ASIAN JOURNAL OF PHARMACEUTICAL AND CLINICAL RESEARCH

INNOVARE ACADEMIC SCIENCES
Knowledge to Innovation

Vol 18, Issue 8, 2025

Online - 2455-3891
Print - 0974-2441

Case Report

DECODING THE RETROPERITONEAL MYSTERY: A CASE OF FOLLICULAR LYMPHOMA

RAVITEJA NALABOTHULA, TARUGUVANDLA SANDHYA RANI, BUDUMURI VENKATA SIVA KARTHIK, KEERTHI REDDY KARRA*

Department of General Medicine, SRM Medical College Hospital and Research Centre, SRM Institute of Science and Technology, Kattankulathur, Chengalpattu, Tamil Nadu, India.
***Corresponding author: Keerthi Reddy Karra; Email: karrakeerthireddy@gmail.com**

Received: 14 June 2025, Revised and Accepted: 26 July 2025

ABSTRACT

Retroperitoneal masses are rare and often difficult to diagnose due to similarities between benign and malignant conditions. Follicular lymphoma (FL), a common indolent subtype of non-Hodgkin lymphoma, can present atypically, complicating early identification. We report a 60-year-old male with abdominal distension, weight loss, and breathlessness. Imaging revealed a large retroperitoneal mass encasing major vessels and associated lymphadenopathy. Biopsy of an enlarged inguinal lymph node confirmed FL through characteristic immunohistochemical markers. The patient was treated with six cycles of R-CHOP chemotherapy, resulting in complete remission. This case highlights the importance of histopathological evaluation in differentiating FL from other retroperitoneal pathologies like retroperitoneal fibrosis. Early and accurate diagnosis enables timely initiation of appropriate therapy, improving patient outcomes. Clinicians should consider FL in the differential diagnosis of retroperitoneal masses with atypical clinical presentations.

Keywords: Retroperitoneal mass, Follicular lymphoma, Non-Hodgkin lymphoma, R-CHOP chemotherapy, Immunohistochemistry.

 DOI: http://dx.doi.org/10.22159/ajpcr.2025v18i8.55592. Journal homepage: https://innovareacademics.in/journals/index.php/ajpcr

INTRODUCTION

Retroperitoneal mass lesions are defined as lesions that arise from the retroperitoneal space. Based on the origin of the lesion, these lesions are categorized as primary or secondary, neoplastic or non-neoplastic masses. Compared to secondary lesions, primary lesions are rare. Of all primary retroperitoneal tumors, 33% are lymphomas. A prominent retroperitoneal mass is observed in 14% of non-Hodgkin lymphomas (NHL) patients. The presence of multiple subclasses and clinical presentation makes the diagnosis and course of treatment vary according to diagnosis [1].

Lymphomas are solid malignant neoplasms originating in lymphoid tissues, which include Hodgkin and NHL. NHL arises from precursors of B, T cells, and mature B, T cells and is more prevalent than Hodgkin's lymphoma. Depending on the rates of growth and dissemination, they are further divided into indolent and aggressive. The most common subtypes of NHL are diffuse large B-cell lymphoma (Bcl 2) (30%) and follicular lymphoma (FL) (20%). Furthermore, diagnosing NHL might be challenging due to their diverse histological appearances, clinical features at presentation, and several subgroups [2,3].

FL is a subtype of NHL, the most common indolent type. The presence of *Bcl 2* gene mutation with the t (14;18) chromosomal translocation causes anti-apoptosis, which leads to the accumulation of cells causing FL. A mixture of bigger non-cleaved B-lymphocytes (known as centroblasts) and smaller cleaved B-lymphocytes (known as centrocytes) with co-expression of CD10 and Bcl-2 immunohistochemistry markers is a pathognomonic characteristic of FL [3-5].

CASE PRESENTATION

A 60-year-old male was presented with breathlessness for 2 weeks and a 3-month history of abdomen distention and weight loss of 25 kg. The patient had a history of type 2 diabetes mellitus a year ago, which was controlled on a diet plan. On the admission, vital signs were stable. Physical examination revealed cachexia, pallor, bilateral painless external iliac and inguinal lymphadenopathy, decreased breath sounds on the left side of the chest, the abdomen was soft and distended, and fluid thrill was present.

Laboratory workup shows a hemoglobin level of 10.5 g/dL, white blood cell, and platelet count within normal Limits. Peripheral smear shows microcytic hypochromic red blood cells; total and differential counts are within normal ranges and exhibit normal morphology. A routine urine examination shows a sugar level of 2+; the rest were within normal ranges. Renal function tests, liver function tests, and serum electrolytes are all within normal ranges. Chest X-ray PA view revealed mild pleural effusion on the left side. Abdomen ultrasound revealed left-sided pleural effusion, massive ascites, and grade 1 prostatomegaly.

Contrast-enhanced computed tomography (CECT)-abdomen revealed large soft-tissue density mass lesion with lobulated margin seen in the retroperitoneum extending from pancreatic regions at D12-L1 to L4 vertebral level up to aortic bifurcation (Fig. 1a and b).The lesion encases the aorta, inferior vena cava, and the bilateral ureters without compression (Fig. 1c) with gross ascites. Multiple enlarged lymph nodes were seen in the bilateral external iliac and inguinal region (Fig. 1d). The possibility of retroperitoneal fibrosis and differential diagnosis of retroperitoneal lymphoma are to be considered.

CECT abdomen, the yellow arrow in the coronal section (Fig. 1a) indicates a large soft-tissue density mass with a lobulated margin in the retroperitoneum. In the axial section (Fig. 1b), the yellow arrow also points to the retroperitoneal mass, and the red arrow highlights the Aorta, with the contrast phase revealing the same lobulated mass in the retroperitoneum. In the sagittal section (Fig. 1c), the yellow arrow represents retroperitoneal fibrosis, and the red arrow points to the aorta in the contrast phase. The lesion encases the aorta, inferior vena cava, and bilateral ureters without causing compression, extending from the pancreatic region at the D12-L1 level to the L4 vertebral level and reaching up to the aortic bifurcation. In the axial section (Fig. 1d), the white arrow points to the enlarged inguinal and external iliac lymph nodes.

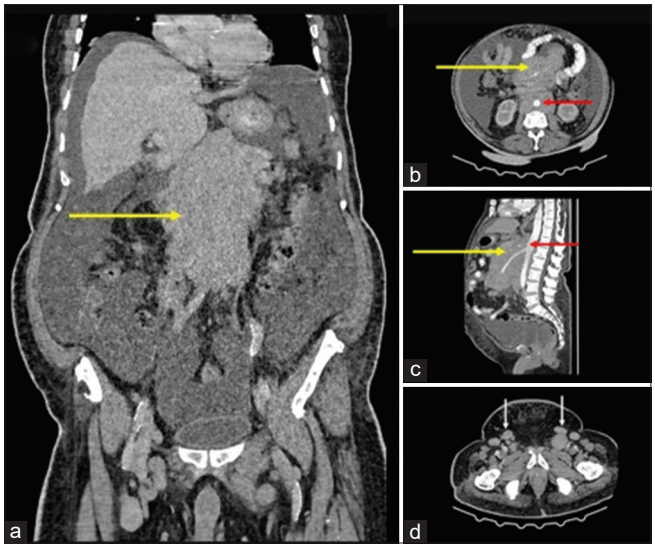

Fig. 1: (a-d) Contrast-enhanced computed tomography

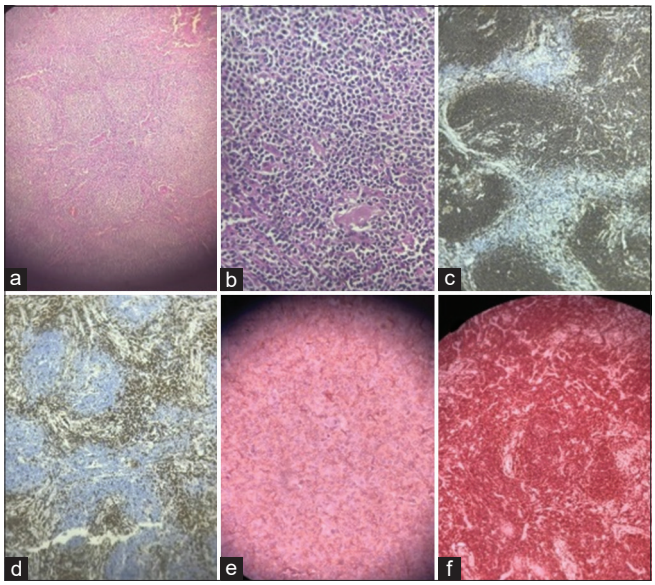

Fig. 2: Histopathology of the inguinal lymph node biopsy revealed (a) multiple follicles, (b) morphology of cells showing reactive changes, (c) CD20 membranous positivity predominantly observed in germinal centers and interfollicular areas, (d) CD3 positivity noted in reactive T-cells around follicles, (e) faint CD10 positivity in germinal centers, and (f) Bcl2 positivity in germinal centers and interfollicular areas (85–90%)

Histopathological examination of the inguinal lymph node biopsy demonstrated multiple follicles (Fig. 2a), with cellular morphology indicative of reactive changes (Fig. 2b). Immunohistochemical analysis revealed strong membranous CD20 positivity predominantly localized within the germinal centers and extending into the interfollicular areas (Fig. 2c). CD3 positivity was distinctly observed in reactive T-cells surrounding the follicles, emphasizing a reactive lymphoid environment (Fig. 2d). Additionally, faint CD10 positivity was noted within the germinal centers (Fig. 2e). Significantly, Bcl2 immunoreactivity was prominently positive in approximately 85–90% of the germinal centers and interfollicular regions, further supporting the reactive nature of the lymph node (Fig. 2f).

DISCUSSION

Lesions originating from the retroperitoneal area can be classified as primary or secondary, neoplastic, or non-neoplastic. While primary lesions are less common, they frequently include neoplastic lesions, among which lymphomas account for approximately 33% and are present in 14% of patients with NHL. Lymphomas, originating from lymphoid tissue, can be classified as either non-Hodgkin or Hodgkin, with non-Hodgkin types being more prevalent and diverse. FL, an indolent form of NHL, is characterized by a mutation in the *Bcl-2* gene and identified through immunohistochemistry.

FL has various presentations, ranging from asymptomatic, painless lymphadenopathy to different unique presentations like acute pancreatitis [6], and pleural effusion [7], which are not directly related to FL, making FL diagnosis difficult.

In this case, the patient initially presented with ascites and breathlessness. On evaluation, a retroperitoneal mass was identified on CECT abdomen imaging. Retroperitoneal masses can arise from neoplastic or non-neoplastic conditions, and treatment approaches vary significantly. The presence of the mass around the aorta and ureters led to suspicion of retroperitoneal lymphoma and retroperitoneal fibrosis. The presence of lymphadenopathy and weight loss is seen in both conditions, with ascites as rare presentations [7,8]. Distinguishing a retroperitoneal mass from inflammatory or malignant conditions is challenging. Morphological CECT imaging alone may not rule out malignancy. Some studies suggest that specific imaging findings, such as extension above renal vessels, enlarged lymph nodes, high fluorodeoxyglucose-positron emission tomography/CT uptake with an SUV max ≥4, splenomegaly, and lack of compression of the inferior vena cava or aorta, mainly if the location is centered cranially to L4/5, are more commonly associated with malignancy. However, histopathological confirmation may still be necessary when malignancy is suspected [9].

A lymph node biopsy was advised due to the patient's high risk for a retroperitoneal biopsy. The biopsy revealed BCL2 positivity, CD20 membranous positivity, CD10 faint positivity, CD3 positivity in reactive T cells around follicles, and a Ki67 index of 15% indicative of FL [10].

Retroperitoneal lymphoma and fibrosis closely resemble each other in clinicoradiological aspects and are often misdiagnosed. On the other hand, retroperitoneal fibrosis is a rare condition with an incidence rate of only one in a million, and its association with lymphoma is also rare. A case report by Lan *et al*. described a patient initially presenting with idiopathic retroperitoneal fibrosis, later found to be retroperitoneal fibrosis secondary to FL. Therefore, even if the condition is retroperitoneal fibrosis, close follow-up is needed, as lymphomas like FL are indolent and may take time to manifest. A review of previous literature and case reports revealed nine case reports in PubMed of lymphoma mimicking retroperitoneal fibrosis. These studies included one case of diffuse large Bcl 2, four cases of FL, two cases of anaplastic large cell lymphoma, and two cases of B-cell non-Hodgkin lymphoma [11].

FL International Prognostic Index (FLIPI) is bed side tool used for enhancing the prognostic evaluation of patients with FL.

The FLIPI score is based on the following five clinical factors; each positive factor carries 1 point, making the total score 5 points,

Clinical factors:

1. Age > 60 years old
2. Ann Arbor Stage III or IV disease
3. Hemoglobin level: A hemoglobin level of <12 g/dL in women or <13 g/dL in men
4. Number of lymph nodes: More than 4 involved lymph node areas (outside the abdomen)
5. Serum lactate dehydrogenase (LDH) level: Elevated LDH (greater than the upper limit of normal).

FLIPI Score	Risk group	10-year overall survival (%)
≤1	Low	70
2	Intermediate	50
≥3	High	35

Based on the FLIPI score, the prognosis of the patient varies, as shown in the table above [12,13].

The patient's FLIPI score is three, and based on the body surface area of the patient, he was treated with 712.5 mg of rituximab IV, 1425 mg of cyclophosphamide IV, 95 mg of doxorubicin IV, 2 mg of vincristine IV (maximum dose) on day 1, along with 100 mg of prednisone orally on day 1, and then 100 mg orally every 24 h from days 2 to 5 [14,15].

After six cycles of the R-chop regimen, the patient achieved complete remission status.

CONCLUSION

Due to their similar clinical and radiological characteristics, retroperitoneal lymphoma and retroperitoneal fibrosis can be challenging to diagnose. The biopsy results in this case show how vital histopathological examination, including immunohistochemical staining, is in making a firm diagnosis. This case also highlights the need to rule out lymphoma when making a differential diagnosis for retroperitoneal masses, especially in cases when the initial presentation points to benign or inflammatory disorders. This case illustrates this difficulty. Despite being an uncommon disorder, retroperitoneal fibrosis may be associated with lymphoma, especially FL, emphasizing the significance of careful monitoring and comprehensive assessment. To enhance patient outcomes, it is imperative to identify such situations early and handle them appropriately.

ACKNOWLEDGMENTS

Nil.

CONFLICTS OF INTEREST

None to declare.

FUNDING SOURCE

Nil.

REFERENCES

1. Gulati V, Swarup MS, Kumar J. Solid primary retroperitoneal masses in adults: An imaging approach. Indian J Radiol Imaging. 2022 Jul;32(2):235-52. doi: 10.1055/s-0042-1744142, PMID 35924125
2. Shankland KR, Armitage JO, Hancock BW. Non-Hodgkin lymphoma. Lancet. 2012 Sep;380(9844):848-57. doi: 10.1016/S0140-6736(12)60605-9, PMID 22835603
3. Singh R, Shaik S, Negi BS, Rajguru JP, Patil PB, Parihar AS, *et al*. Non-Hodgkin's lymphoma: A review. J Fam Med Prim Care. 2020 Apr;9(4):1834-40. doi: 10.4103/jfmpc.jfmpc_1037_19, PMID 32670927
4. PDQ Adult Treatment Editorial Board. Non-hodgkin lymphoma treatment (PDQ®). patient version. In: PDQ Cancer Information Summaries. Bethesda: National Cancer Institute (US); 2002.
5. Gaziano D, Akbar A, Lakhani K, Okafor TL, Bhesania S. Concurrent incidental presentation of primary retroperitoneal follicular lymphoma with acute pancreatitis. Cureus. 2020 Nov;12(11):e11687. doi: 10.7759/cureus.11687, PMID 33274167
6. Carreras J. The pathobiology of follicular lymphoma. J Clin Exp Hematop. 2023 Sep;63(3):152-63. doi: 10.3960/jslrt.23014, PMID 37518274
7. Weber B, Luke ND. Rapid onset chylous ascites presenting as the initial manifestation of follicular lymphoma: A case report. Cureus. 2022;14(7):e27199. doi: 10.7759/cureus.27199, PMID 36035034
8. Ur Rahman A, Ramaraj R, Humphreys E, Slowinska E. Retroperitoneal fibrosis as a cause of chylous ascites. ACG Case Rep J. 2017;4:e115. doi: 10.14309/crj.2017.115, PMID 29090239
9. Zhang S, Chen M, Li CM, Song GD, Liu Y. Differentiation of lymphoma presenting as retroperitoneal mass and retroperitoneal fibrosis: Evaluation with multidetector-row computed tomography. Chin Med J (Engl). 2017 Mar;130(6):691-7. doi: 10.4103/0366-6999.201606, PMID 28303852
10. Cho J. Basic immunohistochemistry for lymphoma diagnosis. Blood Res. 2022 Apr;57(Suppl 1):55-61. doi: 10.5045/br.2022.2022037, PMID 35483927
11. Lan X, Hu R, Song T, Gan L, Sun W. A case of follicular lymphoma mimicking idiopathic retroperitoneal fibrosis. Cureus. 2022 Nov;14(11):e32031. doi: 10.7759/cureus.32031, PMID 36600821
12. Solal-Céligny P, Roy P, Colombat P, White J, Armitage JO, Arranz-Saez R, *et al*. Follicular lymphoma international prognostic index (FLIPI). Blood. 2004;104(5):1258-65. doi: 10.1182/blood-2003-12-4434, PMID 15126323
13. Van de Schans SA, Steyerberg EW, Nijziel MR, Creemers GJ, Janssen-Heijnen ML, Van Spronsen DJ. Validation, revision and extension of the follicular lymphoma international prognostic index (FLIPI) in a population-based setting. Ann Oncol. 2009;20(10):1697-702. doi: 10.1093/annonc/mdp053, PMID 19549712
14. Watanabe T, Tobinai K, Wakabayashi M, Maruyama D, Yamamoto K, Kubota N, *et al*. R-CHOP treatment for patients with advanced follicular lymphoma: Over 15-year follow-up of JCOG0203. Br J Haematol. 2024;204(3):849-60. doi: 10.1111/bjh.19213, PMID 37996986
15. Tomita N, Takasaki H, Fujisawa S, Miyashita K, Ogusa E, Kishimoto K, *et al*. Standard R-CHOP therapy in follicular lymphoma and diffuse large B-cell lymphoma. J Clin Exp Hematop. 2013;53(2):121-25. doi: 10.3960/jslrt.53.121, PMID 23995108