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Retrocardiac mediastinal foregut duplication cyst

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ABSTRACT

Foregut duplications occur at various locations throughout the upper gastrointestinal tract, including the thoracic cavity. Thoracic foregut duplications are typically intimately associated with the esophagus and therefore are located in either the posterior mediastinum or in pleural cavity. Here we report a case of foregut duplication of the middle mediastinum, intimately associated with the pericardium and great vessels that contained gastric mucosa, ciliated respiratory epithelium, bronchial-type epithelium, pancreatic tissue and hepatocytes. The literature of mediastinal foregut duplications is also reviewed.

Foregut duplications are rare developmental anomalies that are thought to originate from failure of developing vacuoles to coalesce into the developing esophagus [1,2]. Thus aberrant structures are created in the lateral chest or mediastinum that can be intimately related to the esophageal wall or completely separate. Occasionally these structures jointly occur with vertebral defects [3]. Traditional criteria for alimentary tract duplication are the presence of a well-developed smooth muscle wall, internal gastrointestinal mucosa and close anatomic relation to a part of the digestive tract [1]. Most cases of intra-thoracic foregut duplication occur in the posterior mediastinum. Of highest concern for anterior and middle mediastinal masses are lymphoma or teratoma, amongst other malignant possibilities. Here, we report a case of foregut duplication cyst in the retrocardiac space between the left atrium and right pulmonary artery.

1. Case report

The patient is a 3-year old boy who initially presented with an asymptomatic heart murmur. At his two-and-a-half-year old well-child check, he had met all developmental milestones, and had a normal physical exam along with updated immunizations. He presented 6 weeks later with a fever and multiple sick contacts at day care, at which time a vibratory systolic heart murmur was appreciated. He was seen six months later for his 3-year old well-child visit at which time persistent 2/6 systolic murmur was again noted.

He was referred to cardiology and an echocardiogram was performed. He was found to have a structurally normal heart with normal great vessel and pulmonary arterial and venous anatomy with mild physiologic pulmonary insufficiency and normal coronary origins and anatomy. There was no pericardial effusion and no masses, vegetations or thrombi seen. There was, however, a suspicious echogenic shadow posterior to the aorta either in or behind the left atrium and an echogenic area could not be ruled out in the left atrioventricular groove as well.

At the time of cardiology evaluation, he remained asymptomatic, specifically without cyanosis, tachypnea, tachycardia/palpitations, color changes, respiratory distress, chest pain, feeding difficulties, shortness of breath, dizziness, syncope or exertional symptoms. Cardiac exam demonstrated a grade 1/6 medium frequency systolic ejection murmur of the Still's variant heard best at the left lower sternal border with no radiation and no diastolic murmur. The apical impulse was normal and no thrill was present. Peripheral pulses were normal. Electrocardiogram showed sinus rhythm with two premature ventricular contractions. 24 h Holter monitoring showed no premature atrial contractions or supraventricular ectopic beats and 150 premature

Abbreviations: MRI, magnetic resonance imaging; TEF, tracheoesophageal fistula.

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ventricular contractions/h, a load of 2.5%. There were no runs of ventricular tachycardia. A repeat echocardiogram further characterized the shadow as a 2.5 cm complex multicystic mass superior to the left atrium and posterior to the ascending aorta. It had no Doppler flow and was not compressing the atria nor the surrounding vascular structures. It was thought to be in the pericardial space.

Magnetic resonance imaging (MRI) of the heart and chest was performed (Fig. 1a). A retrocardiac mass was identified on this study, measuring 4.5 × 3.8 × 3 cm and bordering the superior vena cava and ascending aorta anterolaterally, the right atrium anteriorly, the left atrium and left main and proximal left circumflex arteries inferiorly and the right pulmonary artery superiorly. There was intermediate intensity on T1-weighted images and very high signal intensity on T2-weighted images with multiple internal septa. The great vessel and cardiac anatomy was normal as was cardiac function. There were no atrial or ventricular septal defects. There was no component of the mass along the posterior aspect of the sternum anterior to the heart. The thymus appeared normal for age.

At multidisciplinary conference, cystic lymphangioma or teratoma was thought to be the likely diagnosis and that surgical resection would be required. He underwent surgical resection via a median sternotomy. The tumor was identified protruding between the ascending aorta and superior vena cava (Fig. 1b). It was not attached to the superior vena cava but was adherent to the aortic adeventitia and right pulmonary artery. In the course of separating the tumor from the pulmonary artery, the arterial lumen was entered. After resection, a small pericardial patch was used to repair the arteriotomy (Fig. 1c, arrowhead). There were no connections between the tumor and the posterior pericardial lining of the oblique sinus or the carina. There was no visible associated lymphadenopathy and the tumor was removed intact. Cardiopulmonary bypass was not required for safe removal of the mass. Postoperatively, he recovered uneventfully and was discharged from the hospital on the fourth day. He had no subsequent issues related to the resection.

Histopathologic examination confirmed the multicystic nature of the mass (Fig. 2). Multiple types of epithelial linings in the cyst wall were identified including single layer cuboidal, ciliated respiratory epithelium, bronchial-type mucigenic epithelium, small nodules of cartilage and gastric epithelium. There were well-developed layers of smooth muscle associated with the gastric epithelium and these contained hyperplastic neurenteric-type neural tissue. Furthermore, within the fibrous tissue there was a small remnant of pancreatic tissue with duct-like structures and a minute nodule of hepatocytes. The final pathologic diagnosis was that of a foregut duplication cyst.

2. Discussion

In an asymptomatic or minimally symptomatic mediastinal lesion, such as the one reported here, the primary concern is establishing a diagnosis. Malignant potential is the major consideration, and operative intervention to confirm or exclude a cancer diagnosis is the recommended clinical course. Over one-half of mediastinal lesions will be either lymphoma or a neurogenic tumor. Another common finding is germ cell tumor, of which teratoma is the most common [4,5]. Once these have been excluded, foregut duplication is the next most common diagnosis. The risk of malignant transformation in these lesions is very low. Thus far only four cases of cancer arising from foregut duplications have been reported in the literature [6–9]. The ages of these patients range from 37 to 61 years and in two cases the existence of the mediastinal mass was known for years prior to diagnosis of cancer, while in the other two patients rapid enlargement produced symptoms that prompted further evaluation. These cases suggest not only that cancer is extraordinarily rare within intra-thoracic foregut duplications, but also that its development is very slow, as each of these patients presumably had lived with their duplications since birth. Taken together, timely excision at any point in childhood or early adulthood should suffice to alleviate the risk of developing malignancy.

The only sign our patient showed was a systolic heart murmur identified at thirty months of age. To our knowledge, this is the only report of mediastinal foregut duplication presenting in this way. The only other reported case of a cardiac-related presentation of duplication cyst is a 27-year-old male patient who presented with atrial fibrillation that was refractory to medication, and the mass was discovered on a cardiac MRI obtained prior to planned electrophysiologic ablation procedure [10]. The ablation procedure was canceled, the patient underwent uneventful resection of the mass; it was intimately associated with the right inferior pulmonary vein and also abutted the esophagus and thoracic spine and was tethered with superficial adhesions only. After resection, the patient's dysrhythmia resolved. In an extensive case series of intra-thoracic duplications and bronchogenic cysts from one particular institution, no patients were reported to have cardiac signs or symptoms [11]. Kassner and colleagues report the case of a young child with an associated pericardial defect abutting an esophageal duplication, though this child's symptoms were aerodigestive rather than cardiac [12]. They also cite several earlier case reports of associated pericardial defects, suggesting a shared developmental origin. Foregut duplications most often present in late infancy or at some point in childhood. They rarely present with symptoms in the neonatal period [3,13–15] or in association with esophageal atresia or tracheoesophageal fistula (TEF), which will be diagnosed early

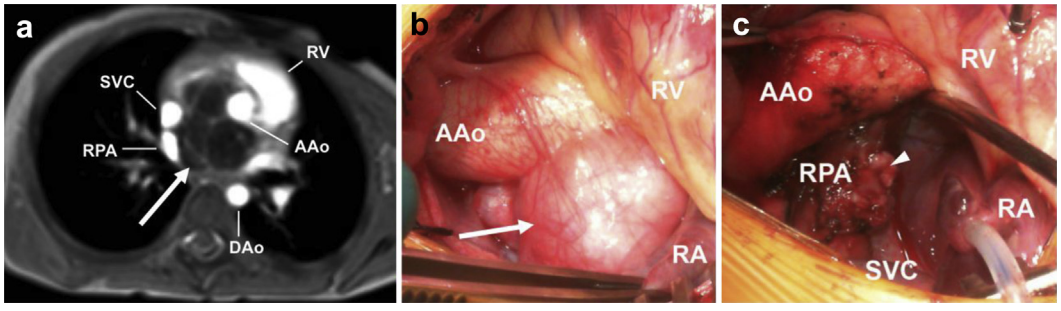

Fig. 1. Foregut duplication cyst in retrocardiac position within the middle mediastinum. (a) MRI image demonstrating close relationship of cystic mass (*arrow*) to superior vena cava, right pulmonary artery and ascending aorta. (b) Intraoperative photograph of exposed surface of the mass (*arrow*) adherent to the ascending aorta. (c) Intraoperative photograph after resection of the mass illustrating the space it occupied, with pericardial patch repair of the RPA (*arrowhead*), covered retraction suture on the RA and suction device retracting the RV. RPA right pulmonary artery, SVC superior vena cava, RV right ventricle, AAo ascending aorta, DAo descending aorta, RA right atrium.

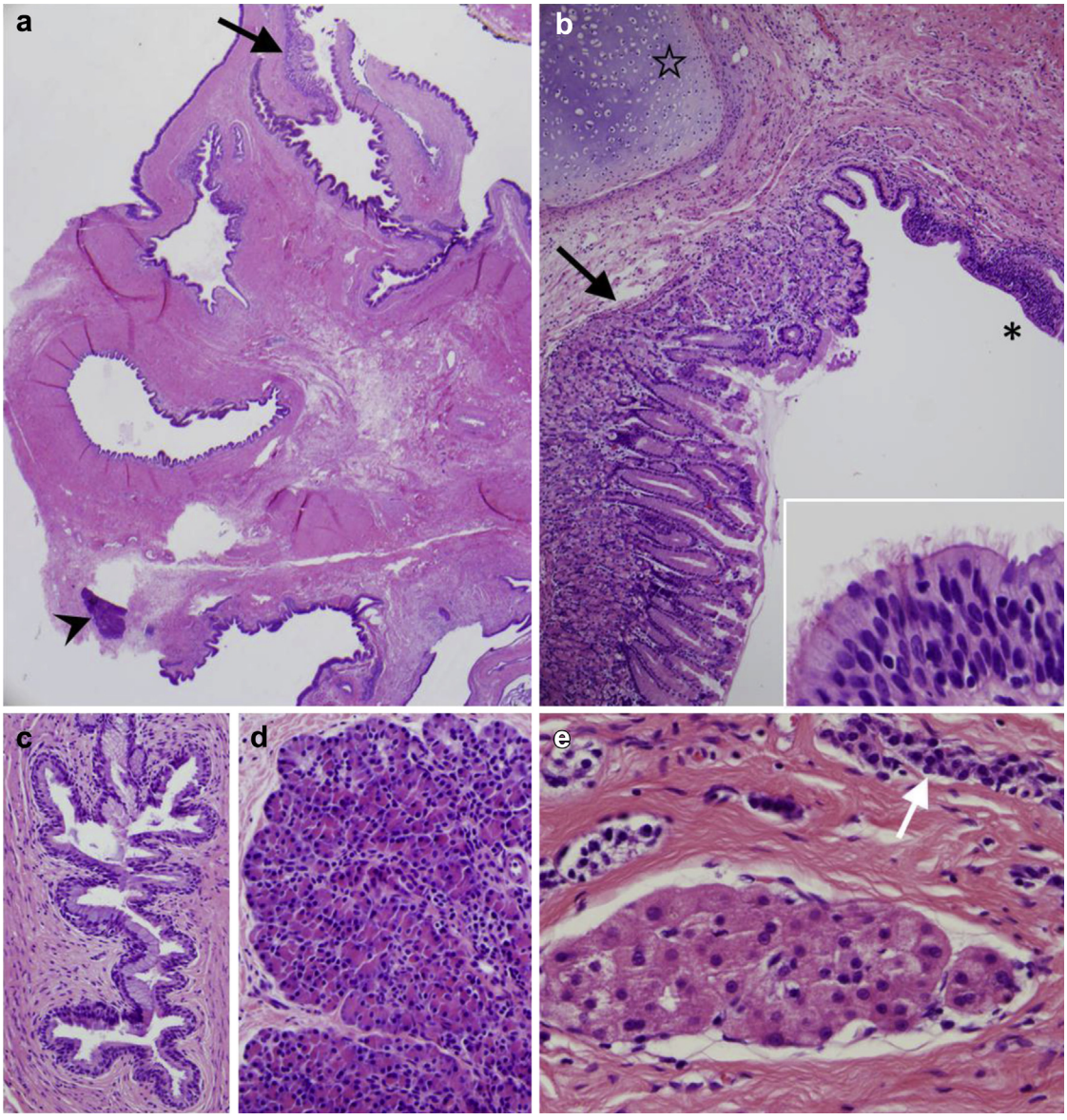

Fig. 2. Histopathologic examination of mediastinal cystic mass (a–e). The multicystic structure features varying layers of surrounding/circumferential smooth muscle, intervening fibrous tissue containing scattered islands of cartilage (*open star*), and displays a spectrum of foregut derived epithelium and tissues. These include gastric type mucosa (*black arrows*), respiratory pseudostratified ciliated columnar epithelium (*asterisks, inset*), (c) bronchial-type mucogenic epithelium, (d) pancreatic tissue (*arrowhead*), and (e) minute nodules of hepatocytes with adjacent biliary-type ductular epithelium (*white arrow*) (a 20×; b 100×, *inset* 600×; c 200×; d 400×; e 600×).

[16–19]. Even more rarely is the diagnosis made based on prenatal imaging [20]. Our patient's mumur was identified at thirty months of age, which fits with the typical timing of presentation despite the highly unusual type of presentation encountered.

As with all developmental anomalies, the search for any associated problems must be considered. For mediastinal foregut duplications, the two major concerns are any coexisting defects in the thoracic vertebrae, which could suggest the presence of a neurenteric cyst with true connection of the cyst to the vertebral structures [3] or non-contiguous vertebral anomalies [15,21]. Evaluation of the thoracic spine by MRI revealed no vertebral anomalies in our patient. In one review, the authors indicated that about one-quarter of patients with a mediastinal foregut duplication will also have a second, intra-abdominal duplication (5 of 21 patients) [1]. However, that report retrospectively identified patients for inclusion in the series by diagnosis of duplication, and therefore does not address the potential for undiagnosed intra-abdominal duplications that remain asymptomatic, so the true rate may be higher. Our patient did not undergo specific abdominal work-up. There is no consensus for what abdominal work-up should be considered in these patients after the diagnosis of intra-thoracic foregut duplication. As mentioned previously, there are also several reports of the co-existence of duplication in the setting of esophageal atresia with or without associated TEF [16–19]. This rare, distinct subset of patients with duplication are highly likely to be identified primarily on the basis of the atresia or TEF, rather than primarily on the basis of the associated duplication.

Potential complications that can arise within alimentary tract duplication include peptic ulceration or bleeding from acid-secreting gastric mucosa or infection of a fluid-filled cyst. Gastric mucosa has been reported to be present in 16.7% of intra-thoracic foregut duplications or bronchogenic cysts [11]. In one case, ulceration from gastric mucosa led to fistulization of a posterior mediastinal duplication to the right lower lobe of the lung and ultimately

to bleeding through the respiratory tract [13]. In another case, bleeding into a second duplication several years after resection of a first, intra-thoracic duplication required transfusion [3]; resection of the duplication resolved the ulcer or bleeding diathesis. Gastric mucosa was present within our patient's cyst, however no stigmata of bleeding or ulceration were identified. Much less common is the occurrence of pancreatic tissue within intra-thoracic foregut duplication; very few cases are reported [1,14–16]. Aberrant intra-thoracic pancreatic tissue is more often found within ventral budding errors such as congenital pulmonary airway malformations or sequestrations, and is thought to arise from failure of the ectopic pancreatic tissue to descend caudally in the developing embryo [11]. Our patient's cyst is also one of the few to contain pancreatic tissue.

3. Conclusion

The case we report here follows many of the typical patterns associated with proximal foregut duplication. It is highly unique for its unusual location in the middle mediastinum and close association with the pericardium and great vessels. Foregut duplications carry a low risk for malignancy, but a definitive diagnosis of a mediastinal mass cannot be determined without histopathologic exam. Therefore a timely excision of any such lesion after discovery should be considered. Our patient will require careful follow up in the case of subsequent unrelated symptoms given the possibility of a secondary developmental anomaly.

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