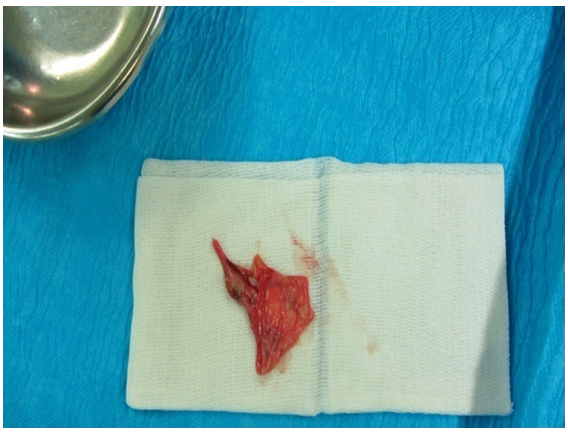

Figure 2. Gross Pathology of the Opened Cyst

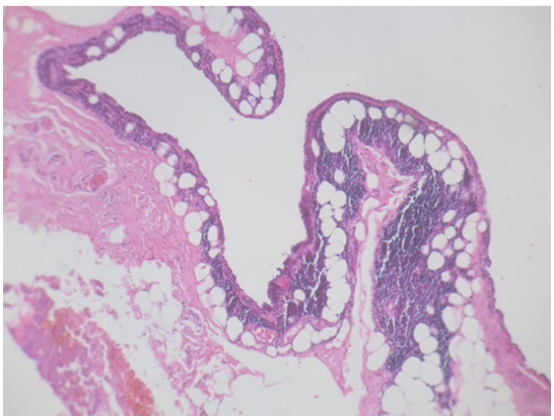

Figure 3. The Cyst Irregularly Lined by Keratinized Squamous Epithelium (magnification × 100)

with a fibrous thin wall, measuring 10 × 6 cm at its maximum width and length. Microscopically, the cystic walls were irregularly lined by keratinized squamous epithelium and focally flattened endothelium (Figures 3 and 4) with varying thickness, and contained scattered nests of thymic tissue including small lymphocytes and epithelial cells. These findings were consistent with a thymic cyst, and the macroscopic features indicated a congenital type.

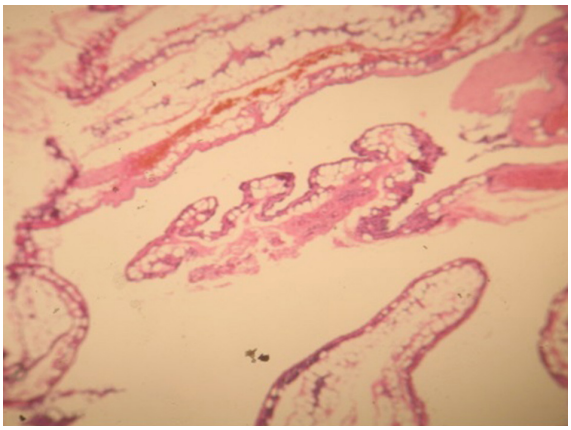

Figure 4. The Cystic Wall Containing Islands of Thymic Tissue (original magnification × 100)

3. Discussion

In our case, an incidentally found mediastina cyst turned out to be a congenital thymic cyst. We successfully treated the patient by removing the cyst through surgery.

congenital thymic cysts are very rare and mostly asymptomatic mediastinal lesions. Ectopic displacement of such cysts to retro-cava position is exceptional and has not been described so far. In cases with CTC at the retrocaval position and intermittent signs and symptoms of SVC syndrome, surgical resection for establishing diagnosis and for definite treatment is recommended. During the first trimester of fetal intra uterine life, the thymus is divided from the third pharyngeal pouch and, it migrates through its course from neck, as a separate lobe of thymus, to the upper part of the mediastinum where these lobes fuse. During this descent, the thymic lobes complete their medial fusion at the end of the second trimester of intrauterine life, occupying their destination site in the upper part of the mediastinum, yet unusual migration of thymic lobes to neck, within the chest to pleura, or around the great vein are possible. Ectopic CTC, around the superior vena cava is an extremely rare phenomenon (3, 4). These CTC are usually unilocular with thin walls, and represent derivatives of embryologic thymic tissue. The acquired type is often multi-locular as a result of an inflammatory or neoplastic process. However, CTC often contains a clear serous fluid, yet rarely; it may be exudative, turbid or sanguineous. Other complications of CTC include, bleeding, rupture, hemothorax, mediastinal hemorrhage or local symptoms. Congenital Thymic Cysts may present with pressure effect on neighboring organs such as pressure on recurrent nerve (stridor, hoarseness and vocal cord paralysis), esophagus (dysphagia), pericardium (chest pain) and lungs (with huge cysts, leading to dyspnea) (5). Lachanas, reported on a case of intermittent obstruction of the left brachiocephalic vein by CTC. Compression of the right ventricle and transudative pleural effusion has also been described (6). Although CTC usually grows

very slowly, yet there are some cases of unilocular thymic cysts that enlarge rapidly by intracystic hemorrhage in children with aplastic anemia (7). The differential diagnosis of CTC includes, a neoplasm with cystic degeneration, post radiation cystic degeneration of the thymus, a complex cystic lesion, bronchopulmonary foregut duplication cyst, cystic lymphangioma, mesothelial cyst, pancreatic pseudocyst herniating through the diaphragm, thoracic aneurysm, and thoracic duct cyst (8). Sahhar et al. (9) described a case of ten-day-old boy that had stridor and respiratory distress by a subglottic CTC. After the cyst was removed endoscopically, the patient's symptoms resolved. Pathology identified the cyst as an ectopic thymic tissue. This case was noteworthy because the authors believed that was the first reported instance of ectopic thymic tissue in the subglottis. Tandon et al. (10) presented a case of five-year-old child with a painless swelling on his neck. Imaging revealed a multi-locular cystic lesion lying in close proximity to carotid vessels. Based on the imaging study, the possibility of an ectopic thymic cyst was considered and confirmed by histopathology exam. Ectopic cervical cyst is an infrequent cause of neck cyst in a child, which is rarely diagnosed preoperatively. Bruno et al. (11) described a 37-year-old man with a huge pleural cyst, presented with symptoms of right heart compression. The mass on the right side of the chest seemed to be initially in connection with the mediastinum. Computed tomography failed to define its relationship with the pericardium, and echocardiography excluded any involvement of the mediastinal structures. The final diagnosis was a congenital thymic cyst exclusively located in the pleural cavity. Congenital Thymic Cysts are uncommon and asymptomatic yet some giant thymic cysts cause cardiac compression and related signs and symptoms. Scharifker (12) described a mediastinal mass in a patient with the sole complaint of persistent cough and found it to be a true thymic hyperplasia associated with a unilocular CTC. The bi-lobed obtained thymus was macroscopically and microscopically normal except for the size (12 × 12 × 3.5 cm) and weight (97 gram). It showed normal thymic tissue was segregated in lobules separated by adipose septa with good corticomedullary separation. Congenital thymic cysts are located along the anatomical course of the third pharyngeal pouch and may also be related to neoplasms. Prasad et al. (13) reported a case, who presented a soft, fluctuating mass on the left side of his neck. Surgical excision revealed an ectopic thymic cyst. Congenital cervical thymic cysts are a very rare cause of neck tumor in children and they can exceptionally induce a laryngeal compression or may be attached to the pericardium and cause respiratory distress, in a newborn (14).

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Authors' Contributions

Freidoun Sabzi developed the original idea and the protocol, abstracted and analyzed the data, wrote the manuscript, and was the guarantor. Babak Eizadi, Nasrin Javid and Reza Faraji contributed to the development of the protocol, abstracted data and prepared the manuscript.

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