

# An Unusual Cause of Main Pulmonary Artery Compression

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## Abstract

Primary mediastinal large B-cell lymphoma rarely causes clinically significant pulmonary artery compression. Acquired or secondary pulmonary stenosis is an uncommon condition and is usually caused by extrinsic compression from mediastinal masses, aneurysms or enlarged lymph nodes. Compression of the main pulmonary artery by an anterior mediastinal tumour is particularly rare because these masses generally enlarge laterally rather than anteroposteriorly. We report a 24-year-old man presenting with dyspnoea and constitutional symptoms. Imaging demonstrated a large anterior mediastinal mass with pleural effusion, while echocardiography showed compression of the main pulmonary artery with turbulent flow and a peak gradient of 40 mmHg. This case highlights an unusual cause of secondary pulmonary stenosis and the value of echocardiography in evaluating mediastinal masses.

## Keywords

Mediastinal Tumour, Pulmonary Artery Compression, Large B Cell Lymphoma

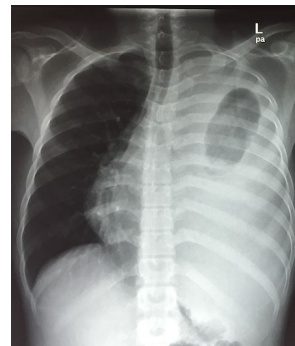
## 1. Introduction

Primary mediastinal large B-cell lymphoma (PMBCL) is a rare and distinct subtype of non-Hodgkin lymphoma that arises from thymic medullary B-cells and typically presents as a bulky anterior mediastinal mass in young adults. Due to its rapid growth and central mediastinal location, it may cause compressive symptoms involving adjacent thoracic structures such as the lungs, pleura, superior vena cava, heart, or pulmonary arteries. We describe a rare case of main pulmonary artery compression caused by a large thymic mediastinal tumour in a young patient, with histopathological confirmation of primary mediastinal large B-cell lymphoma. A

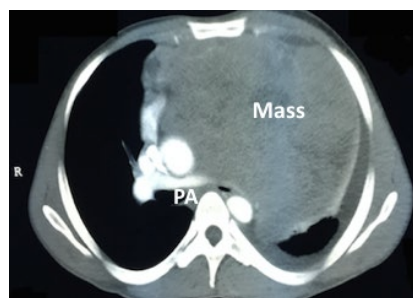
brief overview of mediastinal tumours and secondary causes of pulmonary stenosis will also be discussed, highlighting the importance of multimodality imaging and early recognition of this unusual but clinically significant condition.

## 2. Case Presentation

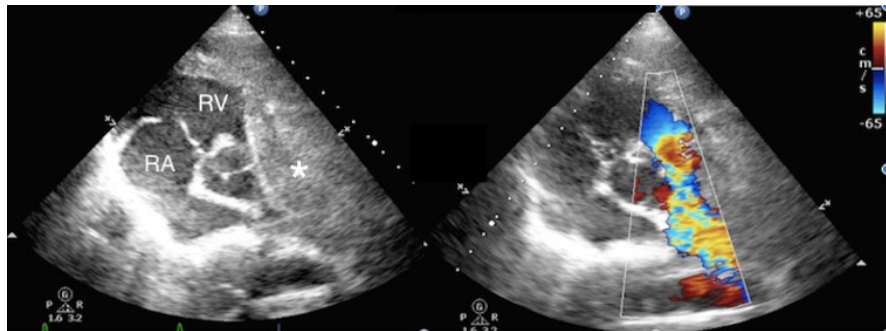
A 24-year old male presented with dyspnoea and constitutional symptoms of 3 months duration. His initial chest X-ray showed a large left-sided opacity with mediastinal shift (**Figure 1**) and was followed by a CT scan showing a large (13.2 cm × 11.4 cm × 10 cm) heterogeneous mass in the anterior mediastinum with an associated left-sided pleural effusion (**Figure 2**). An incidental 2/6 ejection systolic murmur was heard at the right base of the heart which increased on inspiration. There were no signs of right heart failure, and he was normotensive with mild respiratory distress saturating at 89% on room air. A cardiology consult was sought, and an echocardiograph was performed. The parasternal short axis view revealed a large mass compressing the main pulmonary trunk causing flow turbulence (**Figure 3**) and a continuous flow Doppler through the pulmonary artery showed a peak systolic pressure gradient of 40 mmHg over the supravalvular part of the pulmonary valve (**Figure 4**). There was normal right ventricular function with no right heart dilation. The pleural fluid was exudative but showed no malignant cells. The confirmatory diagnosis of Primary Mediastinal (thymic) Large B-cell lymphoma (PMBCL) was made on tru-cut biopsy. Despite the urgency to expedite chemotherapy in our patient with pulmonary artery compression, the patient de-  
ceased before chemotherapy could be instituted.



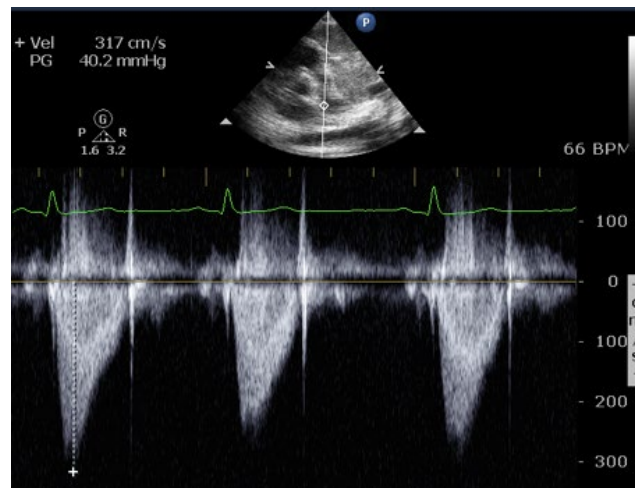
**Figure 1.** Left side opacity with mediastinal shift.



**Figure 2.** CT scan of large mass compressing the Pulmonary Artery (PA).



**Figure 3.** Short axis transthoracic echocardiographic picture illustrating the Right Atrium (RA), Right Ventricle (RV) and the compressive mass (\*) causing turbulence over the pulmonary valve.



**Figure 4.** Continuous-wave measurement shows a moderate peak gradient of 40 mmHg over the pulmonary valve.

### 3. Discussion

Acquired pulmonary stenosis is rare, and usually is a consequence of extrinsic compression. Mediastinal masses can compress vascular structures like the thoracic aorta and vena cava. It is, however, uncommon for it to compress the heart or pulmonary artery sufficiently to produce murmurs of hemodynamic significance [1] [2]. A postulated reason for this is that mediastinal tumours enlarge laterally, rather than antero-posteriorly. Mild to moderate right ventricular outflow obstruction is usually asymptomatic. It is only when there is severe obstruction that patients experience symptoms of exertional dyspnoea, fatigue and chest discomfort. This may, at times, be difficult to distinguish from initial symptoms caused by direct compression of lung parenchyma. With longstanding untreated severe obstruction, tricuspid regurgitation develops, which may lead to right ventricular failure.

Mediastinal masses comprise a heterogeneous group of lesions arising within the anterior, middle or posterior mediastinum. The differential diagnosis depends largely on the compartment involved. Anterior mediastinal masses are the most

common and classically include the “4 Ts”: thymic tumours, thyroid lesions, teratoma and other germ cell tumours, and “terrible” lymphoma.

Middle mediastinal masses are more often due to lymphadenopathy, bronchogenic cysts or vascular abnormalities, whereas posterior mediastinal masses commonly arise from neurogenic tumours. Large mediastinal masses may remain asymptomatic, but can produce symptoms through compression of adjacent structures, resulting in dyspnoea, cough, chest pain, dysphagia, superior vena cava obstruction or, more rarely, compression of the heart or pulmonary artery causing clinically relevant significant obstruction.

With regards to lymphoma, both cHL—classical Hodgkin lymphoma (typically nodular sclerosis classical HL) and Non-Hodgkin lymphoma, such as diffuse Large B-cell lymphoma (DLBCL), T-cell lymphoblastic lymphoma, Primary mediastinal (thymic) large B-cell lymphoma, “gray-zone” lymphoma—*i.e.* B cell lymphoma with features intermediate between DLBCL and HL, and composite lymphoma—*i.e.* both PMBCL and HL occurring in the same individual, are recognized causes of mediastinal tumours [3].

Primary mediastinal (thymic) large B-cell lymphoma (PMBCL) represents a rare subtype of NHL with distinct clinicopathological features [4]. PMBCL has a female predominance (approximate female to male ratio - 2:1), presents at a median age of 35 years and constitutes approximately 2% - 4% of the NHL [5]. PMBCL typically manifests with a mediastinal mass with invasion of the anterior-upper mediastinum. Adjacent structures in the thorax such as the lungs, pleura, pericardium, chest wall and heart may be involved. Systemic symptoms are relatively uncommon, affecting less than 20% of patients [6].

The optimal specific treatment for PMBCL is unclear. Based on retrospective studies, R-CHOP (rituximab, cyclophosphamide, doxorubicin, vincristine, prednisone) is generally considered as the standard of care. The role of consolidation radiotherapy is being challenged by such intensified chemotherapy regimens, such as DA-EPOCH-R (dose-adjusted etoposide, doxorubicin, cyclophosphamide, vincristine, prednisone and rituximab) [7]. Autologous hematopoietic stem cell transplantation is an option for patients with relapsed/refractory disease [8].

## 4. Conclusion

Our case presentation highlights a 24-year-old male with a massive mediastinal mass, constituting an unusual cause of main pulmonary artery compression. Systemic symptoms are uncommon in Primary mediastinal (thymic) large B-cell lymphoma; consequently, the absence of early clinical warning signs may contribute to delayed presentation and, as observed in our patient, a less favourable outcome.

## Ethics Statement

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

## Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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