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Mediastinal widening on chest radiography caused by combined aortic valve disease and primary large B-cell lymphoma

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Abstract

Poststenotic aortic root dilatation in patients with aortic valvular stenosis may result in mediastinal widening on chest radiograph. Main differential diagnosis of mediastinal widening is a tumour. In fact, besides atypical chest pain or dyspnoea at exertion secondary to compression of intrathoracic structures in the latter, symptoms may be absent. We report a case of combined aortic valve disease and additional primary large B-cell lymphoma.

MeSH: Aortic Valve, Lymphoma B-Cell, Echocardiography, Ventricular Outflow Obstruction

Introduction

Aortic valvular stenosis may cause dilatation of the ascending aorta with mediastinal widening on frontal chest radiograph. Still, there are some differential diagnoses to widening of the mediastinum, predominantly tumours.

Primary large B-cell lymphoma, a rare entity of Non-Hodgkin lymphoma, is a rapidly growing tumour that arises from the thymus with predominant mediastinal involvement, first described in the early 1980s.¹⁻² It occurs in adolescents and young adults, with women predominantly affected (2:1). Usually it does not affect other tissues. Patients may clinically present with atypical chest pain and cough, as well as dyspnoea on exertion or superior vena cava syndrome secondary to compression of intrathoracic structures.¹⁻⁴

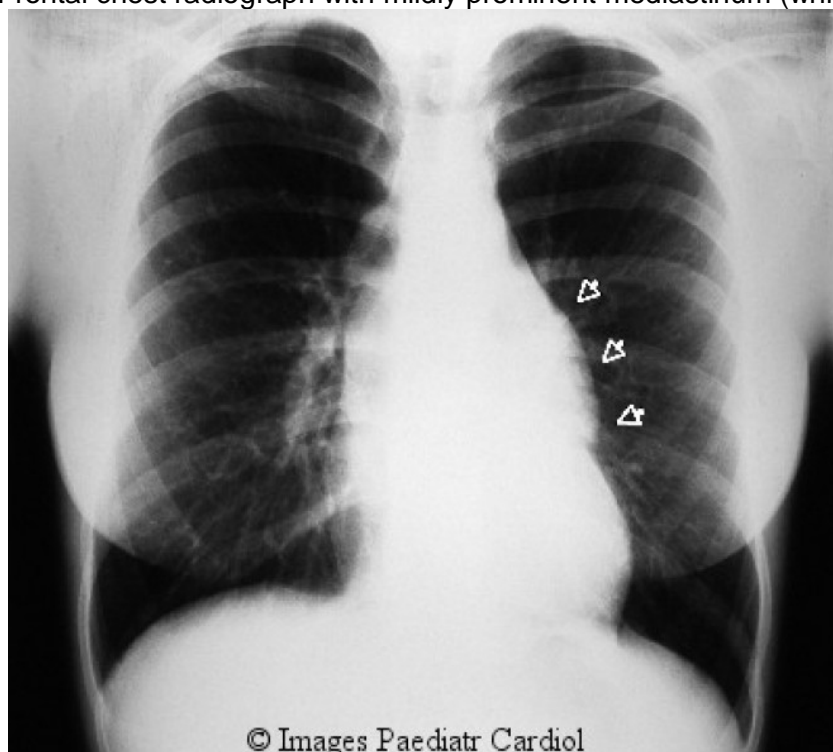
Case

History

A previously healthy 14-year-old girl presented with cardiac murmur. The frontal chest radiograph showed a mildly prominent mediastinum (Figure 1). Trans-thoracic echocardiography revealed combined aortic valve disease with thickening of the right coronary leaflet. Moderate aortic regurgitation resulted in slight left ventricular dilatation. Infective endocarditis was unlikely, as blood cultures were sterile and no suspicious findings on trans-oesophageal echocardiography were detected. Electrocardiogram was normal.

During the following months, she intermittently complained of weariness, dizziness, atypical chest pain, unproductive cough as well as dyspnoea on exertion. Echocardiography at intervals of 3-4 months revealed unchanged moderate aortic regurgitation. Based on an increasing left ventricular dilatation as well as systolic dysfunction despite the use of angiotensin-converting enzyme inhibitors, the decision for surgical aortic valve reconstruction was made.

Figure 1 Frontal chest radiograph with mildly prominent mediastinum (white arrows).



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Investigations and Course

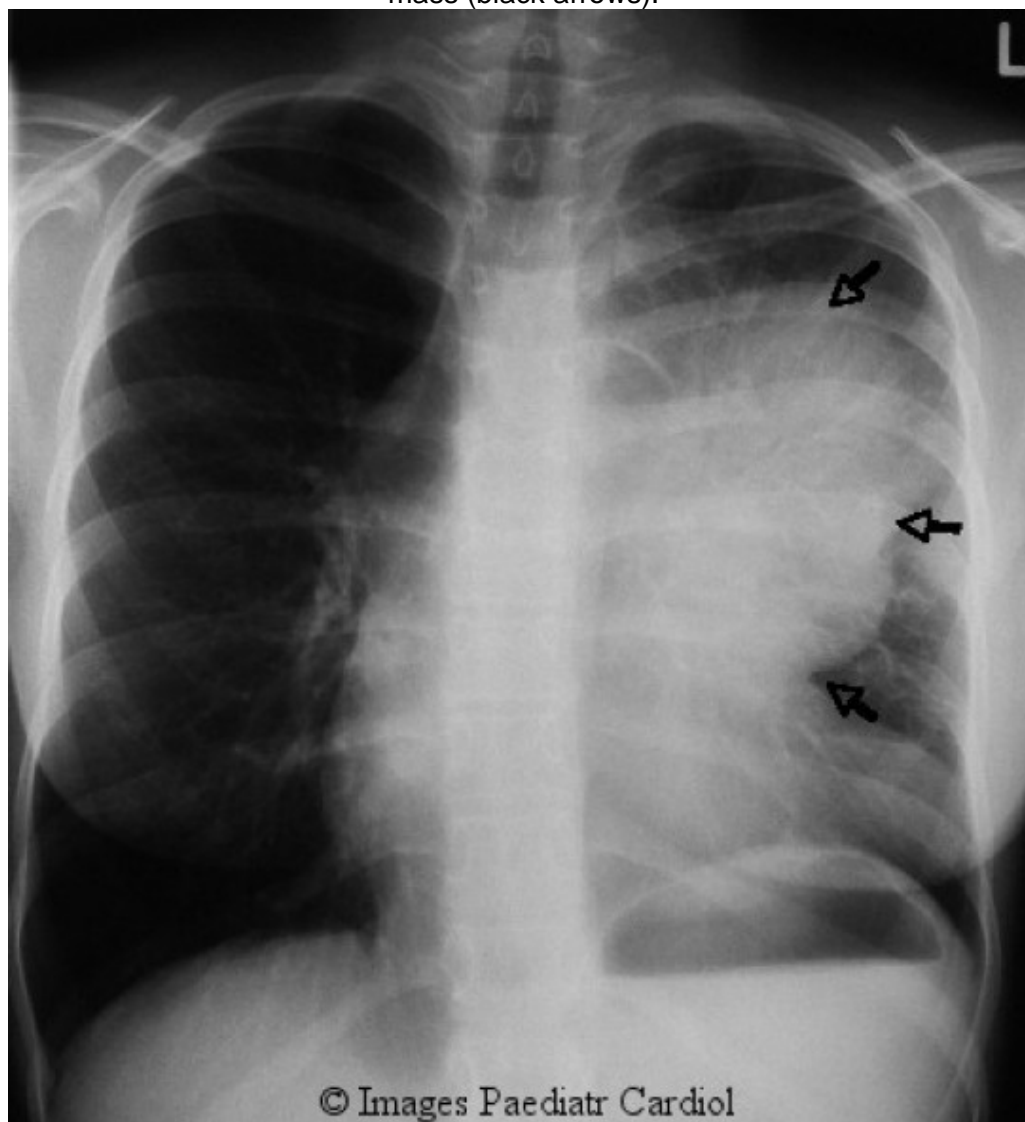
On admission for cardiac surgery, an indolent palpable mass extending from the fifth to seventh rib above the left breast was observed. Besides significant aortic regurgitation, routine pre-operative transthoracic color-coded and pulsed echocardiography revealed turbulent flow in the left pulmonary artery with a peak velocity of 3m/s caused by compression from a large echodense extrinsic mass. Chest radiography revealed marked left-sided mediastinal widening (Figures 2,3).

A subsequent computed chest tomography displayed an anterior mediastinal tumour with compression of the left pulmonary artery, severe narrowing of the left main stem bronchus, as well as infiltration of the middle mediastinum and anterior chest wall (Figures 4,5).

Peripheral blood cell count, uric acid and liver enzymes were normal, lactate dehydrogenase was slightly elevated.

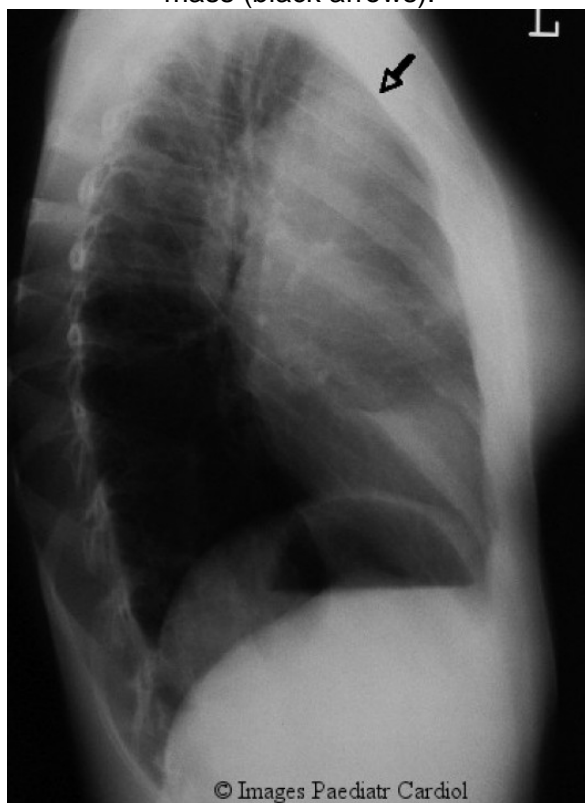
Primary mediastinal large B-cell lymphoma was diagnosed by open biopsy. Combined intensive chemotherapy according to the B-NHL-BFM 04 protocol so far showed a partial response. Tumour volume decreased with resolution of the left pulmonary artery stenosis and bronchus compression.

Figure 2 Biplane chest radiograph 9 months later showing an anterior mediastinal mass (black arrows).

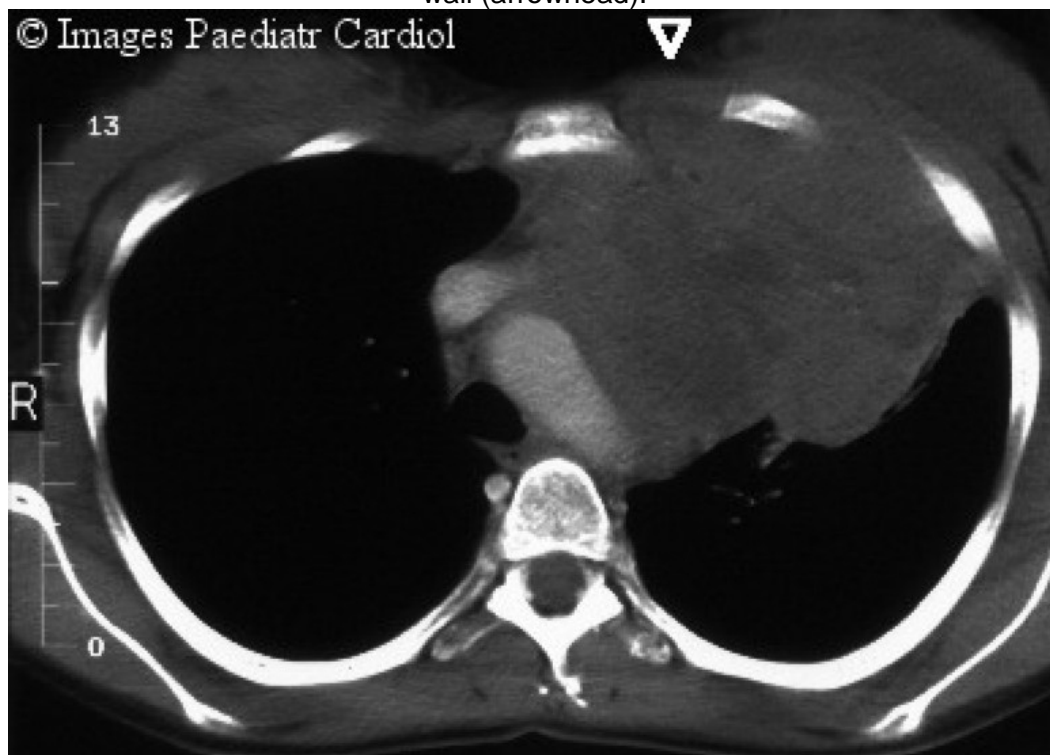


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Figure 3 Biplane chest radiograph 9 months later showing an anterior mediastinal mass (black arrows).

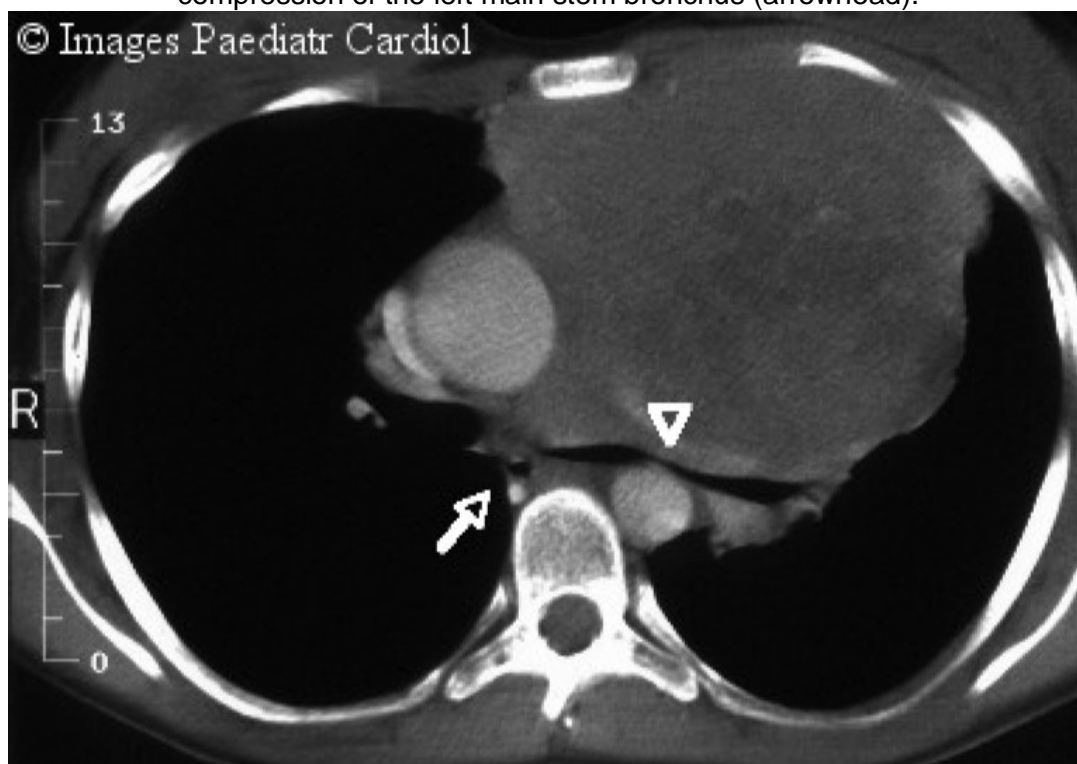


Figures 4 Computed chest tomography on admission. Contrast enhanced axial slices show a large mass in the anterior mediastinum with infiltration of the anterior chest wall (arrowhead).



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Figures 5 Computed chest tomography on admission. Contrast enhanced axial slices show a large mass with infiltration of the middle mediastinum (arrow) leading to compression of the left main stem bronchus (arrowhead).



Discussion

Primary mediastinal large B-cell lymphoma is predominantly affecting and taking its origin in the anterior mediastinum. Clinical symptoms including dyspnoea, atypical chest pain or cough, also suggestive for congestive heart failure, are attributable to the enlarging mediastinal mass with compression of the airways and great vessels. Diagnostic biopsy with distinct morphologic and immunophenotypic features ensures diagnosis.^{1–4}

Primary large B cell lymphoma often is misdiagnosed for dissecting aortic aneurysm or unknown pulmonary infection on chest radiograph as well as acquired pulmonary stenosis or right ventricular outflow tract obstruction on echocardiography.^{5–8}

We describe a patient suffering from aortic valvular stenosis. This was causing dilatation of the ascending aorta with slight mediastinal widening on frontal chest radiograph at first admission. During the next month dyspnoea, unproductive cough and chest pain developed and were interpreted as clinical signs of heart compromise, chest radiograph was not repeated at that point. For radiographic differentiation of mediastinal widening frontal and lateral projections are mandatory. Furthermore, detection of an acquired stenosis of the pulmonary arteries by echocardiography should lead to a thorough investigation of the mediastinum as acquired pulmonary stenosis is predominantly due to external compression.

Conclusion

Even though the majority of symptoms might fit the original cardiac diagnosis, a general work up should be performed, especially if symptoms persist or worsen despite adequate medical therapy.

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