A Rare Presentation of Anterior Mediastinal Teratoma Mimicking Valvular Heart Disease with A Systolic Murmur

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Cite this article as: *Tan Chor Lip H, Jih Huei T, Abdul Wahid A, Jerome Vendargon S. A rare presentation of anterior mediastinal teratoma mimicking valvular heart disease with a systolic murmur. Eurasian J Med 2018; 50: 134-6.*

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Received: December 5, 2017
Accepted: January 21, 2018

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DOI 10.5152/eurasianjmed.2018.17388

ABSTRACT

Extrinsic pulmonary artery stenosis caused by anterior mediastinum teratoma presenting with an ejection systolic murmur is a rare phenomenon. Till date, 15 cases have been reported (inclusive of this case) in the English literatures. Herein we report a 20 year old female with extrinsic pulmonary artery stenosis because of compression by an anterior mediastinal teratoma with a loud ejection systolic murmur. The case report aims to highlight the awareness of such rare presentation of anterior mediastinal teratomas that may mimic congenital valvular heart diseases among clinicians.

Keywords: Teratoma, benign, systolic murmurs, pulmonary artery

Introduction

Pulmonary stenosis caused by extrinsic compression of an anterior mediastinal teratoma is a rare phenomenon [1]. The first case was described by Maier et al. [2] in 1948. In this case, a 4-year-old girl presented with a harsh systolic murmur because of extrinsic compression of the pulmonary artery by a teratoma in the anterior mediastinum [2]. To our knowledge, till date, only 15 cases (including our case) of teratoma within the anterior mediastinum causing extrinsic pulmonary stenosis have been reported in the English literatures [1-7]. Herein, we report on a 20-year-old female with an anterior mediastinal teratoma mimicking a valvular heart disease with a loud ejection systolic murmur. This case report aims to increase the awareness of such rare presentations of anterior mediastinal teratomas that may mimic congenital valvular heart diseases.

Case Presentation

A 20-year-old female previously healthy, presented with symptoms of reduced effort tolerance and chest discomfort for four months without orthopnea or paroxysmal nocturnal dyspnea. The electrocardiogram and blood investigations were within normal limits. Physical examination by the primary care health team revealed that the ejection systolic murmur was loudest over the pulmonary area. There were no other constitutional signs of heart failure. Based on the findings of a heart murmur, she was referred to the cardiology department with a primary suspicion of congenital valvular heart disease. However, transthoracic echocardiography revealed extrinsic compression of the main pulmonary artery causing turbulent blood flow. The peak pressure gradient was 35 mmHg which led to the presence of an ejection systolic murmur. No other structural abnormalities in the heart were detected on the transthoracic echocardiography. A chest roenterogram disclosed a round, smooth

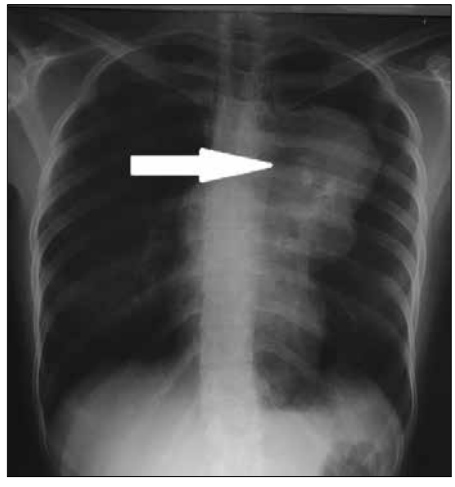

Figure 1. Chest roenterogram showing a suspicious mass over the left upper thorax (green arrow) with clear lung fields

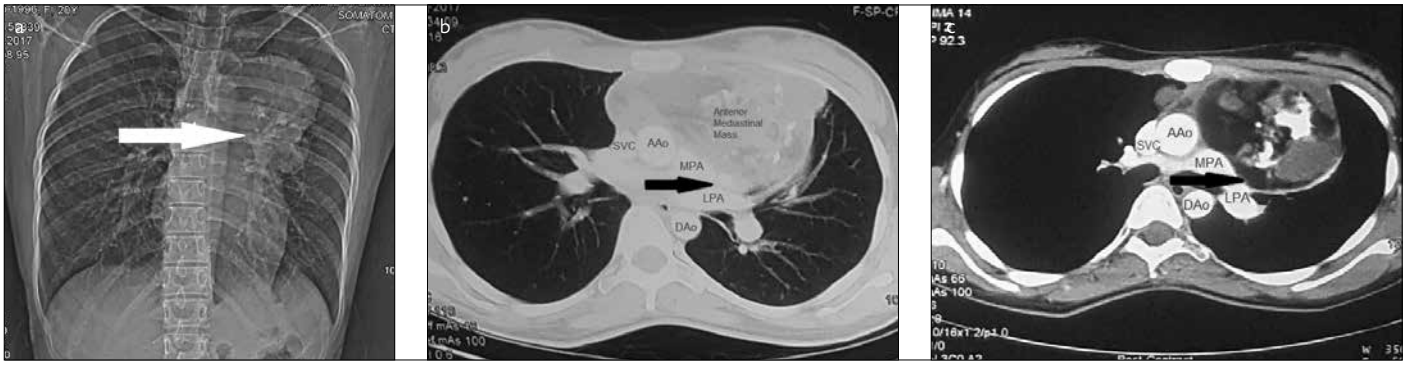

Figure 2. a-c. Coronal view of thorax on CT scan showing anterior mediastinal mass in the left upper thorax (green arrow) (a), Lung window view showing anterior mediastinal mass compressing over the pulmonary artery (green arrow) (b), Mediastinal window view showing anterior mediastinal mass compressing the pumonary artery (green arrow) (c)
SVC: superior vena cava; AAo: ascending aorta; MPA: main pulmonary artery; LPA: left pulmonary artery; DAo - descending aorta

mass in the left upper border of the mediastinum (Figure 1). Computed tomography (CT) of the thorax shows a large anterior mediastinal mass measuring 5.6cm × 10.7cm × 9.5cm causing compression to the pulmonary trunk and left atrium (Figure 2 a-c). There were no enlarged mediastinal lymph nodes and the lung fields were clear. Through a primary median sternotomy, a well encapsulated cystic tumor measuring 15 cm in diameter was found within the anterior mediastinum causing compression to the main pulmonary artery (Figure 3a). Excision was meticulously performed to dissect the tumor from the pericardium. Sectioning of the specimen revealed a yellowish solid multicystic mass containing hair, sebaceous material, cartilage, and bone (Figure 3b). Microscopic examination revealed that the cystic structures were lined by keratinizing stratified squamous epithelium (skin) and ciliated bronchial epithelium (Figure 4b, c). The solid area contained fat, smooth muscles, hair follicles, sebaceous glands, eccrine glands, exocrine pancreas, cartilage, bone, and sero-mucinous salivary glands arranged in haphazard pattern (Figure 4a). The histology of the resected specimen had no immature cells and this led to a diagnosis of a mature teratoma of the mediastinum. Post-operative recovery was uneventful and the murmur disappeared following excision of the mass. Patient was discharged on post-operative day six and was well on subsequent clinical follow-up. Informed consent was taken from the patient prior publication and the consent form is available with the authors and publisher.

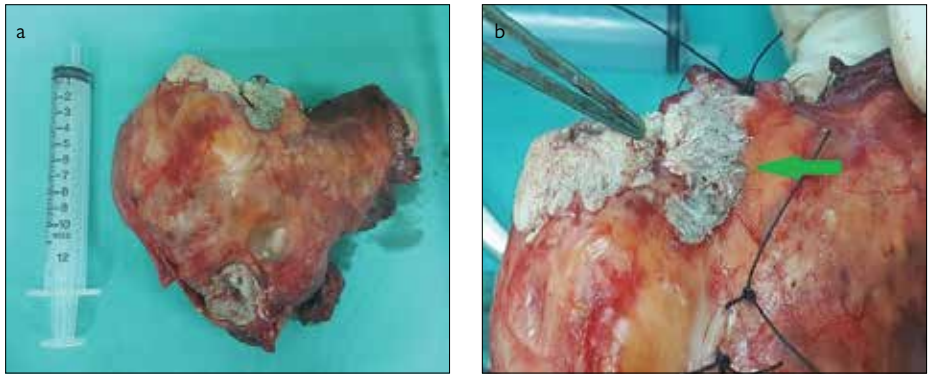

Figure 3. a, b. Gross specimen after excision of a cystic mass with presence of sebum, and hair (a), Close up view showing sebum and hair over the surfaces of the tumor (green arrow) (b)

Discussion

Pulmonary stenosis is defined as the constriction of the right ventricular outflow tract below, above, or at the pulmonary annulus which leads to an increase in right ventricular pressure [3]. The causes may be broadly divided into intrinsic (congenital) or extrinsic compression. Several known causes of extrinsic pulmonary stenosis are Hodgkin's disease, lymphoma, teratoma, lung carcinoma, pericardial sarcoma, thymoma, and chondrosarcoma of the sternum [4]. Including our case, only 15 cases of pulmonary stenosis because of extrinsic compression by a teratoma in the anterior mediastinum have been reported. All of the 15 cases reported in the literatures demonstrated a loud systolic murmur on presentation. Extrinsic compression of the pulmonary artery invariably leads to acute symptoms which differs from pulmonary stenosis due to congenital causes which may have a more prolong and chronic symptoms prior seeking medical attention. Other associated symptoms are exertional dyspnea, pleuritic chest pain with cough, and palpitations [1-7]. In the evaluation of such cases, chest roenterogram may be of great benefit. The presence of a mass on a chest roenterogram may aid the clinician in diagnosis and making necessary referrals to a cardiology center with cardiothoracic surgery backup. Transthoracic echocardiography is equally important to ascertain extrinsic pulmonary stenosis and identify any other related structural heart abnormalities [5]. CT of the thorax gives essential information for pre-operative planning, possibility of benign or malignant tumor, and any suspicious lymph node or invasion in the surrounding organs. Teratoma is a germ cell tumor composed of somatic tissue derived from two or three of the germ cell layers. Teratoma can be further classified as mature teratoma (adult-type tissue) and immature teratoma [8]. Microscopic examination of mature teratoma may demonstrate squamous epithelium, hair follicles, sebaceous sweat glands, smooth and striated muscle, respiratory epithelium, thymus, thyroid, intestines, bone, or cartilage tissue [1]. Patients with anterior mediastinal teratomas of the mature type, generally carries a good prognosis after complete resection of tumour. The majority of the systolic murmurs which occur due to the extrinsic compression disappears after complete excision of the teratoma [3]. Anterior mediastinal teratoma masquerading as valvular heart disease is of particular interest to clinicians, cardiologist, and surgeons alike because of its nature to mimic congenital valvular heart disease. This leads to frequent difficulty and errors in interpretation of physical signs and chest roenterograms of such cases [5]. Delay due to misinterpretation may lead to death in some cases of malignant disease as reported by Fry et al. [7]. Therefore it is of utmost

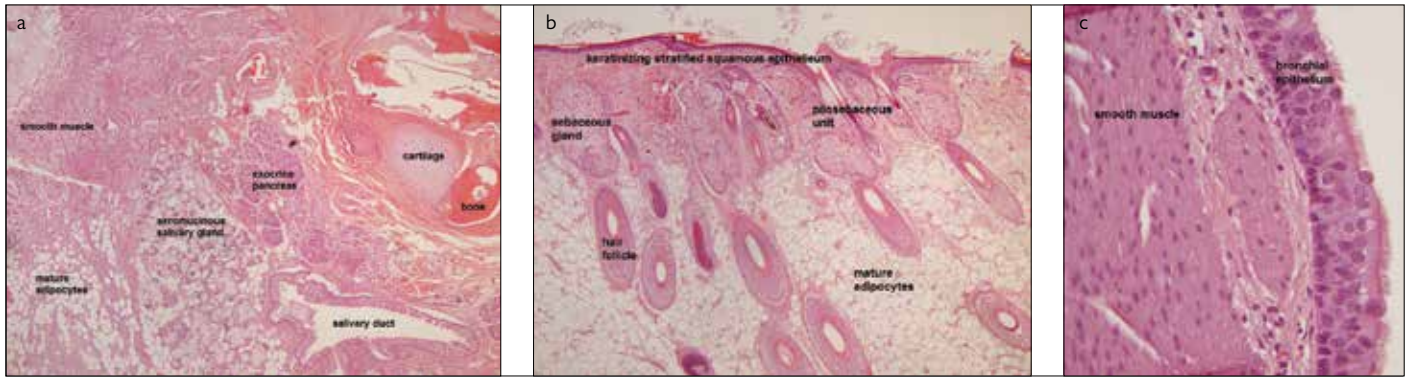

Figure 4. a-c. Microscopic examination showing mature components of smooth muscle, mature adipocytes, seromucinous salivary gland, salivary duct, exocrine pancreas, cartilage, and bone (a), Keratinizing stratified squamous epithelium, hair follicle, and sebacaeous gland (b), Bronchial epithelium (c)

importance to highlight such cases to increase the awareness among medical practitioners. In conclusion, extrinsic pulmonary artery stenosis due to anterior mediastinal teratoma is a rare phenomenon with only 15 cases reported till date. This case report highlights the awareness of anterior mediastinal teratoma which may mimic valvular heart disease with the presence of a systolic murmur. A simple chest roentero-gram revealing a suspicious mediastinal mass may aid in the diagnosis of such cases.

Informed Consent: Written informed consent was obtained from the patient who participated in this study.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept - H.T.C.L., T.J.H.; Design - H.T.C.L., S.J.V.; Supervision - H.T.C.L., S.J.V.; Resources - H.T.C.L.., A.A.W.; Materials - H.T.C.L., S.J.V.; Data Collection and/or Processing - H.T.C.L., A.A.W.; Analysis and/or Interpretation - H.T.C.L., A.A.W., T.J.H.; Literature Search - H.T.C.L, T.J.H; Writing Manuscript - H.T.C.L, S.J.V; Critical Review - H.T.C.L., S.J.V.; Other - A.A.W, H.T.C.L.

Conflict of Interest: Authors have no conflict of interest to declare.

Financial Disclosure: The authors declared that this study has received no financial support.

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