Case study

Acquired extrinsic pulmonary stenosis secondary to a compressing mediastinal tumor

Abstract:

Acquired pulmonary stenosis in adults due to extrinsic compression of the pulmonary artery is a rare manifestation of non-Hodgkin's lymphoma (NHL). We describe here the case of a young patient who was diagnosed with acquired extrinsic pulmonary stenosis, secondary to a type B large cell lymphoma of mediastinal location.Our clinical case affirms the interest of transthoracic echocardiogramin the diagnosis as well as the follow-up of this unusual manifestation.

Keywords:

Pulmonary stenosis; mediastinal tumor; lymphoma; transthoracic echocardiogram

Introduction:

Acquired pulmonary stenosis is a rare lesion whose mechanism can be integrated or extrinsic to the pulmonary artery. External compression of the pulmonary artery or right ventricular outflow tract can lead to extrinsic abnormalities, while intrinsic damage usually involves the pulmonary valve itself.

We describe here the case of a young patient who was diagnosed with acquired extrinsic pulmonary stenosis, secondary to a type B large cell lymphoma of mediastinal location.

Case presentation:

Female patient, 25 years old, with no particular pathological history or cardiovascular risk factors. The patient reported, on admission, NYHA stage II dyspnea, a chronic dry cough, evolving over a period of five months, resistant to antitussives, without sputum or hemoptysis. There have also been reports of intermittent palpitations, feverish sensations, profound asthenia, anorexia and unquantified weight loss.

During her examination, we found a patient, conscious (Glasgow score 15/15), a low BMI of 17, hemodynamically and respiratory stable with a blood pressure of 121/64 mmHg symmetrical to both upper limbs, a heart rate 92 beats per minute, respiratory rate 22 breaths per minute, oxygen saturation 96% on room air, and temperature 37.4°C.

Examination found no signs of left or right heart failure. Auscultation revealed a regular rhythm with a systolic murmur rated 3/6th at the level of the pulmonary focus. We also noted slight jugular turgidity and edema of the neck and face. No adenopathy was found.

The electrocardiogram showed sinus tachycardia of 94 beats per minute and incomplete right bundle branch block without other abnormalities.

A thoracic CT angiography was performed, having ruled out the diagnosis of a pulmonary embolism, and revealing a reduction in the caliber of the trunk of the pulmonary artery as well as its dividing branches. He also showed a voluminous locally infiltrating and compressive left mediastinum-hilar lymph node complex, responsible for thrombosis of the terminal portion of the internal jugular veins, subclavian vein and left brachiocephalic venous trunk(Figures 1-2).

On transthoracic echocardiogram (TTE), the heart is pushed back to the right side; the major and minor axis para-sternal view obtained with difficulty in right latero-sternal. The trunk of the pulmonary artery and its branches are compressed by the mediastinal mass. The diameter of the trunk of the pulmonary artery is reduced to 9 mm, responsible for a maximum gradient at 55 mmHg with a Vmax at 3.5 m/s. A gradient of obstruction is found at the pulmonary infundibulum. The right ventricle is of limited size, with a basal diameter of 40 mm, and a preserved systolic function. The gradient between the right atrium and the right ventricle was 80 mmHg. TTE also revealed moderate pericardial effusion with no haemodynamic impact(Figures 3-6).. The other TTE parameters were normal

A biopsy with pathological study confirmed the diagnosis of non-Hodgkin type B large cell lymphoma.

Initially, our patient received 3 sessions of chemotherapy, over a period of 4 weeks, with clinical improvement. A weekly TTE control was carried out highlighting the regression of the pericardial effusion, the regression of the tumor compression, the progressive recovery of normal diameter of the trunk of the pulmonary artery and the right ventricle, and reduction of the maximum pulmonary gradient arriving at a value of 23 mmHg(Figures7-9). Regarding venous thromboses, we opted for anti-coagulation with Rivaroxaban.

Discussion:

Serious cardiovascular and tracheobronchial complications can result from anterior mediastinal masses. Right ventricular outflow tract obstruction and pulmonary stenosis are two such rare but increasingly well-known manifestations (1,2). This may be due to the tendency of mediastinal tumors to grow laterally rather than anteroposteriorly(3).

According to a recent literature review, the following conditions can lead to acquired extrinsic pulmonary stenosis: anterior mediastinal tumors, aortic aneurysms, mediastinal cysts, benign or malignant sternal tumors, fibrosing mediastinitis, and pericardial disease(4). In a series of case reports, NHL was responsible for only 9% of cases of extrinsic compression of the pulmonary artery by tumor (5).

The most common complaints are chest pain (69%) and dyspnea (60%), with a systolic ejection murmur occurring in 81% of cases(6). Cough (14%), fatigue (11%), palpitations (11%) and weight loss (14%) were sporadic observations(5).

Diagnosis of pulmonary stenosis and right ventricular outflow tract obstruction caused by cardiac and mediastinal neoplastic tumors has recently been facilitated by noninvasive echocardiographic techniques (2,7).

For malignant compression of the pulmonary artery, the goal of treatment is primarily symptomatic (8,9). Intervention is generally considered in the event of significant elevation of right ventricular pressure and/or right ventricular dysfunction, severe pulmonary insufficiency or worsening hemodynamic(9). Catheter-based pulmonary artery stenting was first reported in 1998 by Muller-Hulsbeck and colleagues(10).

Conclusion:

Although rare, acquired extrinsic pulmonary stenosis can cause serious complications. Transthoracic echocardiogramis a non-invasive tool allowing the follow-up and the choice of the adequate therapeutic intervention when it proves to be necessary.

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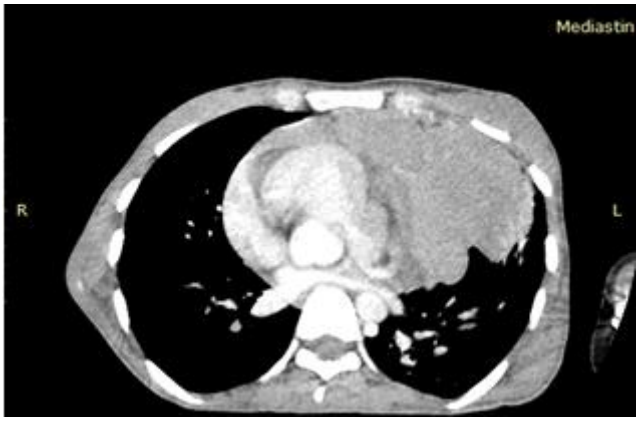
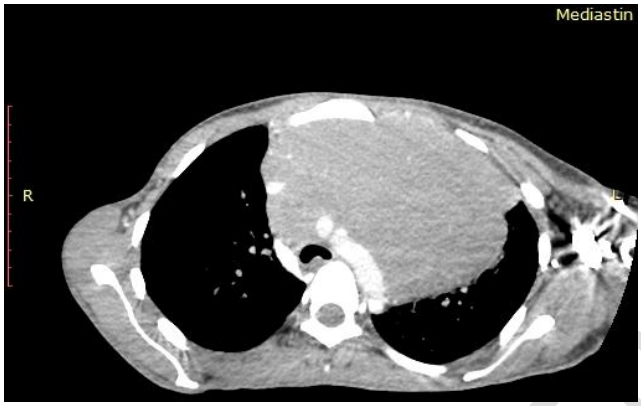
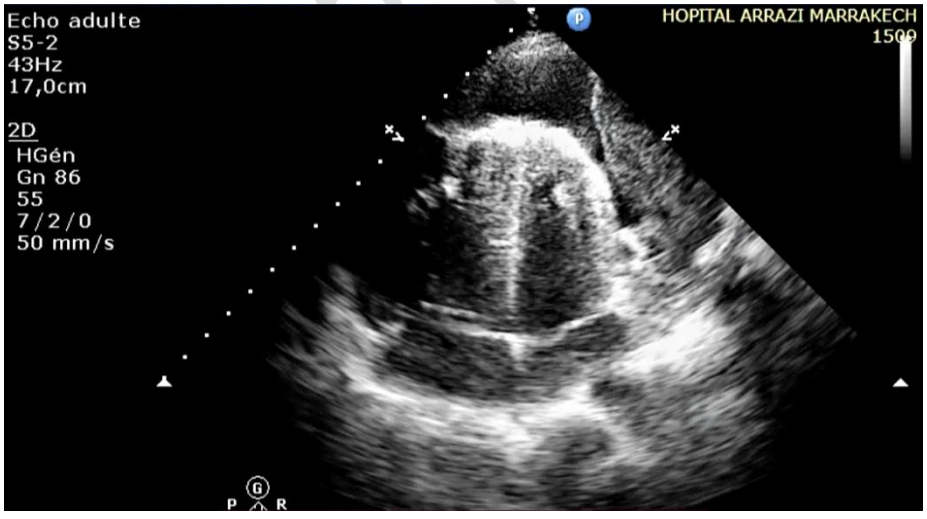
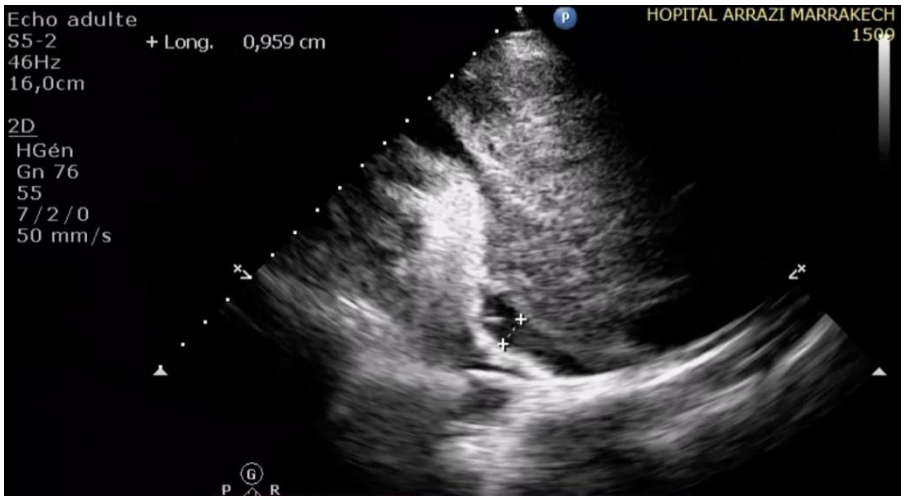


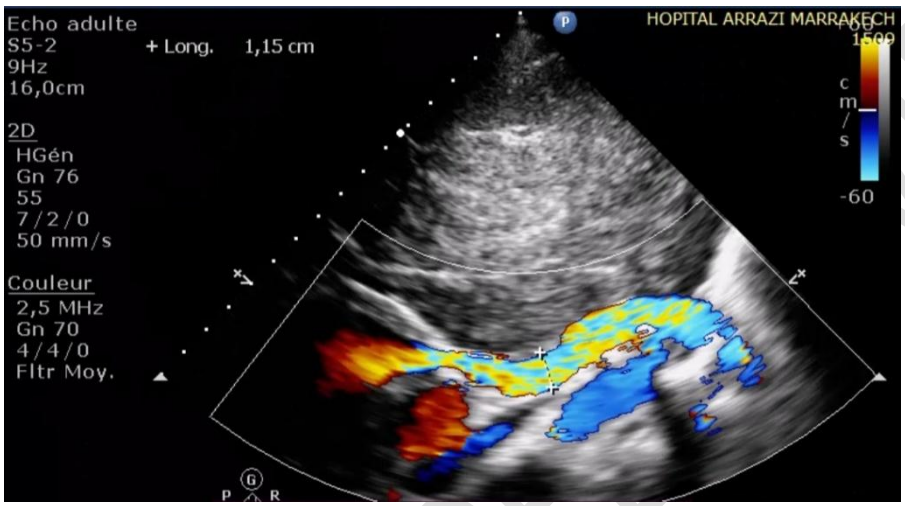


- **Figures 1 and 2:** thoracic CT angiography revealing:
    - Reduction in the caliber of the trunk of the pulmonary artery as well as its dividing branches.
    - Voluminous left mediastinum-hilar lymph node complex.

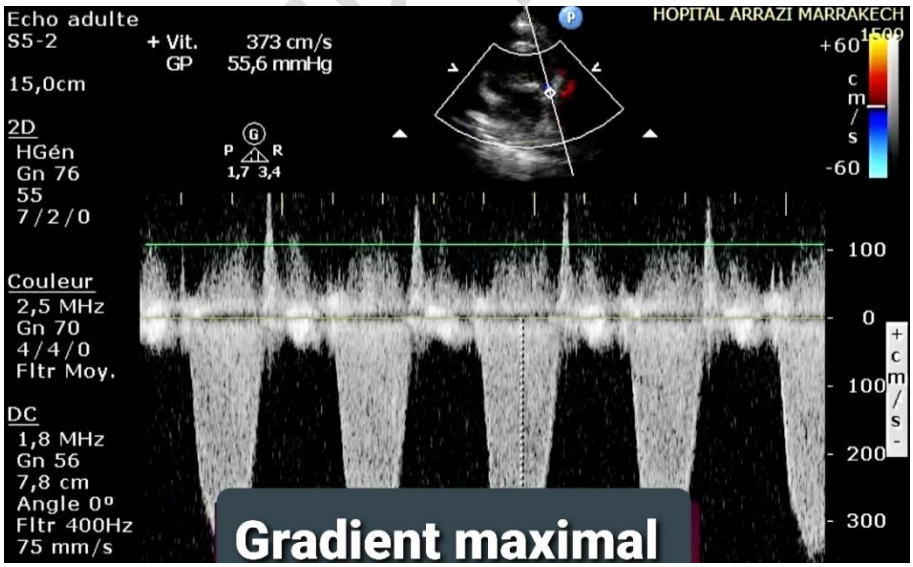


- **Figure 3 (TTE, Apical four chamber view):** heart pushed back to the right; right ventricle of limited size (40 mm); moderate pericardial effusion.

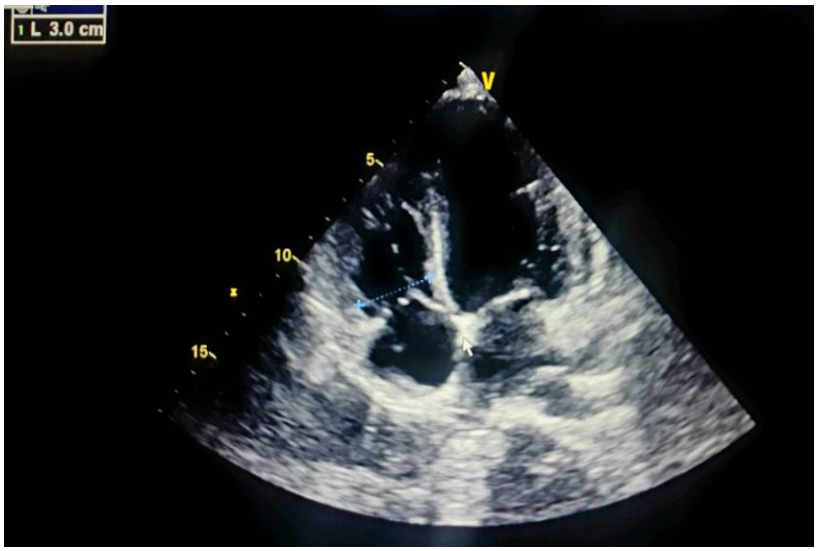




- **Figures 4 and 5 (TTE, Parasternal short axis view):**trunk of the pulmonary artery compressed by the mediastinal mass (diameter of 9 mm), with an aliasing in Color Doppler.



- **Figure 6:**maximum pulmonary gradient at 55.6 mmHg.



- **Figure 7 (TTE, Apical four chamber view):**regression of the tumor compressionand the pericardial effusion; recovery of normal diameter of the right ventricle.

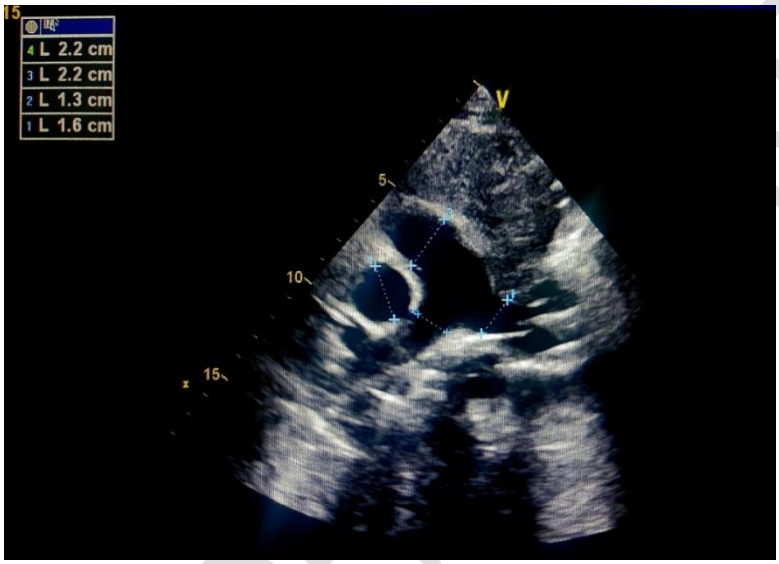


Figure 8 (TTE, Parasternal short axis view):recovery of normal diameter of the trunk of the pulmonaryarteryand its branches.

Figure 9:reduction of the maximum pulmonary gradient arrivingat 23 mmHg.