

Acquired Pulmonary Stenosis in an Adolescent with Lymphoma

Estenose Pulmonar Adquirida em Adolescente com Linfoma

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Case report

A previously healthy 14-year-old male patient sought emergency care complaining of dyspnea. He was New York Heart Association (NYHA) functional class III and had a 1-month history of orthopnea and night sweats. Auscultation abolished in the left hemithorax, while a chest X-ray revealed extensive opacity in that region. Chest computed tomography (CT) showed a large mediastinal mass with concomitant involvement of the left pleura and retroperitoneal lymph nodes. No other palpable lymph nodes were present on physical examination.

The patient underwent pleuroscopy for pleural biopsy, which identified lymphoid cell proliferation in a “starry sky” pattern compatible with lymphoma and a histochemical pattern compatible with precursor T-cell lymphoblastic non-Hodgkin lymphoma (NHL) (pan-T and TdT, CD4 and CD4 co-expression, and high cell proliferation rate with positive Ki67 test results in 90%). Blast tests in the medulla and cerebrospinal fluid were negative.

Transthoracic echocardiography (TTE) was part of the workup to assess dyspnea and a potentially cardiotoxic pre-chemotherapy protocol. A mediastinal 12 × 5.5 cm mass with an anechoic and heterogeneous pattern adjacent to the great vessels and causing extrinsic compression with reduced pulmonary artery trunk diameter was visualized (Figure 1). Pulmonary artery flow was turbulent, with a high peak gradient measuring 44 mmHg. The pulmonary valves seemed normal (Figure 2 and Video 1).

He also had a mild anechoic pericardial effusion of 10 mm without hemodynamic repercussions. The longitudinal function was preserved with mitral annular plane systolic excursion of 12 mm and mitral S' wave of 9.9 cm/s. No other changes were identified, with an anatomically normal heart and acquired extrinsic valve disease related to the volume and site of the mediastinal mass. Chemotherapy was promptly initiated with a regimen consisting of cyclophosphamide, vincristine, and prednisone as well as daunorubicin.

Imaging tests (chest CT and TTE) were repeated after the treatment started, and the patient reported complete symptom improvement. CT showed complete mediastinal mass and pulmonary involvement remission in addition to a reduced number and volume of mesenteric and retroperitoneal lymph nodes (Figure 1). TTE showed preserved left ventricular

longitudinal function (GLS, -21%) and normal pulmonary artery flow without turbulence or systolic gradients. The patient was discharged on prophylactic Bactrim® and scheduled to return for chemotherapy maintenance.

From an oncological point of view, NHL is the fourth most common neoplasm in children and adolescents. Survival rates with current treatment exceed 80%, with T-cell or follicular lymphomas representing less than 5% of cases in this population. The Ann Arbor classification, widely used in the adult population, was not adapted for the pediatric population, with a preference for staging using the modified St. Jude classification, which considers the primary site of involvement, extranodal sites, and lymph node chain involvement above or below the diaphragm. Lymphoblastic lymphomas most commonly have a T-lineage (90%) and are diagnosed at more advanced stages (stage III-IV > 90%) with bone marrow (30%) and central nervous system (CNS) (5%) involvement. Treatment protocols include several potentially cardiotoxic drugs, such as anthracyclines and vincristine, in addition to corticosteroids and specific protocols for preventing CNS involvement (considered a “sanctuary” for lymphoma).

Mediastinal lymphomas commonly involve the great vessels with hemodynamically significant obstructions capable of generating murmurs or symptoms since they tend to grow laterally and not anteroposteriorly. In case of cardiac involvement, the most common symptoms are chest pain and dyspnea, with an audible murmur noted in 81% of patients. The prognostic implication of the presence of acquired pulmonary stenosis in NHL cases is unknown.

Acquired pulmonary stenosis is rare, with the congenital form being the most common and affecting up to 10% of the pediatric population in varying degrees of importance. Cases of extrinsic compression with supravulvar pulmonary stenosis or right ventricular outflow tract obstruction are even more uncommon, being reported only in case reports or series. Obstruction is usually caused by mediastinal tumors, the most frequent being lymphoma. Other possible causes include aortic aneurysm, mediastinal cysts, extra-mediastinal tumors, and fibrosing mediastinitis. Fixed pulmonary artery obstruction may contribute to symptoms of exertional dyspnea and orthopnea.

Long-term persistent obstruction can increase right chamber pressure, causing tricuspid insufficiency and, in the presence of patent foramen ovale, a right-to-left shunt with cyanosis, and an increased risk of paradoxical embolism. TTE has paramount importance in defining the etiology of pulmonary stenosis — in the case reported here, the mediastinal mass and its compressive effect are clearly visible in the parasternal short-axis window as well as the pulmonary valves with normal morphology, suggesting an external cause of the flow turbulence.

According to Tesoro-Tess et al., basal vessel involvement is more frequent in NHL (42%) than in Hodgkin's lymphoma

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Case Report

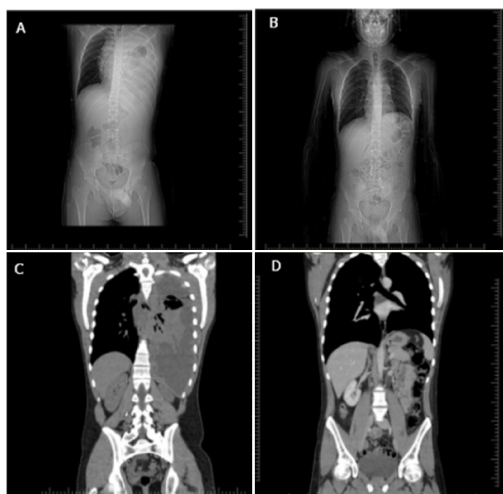


Figure 1 – Comparison of diagnostic (left images – A and C) and post-chemotherapy (right images – B and D) chest tomography images showing complete regression of the mediastinal mass. The images below (C and D) show the extrinsic pulmonary artery compression and its subsequent relief in the post-treatment period.

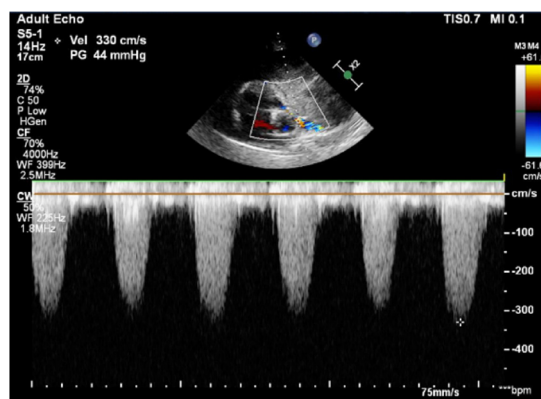


Figure 2 – Transthoracic echocardiogram in the parasternal short-axis window with continuous Doppler of the pulmonary artery showing a maximum velocity of 3.3 m/s and peak gradient of 44 mmHg due to extrinsic compression by the mediastinal mass.

(23%). Treatment is based on the underlying cause, consisting of chemotherapy, radiotherapy, or surgery depending on the type of mass identified. When the mass is successfully reduced, the pulmonary circulation flow is restored to normal and symptoms are expected to be resolved. With intensive treatment, refractory or recurrent disease occurs in 10–20% of patients, mainly with local recurrence in cases of T lymphoblastic lymphoma. In these cases, the cure rate decreases to 30%, so patients must maintain regular follow-up and closely monitor their symptoms.

References

1. Tesoro-Tess JD, Biasi S, Balzarini L, Ceglia E, Matarazzo C, Piotti P, et al. Heart involvement in lymphomas. The value of magnetic

Authors' contributions

Research conception and design: Voss TH; data collection: Voss TH; funding obtention: Voss TH; manuscript writing: Arantes FBB.

Conflict of interest

The authors have declared that they have no conflict of interest.

resonance imaging and two-dimensional echocardiography at disease presentation. *Cancer*. 1993;72(8):2484-90. doi: <https://doi.org/10.1002/cn.21000>

- doi.org/10.1002/1097-0142(19931015)72:8<2484::aid-cncr2820720828>3.0.co;2-w
2. WALDHAUSEN JA, LOMBARDO CR, MORROW AG. Pulmonic stenosis due to compression of the pulmonary artery by an intrapericardial tumor. *J Thorac Surg.* 1959;37(5):679-86. PMID: 13655327.
 3. Minard-Colin V, Brugières L, Reiter A, Cairo MS, Gross TG, Woessmann W, et al. Non-Hodgkin Lymphoma in Children and Adolescents: Progress Through Effective Collaboration, Current Knowledge, and Challenges Ahead. *J Clin Oncol.* 2015;33(27):2963-74. doi: <https://doi.org/10.1200/JCO.2014.59.5827>
 4. Lee HH, Wu XM, Chen YH, Lin LC, External compression of the main pulmonary artery by a diffuse large B-cell lymphoma, *Acta Cardiol Sin.* 2008;24:109-12.
 5. Ozer N, Deveci OS, Kaya EB, Demircin M. Mediastinal lymphoma causing extrinsic pulmonary stenosis. *Turk Kardiyol Dern Ars.* 2009;37(6):421-4. PMID: 20019459.
 6. Pugliatti P, Donato R, Grimaldi P, Nunnari F, de Gregorio C, Zito C, et al. Extrinsic pulmonary stenosis in primary mediastinal B-cellular lymphoma. *J Clin Ultrasound.* 2015;43(1):68-70. doi: <https://doi.org/10.1002/jcu.22133>
 7. Robinson T, Lynch J, Grech E. Non-Hodgkin's lymphoma causing extrinsic pulmonary artery compression. *Eur J Echocardiogr.* 2008;9(4):577-8. doi: <https://doi.org/10.1093/ejehocard/jen069>.