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EC CARDIOLOGY
Case Report

Mediastinal Germ Cell Tumor Compressing the Main Pulmonary Artery and Part of the Left Pulmonary Artery with Consequent Right Sided Heart Failure and Dilatation

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Abstract

Pulmonary stenosis could be either congenital, which is not un common or acquired, which is extremely rare. Compression of the pulmonary artery by mediastinal tumors is the most common cause of the acquired pulmonary artery stenosis. Reviews and case reports suggest that tumors of the mediastinum (teratomatas and Hodgkin's disease) are the most frequent culprits leading to compression of the main pulmonary artery. This is a case report of a 21-year-old male patient who presented with retrosternal chest pain lasting for 2 mo. Compression of the main and left pulmonary artery by an anterior mediastinal seminomatous germ cell tumors (GCTs) was diagnosed by transthoracic echocardiography and CT scan.

Keywords: *Germ Cell Tumor; Acquired Pulmonary Artery Stenosis; Echocardiography; Computed Tomography Scan*

Introduction

Acquired pulmonary stenosis is uncommon, particularly so when it is due to extrinsic compression by a mediastinal tumor. extra gonadal metastasis of germ cell tumors is not un common, it is usually localized in the mediastinum. Approximately 1 and 5% of all germ cell tumors occur in the mediastinum, of which about 85% are benign. They typically occur in twenties to forties, with equal sex distribution. These are usually slow growing tumors and are often detected incidentally on chest radiographs.

Case report

21 years Old male presented to ER with history of shortness of breath, dull aching retrosternal chest pain, swelling of both lower limbs and abdominal distention 2 months ago; He was diagnosed 2 years ago in other hospital as a case of Germ cell tumor of the left testis on chemotherapy. Patient was tachypnea T: 36.5, BP: 110/75.

PSO2: 85% RA RR: 24 P: 78 B/M. On examination: Congested neck vein up to ear lobe.

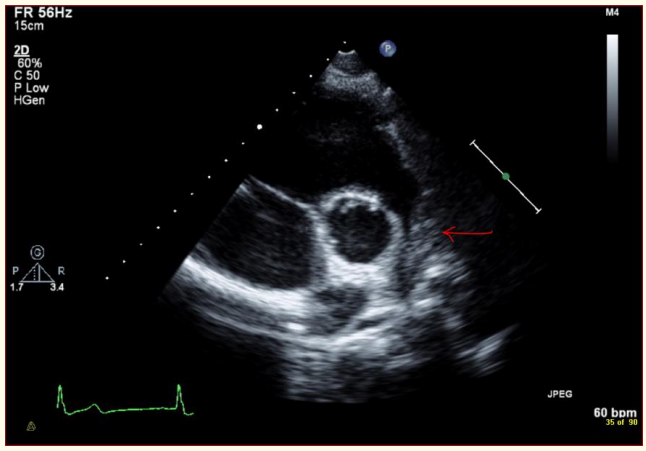
Chest: bilateral equal air entry no added sound.

CVS: Left parasternal heave, systolic thrill over Lt parasternal area, Normal S1, Accentuated S2, Systolic murmur increased with inspiration, a harsh grade 3/6 ejection systolic murmur was heard in the left second intercostal space along the sternal border.

Abdomen: moderately distended with moderate ascites and painful hepatomegaly bilateral Lower limbs pitting edema, no peripheral stigmata of infective endocarditis.

ECG: sinus tachycardia, right atrial enlargement, tall R in V1 with right ventricular strain pattern.

CXR: Showed wide mediastinum otherwise normal lung parenchyma. We examined the patient, requested for echocardiography which showed (Figure 1- 5): LV: Normal LV function and dimensions, normal mitral valve and normal aortic valve. RV: Severely dilated right atrium and right ventricle, severely RV dysfunction, sever tricuspid regurge, significant supravalvular pulmonary stenosis [1] (peak gradient = 65 mmHg), sever pulmonary hypertension, no Intracardiac shunt and no pericardial effusion. In context of patient complain with history of germ cell tumor and echo data, we highly recommended for Contrast-enhanced computed tomography of the chest (CT chest); which showed (Figure 6): Contrast-enhanced computed tomography image showing the mass in the mediastinum compressing the main pulmonary artery and left pulmonary artery [2] with clear lung fields. Ventilation perfusion scan was done; showed well perfused right lung while left lung non-perfused and almost absent (Figure 7).



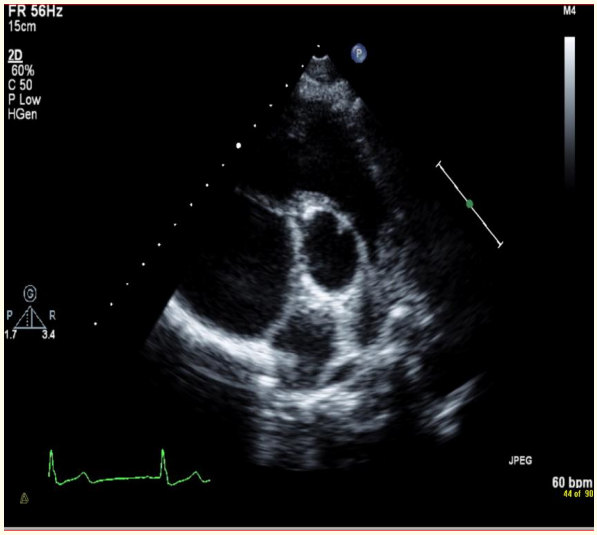


Figure 1 and 2: *Parasternal short axis view shows: supravalvular pulmonary stenosis due to mass compressing main pulmonary artery with part of left pulmonary artery (arrow) (figure 1 and 2).*

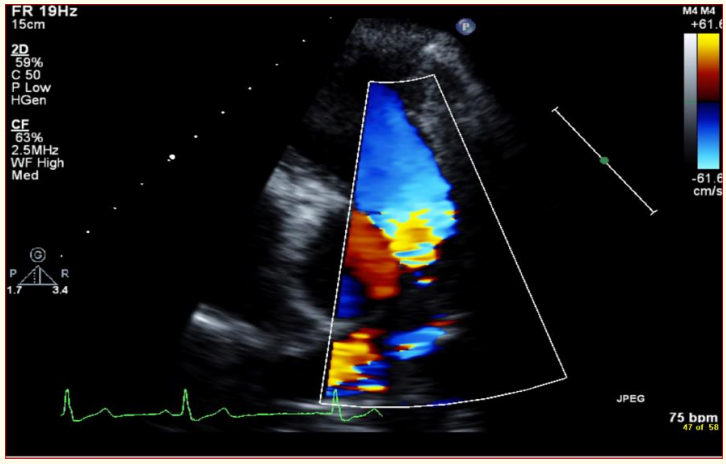


Figure 3: *Parasternal short axis color flow shows flow acceleration and signal aliasing at area of compression supravalvular with no jet at level of pulmonary valve.*

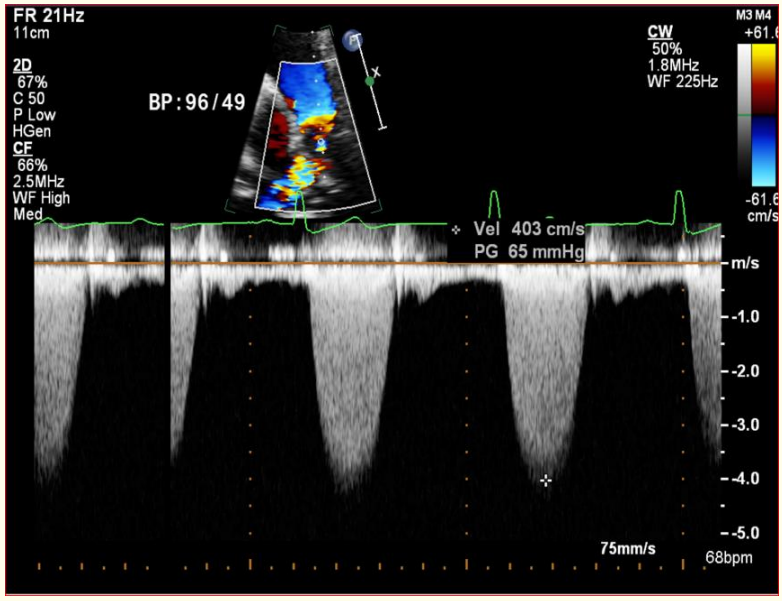


Figure 4: *A 2 dimension (2D) guided continuous wave Doppler image from PSAX at level of right ventricular outflow tract (RVOT) shows a maximum velocity of 4.03 m/s, and a pressure gradient of 65 mmHg represented sever supravalvular pulmonic stenosis.*

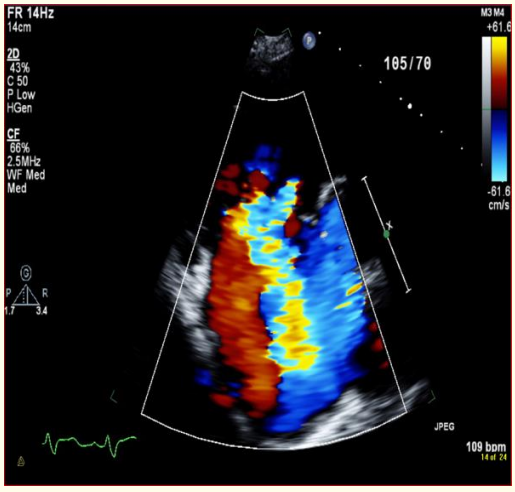


Figure 5: *A 2d color doppler over parasternal long axis inflow shows sever right atrial, right ventricle dilatation with sever tricuspid regurge.*

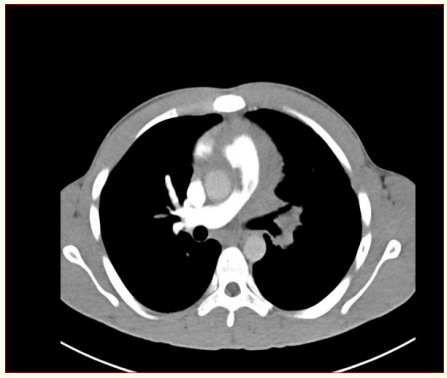


Figure 6: *Contrast-enhanced computed tomography image showing the mass in the mediastinum compressing the main pulmonary artery and left pulmonary artery with clear lung field.*

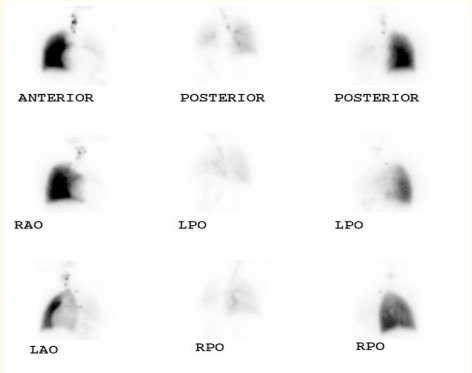


Figure 7: *Ventilation perfusion scan was done; showed well perfused right lung and non-perfused left lung which almost absent.*

Eventually, we diagnosed the patient as a case of mediastinal germ cell tumor compressing the main pulmonary artery and part of the left pulmonary artery with consequent right sided heart failure and dilatation; patient was promptly admitted to our center, kept on medical treatment then, emergently sent to operation room [3]. A palliative conduit from right ventricular out flow tract to left pulmonary artery was done. CT chest was done post-surgery confirmed patency of the conduit (Figure 8), patient followed by cardiology team showed well recovery [4-7].

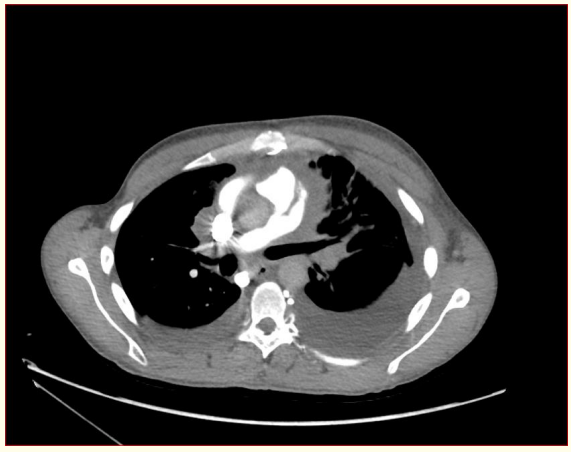


Figure 8: *CT chest was done post-surgery confirmed patency of the conduit.*

Discussion

Congenital pulmonary stenosis is not an uncommon lesion, but acquired stenosis of the pulmonary outflow tract due to external compression by mediastinal masses which compress the pulmonary artery or right ventricular outflow tract has also been reported. Compression could be due to many causes (e.g.: aortic aneurysm, Hodgkin's disease, Carcinoid, germ cell tumors (as in our case) [8] ……etc).

There are many treatment options ranged from chemotherapy [9], radiotherapy to surgical resection, the earliest the surgery done, the least deleterious effects loaded on the right sided of the heart [10].

Conclusion

There are two types of pulmonary stenosis ; The first is due to diseases affect the pulmonary valve itself, the others are due to external compression of the pulmonary artery by surrounding structures as mediastinal tumours, Hodgkin's disease, and aneurysm of the ascending aorta. As early pick up of these treatable acquired compression of the pulmonary valve is paramount, the new diagnostic modalities have been revolutionized over decades, made diagnosis of such rare diseases much more feasible.

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