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). The incidence remains poorly known in the literature since only a few sporadic cases are described.

The clinical management is directed towards the treatment of the underlying disease. According to the latter, the patient may benefit from chemotherapy, radiotherapy, angioplasty, stenting, or surgery.

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 a transthoracic echocardiogram in 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, resistance to antitussives, and no 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 who was conscious (Glasgow score 15/15), had a low BMI of 17, was hemodynamically and respiratoryly stable, with a blood pressure of 121/64 mmHg symmetrical to both upper limbs, a heart rate of 92 beats per minute, a respiratory rate of 22 breaths per minute, oxygen saturation of 96% on room air, and a temperature of 37.4°C.

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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 an incomplete right bundle branch block without other abnormalities.

A thoracic CT angiography was performed, ruling 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 a transthoracic echocardiogram (TTE), the heart is pushed back to the right side; the major and minor axis para-sternal views are obtained with difficulty in the 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 of 55 mmHg with a Vmax of 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 preserved systolic function. The gradient between the right atrium and the right ventricle was 80 mmHg. TTE also revealed moderate pericardial effusion with no hemodynamic impact (Figures 3–6). The other TTE parameters were normal.

A biopsy and 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 the normal diameter of the trunk of the pulmonary artery and the right ventricle, and the reduction of the maximum pulmonary gradient to a value of 23 mmHg (Figures 7-9). Regarding venous thromboses, we opted for anticoagulation 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). While thymomas are the most common tumors of the anterior mediastinum that can cause extrinsic pulmonary stenosis (6,7).

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The most common complaints are chest pain (69%) and dyspnea (60%), with a systolic ejection murmur occurring in 81% of cases (8). Cough (14%), fatigue (11%), palpitations (11%), and weight loss (14%) were sporadic observations (5).

The 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 (2,9,10). This diagnosis can be verified, and the degree of obstruction can be determined, using right cardiac catheterization with pulmonary angiography. CT and MRI are less invasive yet nonetheless effective diagnostic tools (11).

On cross-sectional imaging, extrinsic compression of the pulmonary artery or its central branches manifests as compression, displacement, distortion, extrinsic indentation, luminal narrowing, and, in extreme cases, as a slit-like lumen or total occlusion. Right ventricular hypertrophy, pulmonary regurgitation, and tricuspid valvular regurgitation can all be secondary effects of chronic compression (11).

For malignant compression of the pulmonary artery, the goal of treatment is primarily symptomatic (12,13). Intervention is generally considered in the event of significant elevation of right ventricular pressure and/or right ventricular dysfunction, severe pulmonary insufficiency, or worsening hemodynamics (13). Catheter-based pulmonary artery stenting was first reported in 1998 by Muller-Hulsbeck and colleagues (14).

Several cases of angioplasty with stenting of the pulmonary arteries and sometimes of the superior vena cava are reported in the literature; they are performed in specific situations where chemotherapy and radiotherapy fail to reduce the mass or in the case of acute respiratory distress (15–17).

Total surgical resection may form the mainstay of treatment and be applied in combination with radiotherapy and chemotherapy, as in the case of advanced invasive thymomas or teratomas (6,18,19).

Conclusion:

Although rare, acquired extrinsic pulmonary stenosis can cause serious complications. Transthoracic echocardiogram is 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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Ethical Approval:

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

Consent

As per international standard or university standard, patient(s) written consent has been collected and preserved by the author(s).

References:

1. Gough JH, Gold RG, Gibson RV. Acquired pulmonary stenosis and pulmonary artery compression. Thorax. juill 1967;22(4):358‑67.

2. Putterman C, Gilon D, Uretzki G, Bar-Ziv J, Polliack A. Right ventricular outflow tract obstruction due to extrinsic compression by non-Hodgkin's lymphoma: importance of echocardiographic diagnosis and follow up. Leuk Lymphoma. juin 1992;7(3):211‑5.

3. Waldhausen JA, Lombardo CR, Morrow AG. Pulmonic stenosis due to compression of the pulmonary artery by an intrapericardial tumor. J Thorac Surg. mai 1959;37(5):679‑86.

4. Dalby AJ, Forman R. Acquired pulmonary stenosis. S Afr Med J. 10 févr 1979;55(6):218‑20.

5. Marshall ME, Trump DL. Acquired extrinsic pulmonic stenosis caused by mediastinal tumors. Cancer. 1 avr 1982;49(7):1496‑9.

6. Çap M, Erdoğan E, Akyüz A, Çap NK, Erdur E. Progressive pulmonary stenosis due to huge mediastinal thymoma. Anatol J Cardiol. 1 juill 2021;25(7):E-28-E-29.

7. Bushan K, Sharma S, Verma H. A Review of Thymic Tumors. Indian J Surg Oncol. juin 2013;4(2):112‑6.

8. Robinson T, Lynch J, Grech E. Non-Hodgkin's lymphoma causing extrinsic pulmonary artery compression. European Journal of Echocardiography. 1 juill 2008;9(4):577‑8.

9. Hsiung, M. C., Chen, C. C., Wang, D. J., Shieh, S. M. and, Chiang, B. N. Two-dimensional echocardiographic diagnosis of acquired right ventricular outflow obstruction due to extrinsic cardiac compression. Amer J Cardiol. 53^{e} éd. 1984;973-974.

10. Darbha K, Gupta AR, Suen IH, Toreli A. Rare anterior mediastinal mass causing pulmonary stenosis. Journal of the American College of Cardiology. 8 mars 2022;79(9_Supplement):3265‑3265.

11. Batra K, Saboo SS, Kandathil A, Canan A, Hedgire SS, Chamarthy MR, et al. Extrinsic compression of coronary and pulmonary vasculature. Cardiovasc Diagn Ther. oct 2021;11(5):1125‑39.

12. Fierro-Renoy C, Velasquez H, Zambrano JP, Ridha M, Kessler K, Schob A. Percutaneous stenting of bilateral pulmonary artery stenosis caused by malignant extrinsic compression. Chest. oct 2002;122(4):1478‑80.

13. Fang JX, Fang BX, Lam CS, Chan P, Tam FC, Siu C. Extrinsic pulmonary artery compression mimicking acute pulmonary embolism. Clin Case Rep. 17 août 2017;5(10):1591‑3.

14. Müller-Hülsbeck S, Bewig B, Schwarzenberg H, Heller M. Percutaneous placement of a self-expandable stent for treatment of a malignant pulmonary artery stenosis. Br J Radiol. juill 1998;71(847):785‑7.

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15. Giordano G, Fanzone L, Meo D, Piana S, Lentini V, Borracino S, et al. Stenting of the superior vena cava and right pulmonary artery in a woman with a mediastinal mass and acute respiratory distress syndrome (ARDS). Radiology Case Reports. 1 sept 2021;16(9):2437‑41.

16. Ponte C, Kerzmann A, Defraigne JO. [Pulmonary artery percutaneous angioplasty for extrinsic compression induced by squamous cell carcinoma]. Rev Med Liege. juill 2021;76(7‑8):592‑4.

17. Welby JP, Fender EA, Peikert T, Holmes DR, Bjarnason H, Knavel-Koepsel EM. Evaluation of Outcomes Following Pulmonary Artery Stenting in Fibrosing Mediastinitis. Cardiovasc Intervent Radiol. mars 2021;44(3):384‑91.

18. Chaudhry I ul H, Cheema A, Aqeel C, Alshaer AA, Alradei FG, Ali MG. A rare cause of right ventricle out flow tract obstruction: Anterior mediastinal teratoma. Annals of Medicine and Surgery. 1 févr 2021;62:258‑60.

19. Daly H, Horchani A. Mature teratoma of the anterior mediastinum revealed by supravalvular pulmonary stenosis: a case report. Pan Afr Med J. 28 oct 2022;43:109.

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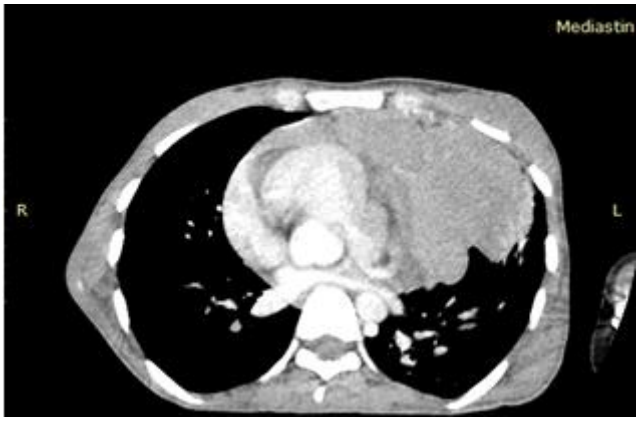
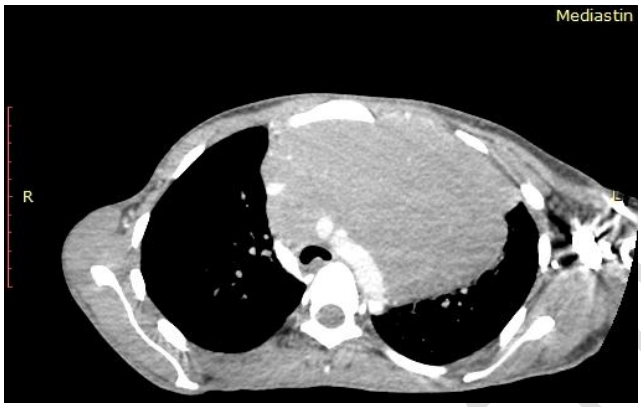
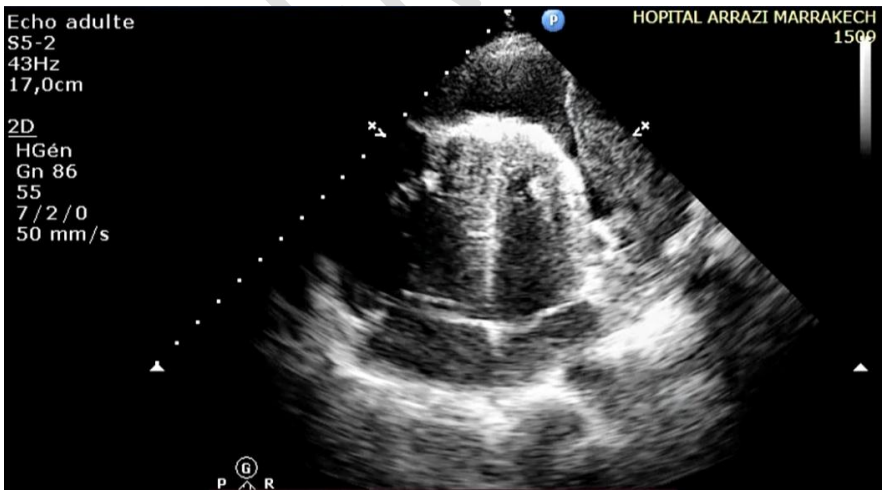




- **Figures 1 and 2:** thoracic CT angiography revealing:
    - Reduction in the caliber of the trunk of the pulmonary artery as well as its dividing branches.
    - Voluminous left mediastinum-hilar lymph node complex.



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

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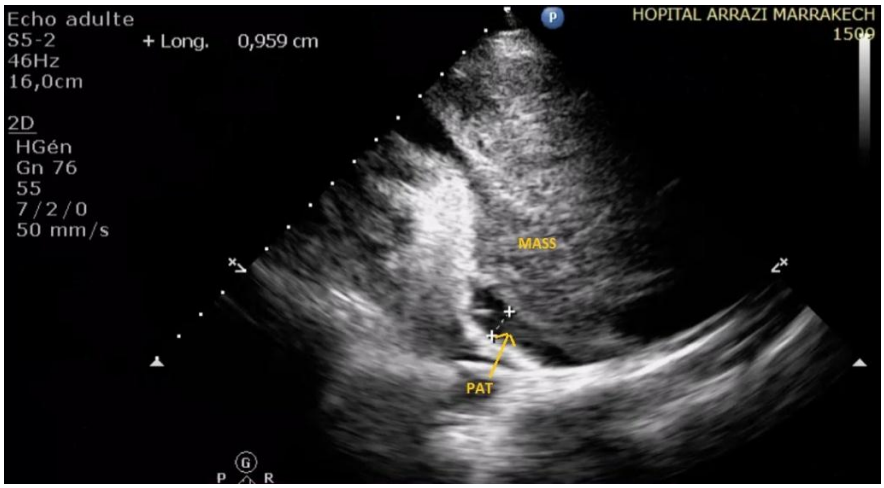
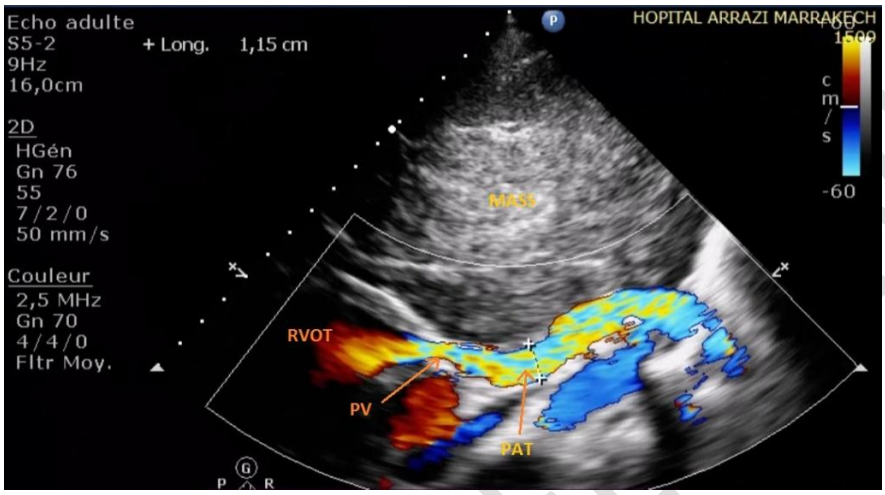
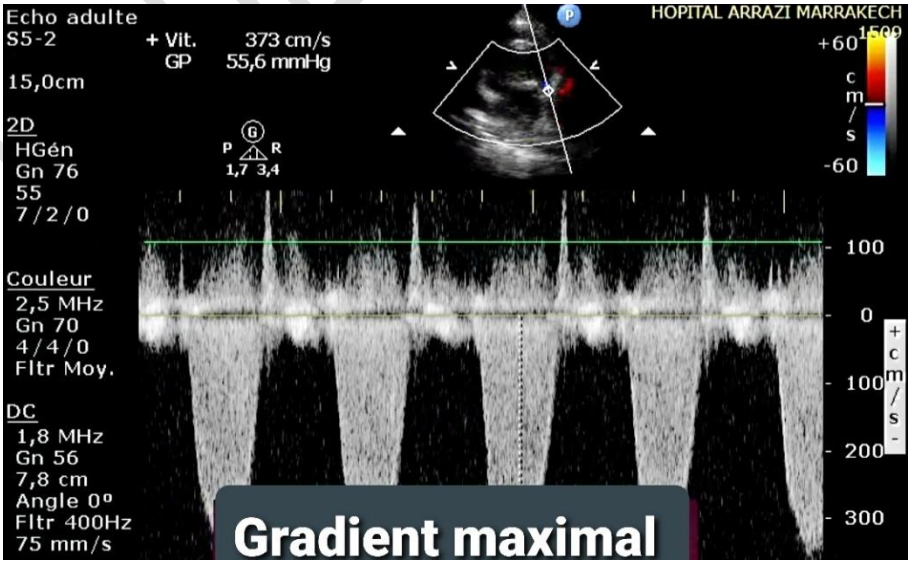
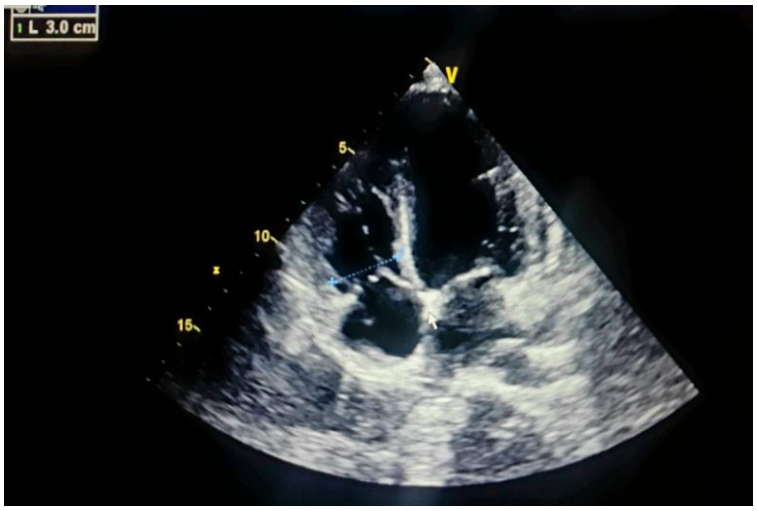




- **Figures 4 and 5 (TTE, parasternal short axis view, obtained with difficulty in the right latero-sternal because the heart is pushed back to the right side by the tumor):** trunk of the pulmonary artery (PAT) compressed by the mediastinal mass (diameter of 9 mm), with an aliasing in color Doppler.
  PAT: pulmonary artery trunk; **RVOT**: right ventricular outflow tract; **PV**: pulmonary valve



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



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

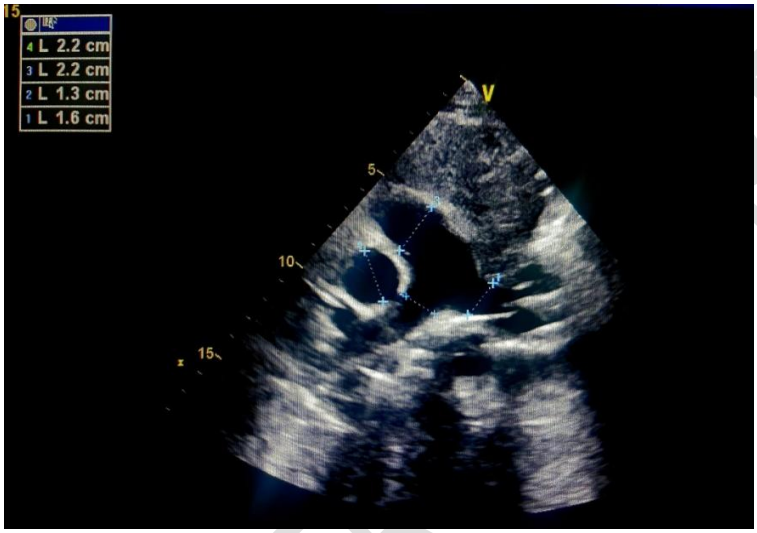


Figure 8 (TTE, Parasternal short axis view): recovery of normal diameter of the trunk of the pulmonary artery and its branches.

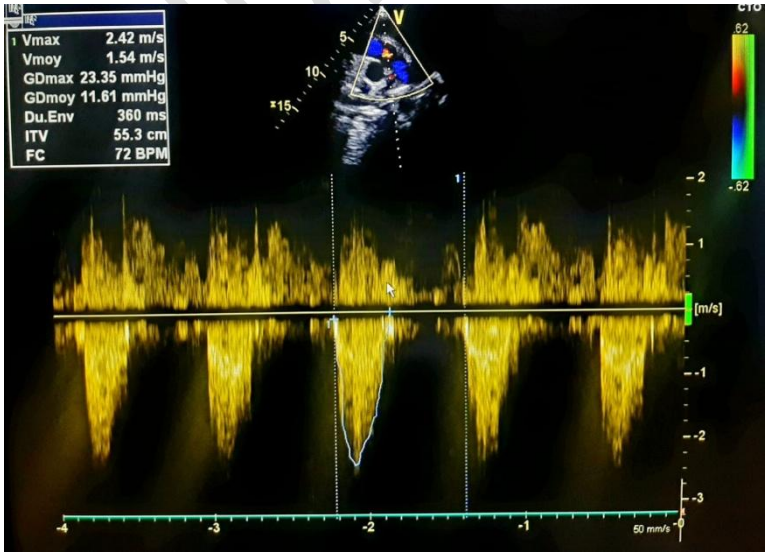


Figure 9: reduction of the maximum pulmonary gradient arriving at 23 mmHg.