Article published online: 2024-06-04

The Cystic Appearance of a Mediastinal Mass Does Not Exclude Lymphoma: A Case Example of Classic Hodgkin Lymphoma in a Thymic Cyst and a Brief Literature Review

Samir Kahwash[1] Sheridan Chaves[1] Ashlie Patton[1]

[1] Department of Pathology and Laboratory Medicine, Nationwide Childrens Hospital, Columbus, Ohio, United States

Ibnosina J Med Biomed Sci

Address for correspondence Samir Kahwash, MD, Department of Pathology and Laboratory Medicine, Nationwide Childrens Hospital, Columbus, OH 43205, United States (e-mail: samir.kahwash@nationwidechildrens.org).

Abstract

Herein, we report a case of a 16-year-old girl who presented with an episode of shortness of breath while playing sports. Upon radiologic workup, she was found to have a large multicystic anterior mediastinal mass. The cystic appearance of mass and lack of constitutional symptoms contributed to favoring a benign cystic process and deferring surgical resection until the end of sport season. When surgery was performed 3 months later, microscopic examination showed findings diagnostic of classic Hodgkin lymphoma arising in a thymic cyst. Despite the rarity of such cases, it is important to remember that the cystic nature of an anterior mediastinal mass does not exclude lymphoma. Definitive diagnosis is not possible without examination of representative tissue.

Keywords

- anterior mediastinal
- cystic thymic masses
- Hodgkin lymphoma
- mediastinal mass
- Reed–Sternberg cells

Introduction

The mediastinum is the component of thorax located between the two lungs on both sides, the sternum anteriorly and spine posteriorly. In the pediatric age group, the mediastinum is the site for most common and most variable mass lesions of the thorax. Location of mediastinal mass lesions matters in considering a differential diagnosis. For example, the leading differential for an anterior mediastinal mass includes lymphomas (Hodgkin than non-Hodgkin), thymomas, thymic cysts, germ cell tumors, and thyroid lesions. The consistency of mass lesions (cystic vs. noncystic per radiologic impression) may influence the differential diagnosis as well, with a cystic nature favoring a benign diagnosis. However, exceptions, as in this case, are important to keep in mind.

Case Report

A 16-year-old girl presented to the emergency department following an episode of shortness of breath while playing sports. She had no pertinent past medical or surgical history. A physical exam noted a III/IV harsh holosystolic ejection type heart murmur devoid of rub, click, or gallop. An echocardiogram revealed a large, cystic anterior mediastinal mass with pulmonary artery stenosis and mass effect of the pulmonary artery. Computed tomographic-scan showed a large anterior mediastinal mass, 9.4 cm in greatest dimension. The mass had a large lobulated cystic component with thick walls. There were several associated enlarged lymph nodes, the largest was 1.5 cm.

The patient was emergently transferred to a pediatric cardiac unit for expedited oncology workup. She was

DOI https://doi.org/10.1055/s-0044-1785529.
ISSN 1947-489X.

Thieme Medical and Scientific Publishers Pvt. Ltd., A-12, 2nd Floor, Sector 2, Noida-201301 UP, India

asymptomatic on arrival at the cardiac unit and laboratory tests were normal and tumor markers were negative.

The results and treatment options were discussed with the family and surgery was decided in an elective setting. The patient and family elected to postpone surgery until after the end of the patient's school and sports season. Surgery was performed approximately 3 months following initial presentation and the mass was resected.

Pathologic Findings

Gross Pathologic Findings

The mass was received fresh and consisted of a 93.8 g, 11.2 cm in greatest dimension diffusely disrupted and cystic thymus with attached soft tissue. Sectioning of the specimen revealed tan-white, diffusely fibrotic, somewhat fleshy, and focally necrotic cut surfaces with a central, multiloculated cystic structure lined by a pale tan, slightly yellow and focally edematous fibrous wall (►**Fig. 1**). The cystic area contained red brown, somewhat diffluent, necrotic material. The remaining soft tissue contained tan-white, diffusely fibrotic, and focally necrotic lymph nodes. Touch preparations were made, and tissue samples of the solid and cystic areas were sent for flow cytometry.

Microscopic Findings

Microscopic examination demonstrated multilocular cystic, hemorrhagic thymic tissue (►**Fig. 2A**) with cystic areas intersected by thick fibrous bands entrapping nodules of Hodgkin lymphoma infiltrates consisting of Reed–Sternberg (R-S) cells and variants in a background of small lymphocytes, eosinophils, and neutrophils (►**Fig. 1B**). In areas, the cystic lining showed infiltrates of R-S cells (►**Fig. 2C** and **D**). Similar Hodgkin lymphoma infiltrates were seen surrounded or intersected by thick bands of collagen fibers (►**Fig. 3A** and **B**) alternating with clusters of R-S cells and variants (►**Fig. 3C** and **D**).

Immunohistochemistry stains were performed showing R-S cells and variant expressing CD30, CD15 with weak nuclear sating for PAX-5 supporting the diagnosis of classic Hodgkin lymphoma (CHL; ►**Fig. 4**).

The diagnosis of CHL, nodular sclerosing type, arising in a thymic cyst was established. Examination of bone marrow showed no involvement and additional radiologic workup determined extent of lymphoma as stage IIA.

The patient underwent port placement and the induction of chemotherapy (doxorubicin, bleomycin, vincristine, etoposide, and prednisone).

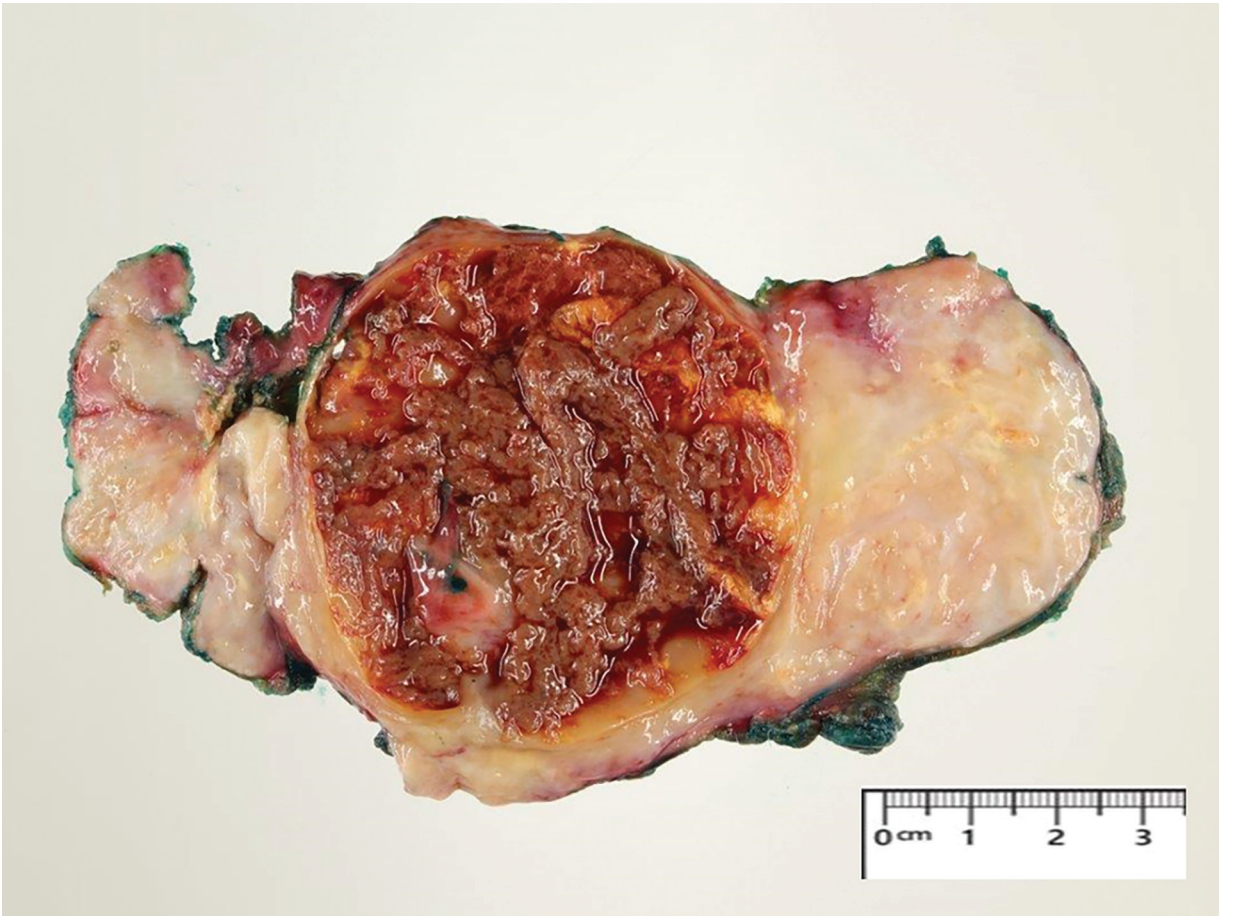

Fig. 1 Cut surface of the mediastinal mass showing a partially cystic central zone surrounded by solid fibrotic rim.

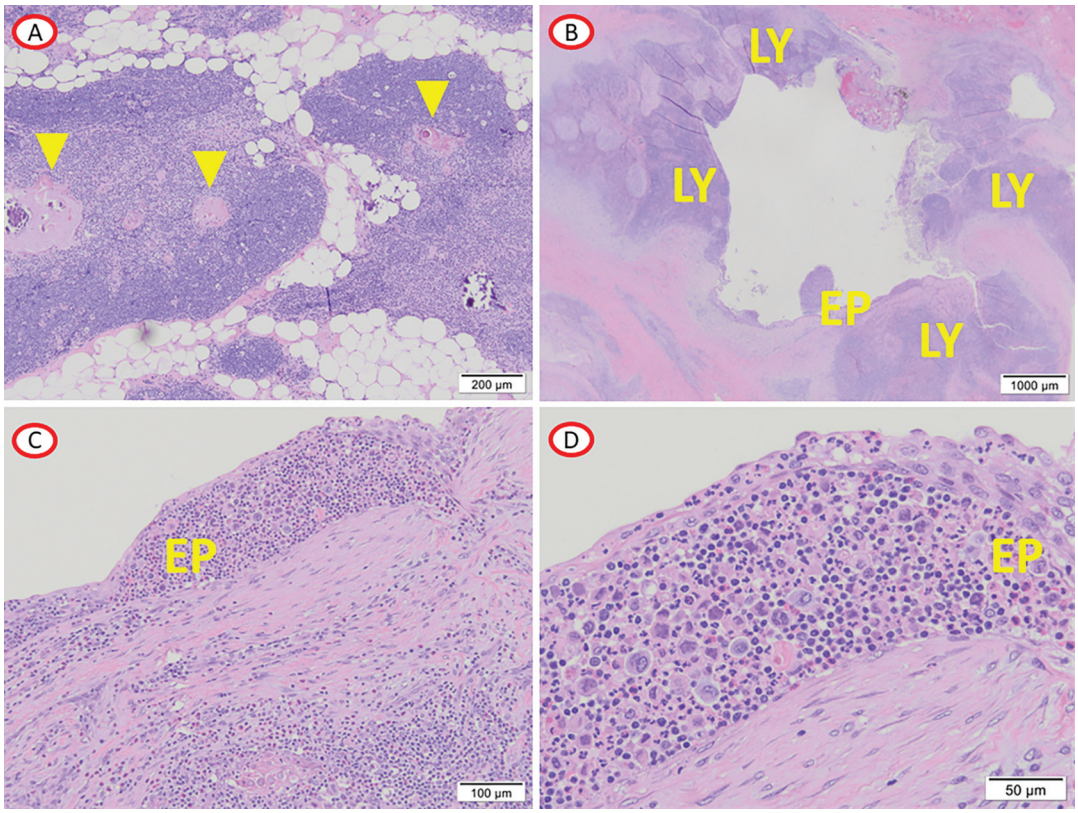

Fig. 2 Representative microscopic views showing thymic tissue (**A**) with Hassall's corpuscles (arrowheads), a cystic space (**B**) lined by lymphoid infiltrates (LY) and areas of squamous epithelial lining (EP). At higher power, the epithelial lining is infiltrated by Reed–Sternberg cells in a background of eosinophils, lymphocytes, and neutrophils (**C** and **D**).

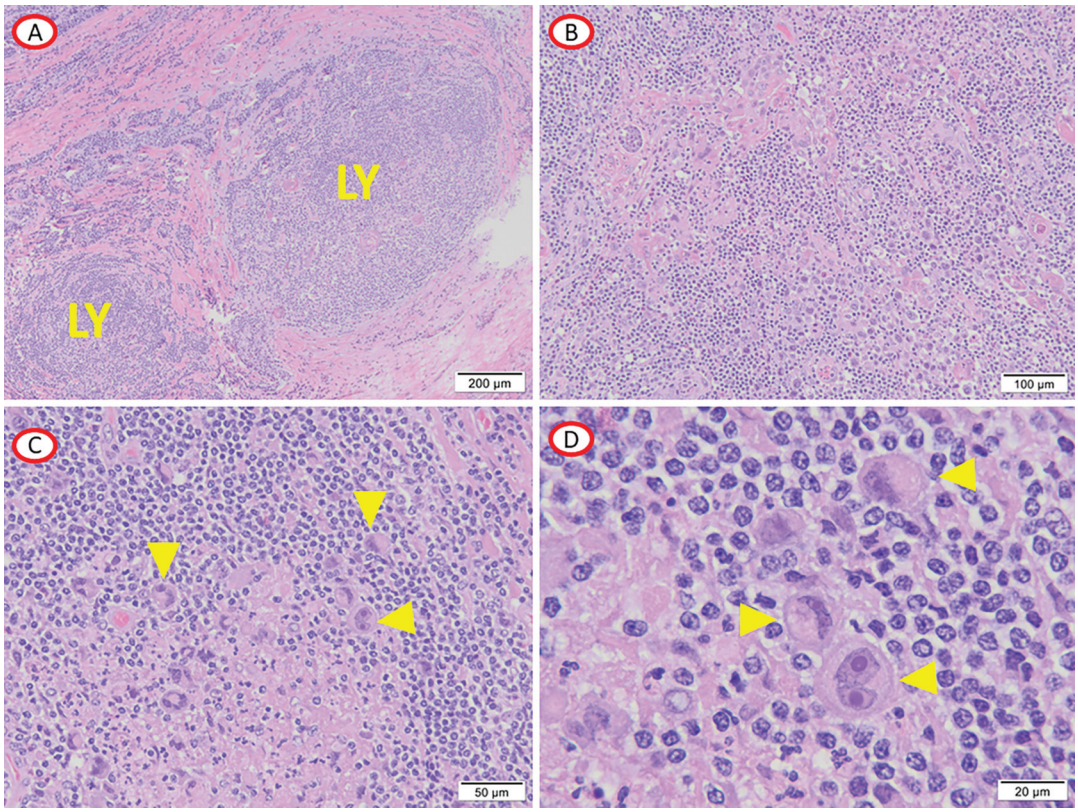

Fig. 3 Nodules of lymphoid infiltrates (LY) are seen surrounded or admixed with by dense fibrous tissue (**A** and **B**). These lymphoid infiltrates consist of Reed–Sternberg cells (arrowheads) in a background of lymphocytes and other inflammatory cells (**C** and **D**).

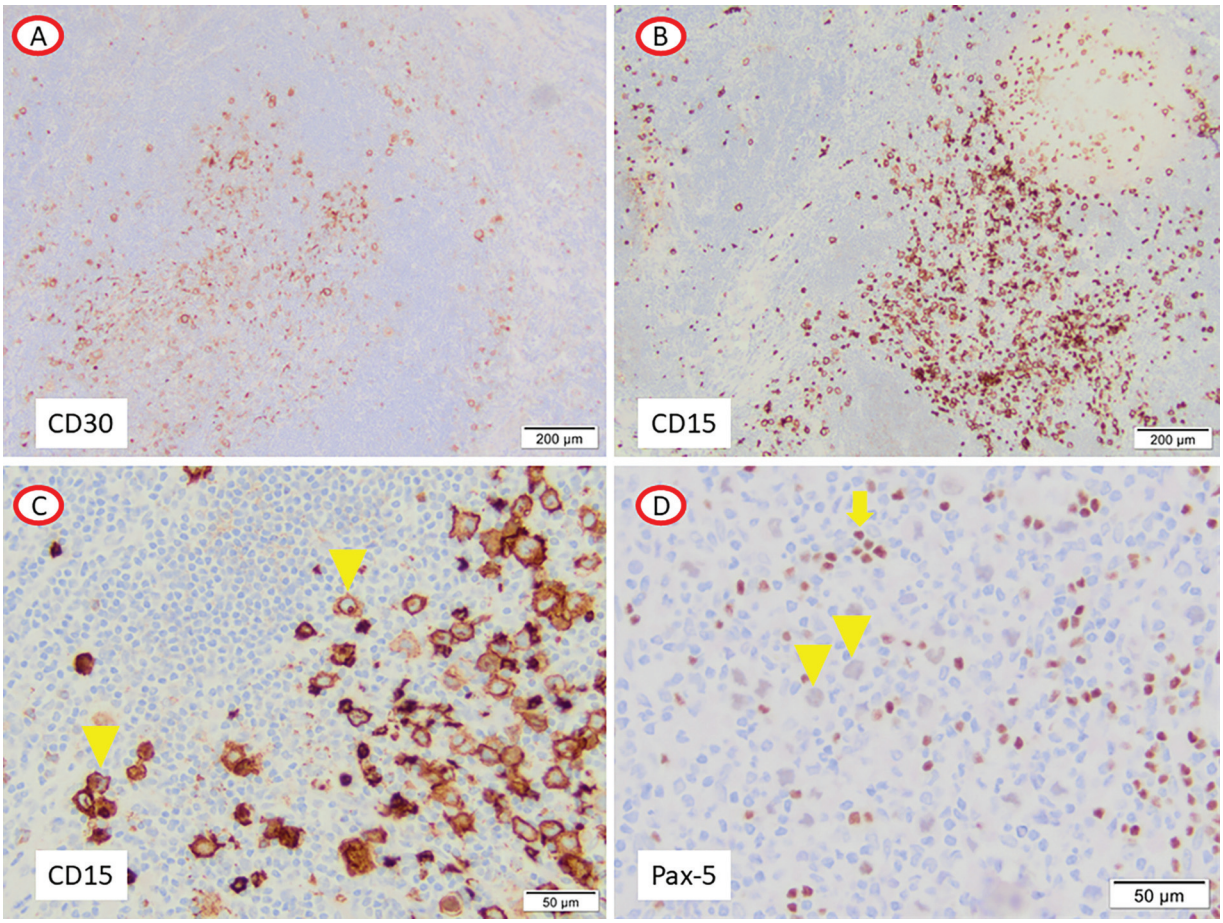

Fig. 4 Reed–Sternberg cells and variants expressed CD30 (**A**) CD15 (**B** and **C**) with a characteristically weak nuclear staining (**D**, arrowheads) compared with the string staining of associated background B-lymphocytes (arrow).

Discussion

Pediatric mediastinal mass lesions can be congenital, vascular, infectious, or neoplastic disorders. The location (anterior, posterior, or superior mediastinal localization) and radiologic appearance (cystic vs. solid) of lesion can help narrow the differential diagnosis. For example, an anterior mediastinal location raises the differential diagnosis of entities such as lymphoma, thymoma, thymic cyst, germ cell tumor, thyroid and parathyroid lesions, hemangioma, paraganglioma, and lipoma.[1] In the posterior mediastinum, commonly encountered entities include neurogenic tumors and gastroenteric cysts.[1] Furthermore, as a rule, a cystic radiologic appearance typically favors such entities as thymic cyst, cystic thymoma, duplication cysts, bronchogenic cyst, pericardial cyst, and lymphangioma.[2]

CHL presenting as a mediastinal cyst is uncommon and can be diagnostically challenging, especially when constitutional symptoms are not present as in the current case.[3–6] Reported cases of CHL involving thymic cysts usually describe a multiloculated cyst lined by banal-appearing squamous, columnar, or cuboidal epithelium.[7,8] The histomorphology of CHL in the present case displayed a nodular architecture with interconnecting bands of thick fibrocollagenous tissue characteristic of the nodular sclerosing subtype and the characteristic infiltrates of R-S cells in the typical inflammatory cells background.

In the largest series of large thymic cysts involved by Hodgkin lymphoma, the nodular sclerosing type was reported in more than half of cases (60%) that is the most common subtype of CHL in the pediatric population and presents within the mediastinum and cervical regions in 60 to 80% of cases.[9] The etiology of this collision phenomenon has not been well elucidated. It is unclear whether the underlying Hodgkin lymphoma promotes cyst formation when present within the thymus or arises concurrently with thymic cyst. Some proposed that thymic cysts arise from cystic transformation of the medullary duct epithelium-derived structures induced by an inflammatory process.[10]

Of note, there are reports of CHL coinciding with cystic thymic masses described following treatment for CHL, often years after the initial diagnosis of lymphoma.[11–13] The clinical history is usually helpful in this context.

An important concern that is raised in the setting of a cystic mediastinal mass is sample error. Percutaneous needle biopsy is increasingly utilized due to the minimally invasive technique and expedited recovery. However, one disadvantage is the limited sample, particularly of the cyst wall that may miss small foci of potential CHL. Other reports documented this concern and highlighted the importance of adequate sampling.[3,5,10] Targeting the most metabolically active site or surgical resection may be a potential alternative if a suspected hematological malignancy is still a consideration.[2]

Conclusion

In conclusion, although CHL presenting as a cystic mediastinal mass is exceptionally rare, cases like the one presented here underscore the importance of keeping CHL in the differential diagnosis when evaluating a mediastinal mass even in the context of a cystic radiologic appearance.

Authors' contributions

All authors contributed to the conception of the case report, its drafting and finalizations. They all reviewed and approved the final version.

Funding

None.

Conflict of Interest

None declared.

References

1 Goldblum JR, Lamps LW, McKenney JK, et al. Eds. Rosai and Ackerman's Surgical Pathology. Eleventh edition. Cambridge, MA, Elsevier; 2018
2 Bhunia N, Ondrejka S, Flagg A, et al. Classical Hodgkin lymphoma of cystic thymus in a patient with infectious mononucleosis: diagnostic challenges and practical considerations. Pediatric Hematology Oncology Journal. 2021;6(02):81–83
3 Steinmüller F, Traeger T, Kühn JP, et al. Rare case of a large mediastinal cyst involved in Hodgkin's lymphoma. Pathol Res Pract 2012;208(05):306–309
4 Almarzooqi S, Hammond S, Kahwash SB. Hodgkin lymphoma in a thymic cyst: report of a case with multiple secondary neoplasms. Case Rep Med 2010;2010:795037
5 Kocaman G, Özakıncı H, Yüksel S, et al. Three in one: a case presented with multiloculated thymic cyst, thymic Hodgkin lymphoma and pericardial cyst. Turk Gogus Kalp Damar Cerrahisi Derg 2023;31(02):289–294
6 Lewis CR, Manoharan A. Benign thymic cysts in Hodgkin's disease: report of a case and review of published cases. Thorax 1987;42 (08):633–634
7 Whitten CR, Khan S, Munneke GJ, et al. A diagnostic approach to mediastinal abnormalities. Radiographics 2007;27(03):657–671
8 Çinar HG, Gulmez AO, Üner Ç, et al. Mediastinal lesions in children. World J Clin Cases 2023;11(12):2637–2656
9 Kahn JM, Pei Q, Friedman DL, et al. Survival by age in paediatric and adolescent patients with Hodgkin lymphoma: a retrospective pooled analysis of children's oncology group trials. Lancet Haematol 2022;9(01):e49–e57
10 Suster S, Rosai J. Multilocular thymic cyst: an acquired reactive process. Study of 18 cases. Am J Surg Pathol 1991;15(04):388–398
11 Case records of the Massachusetts General Hospital. Weekly clinicopathological exercises. Case 47-1982. Persistence of a mediastinal mass after chemotherapy for Hodgkin's disease. N Engl J Med 1982;307(22):1391–1397
12 Kim HC, Nosher J, Haas A, et al. Cystic degeneration of thymic Hodgkin's disease following radiation therapy. Cancer 1985;55 (02):354–356
13 Lindfors KK, Meyer JE, Dedrick CG, et al. Thymic cysts in mediastinal Hodgkin disease. Radiology 1985;156(01):37–41

