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*Case Report*

# A Rare Case of Primary Mediastinal B-Cell Lymphoma – The Great Masquerade

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## ABSTRACT

Primary mediastinal B-cell lymphoma (PMBCL) is a relatively rare lymphoma subtype affecting mainly seen in young adults with female predominance. It constitutes approximately 2–4% of all non-Hodgkin's lymphomas (NHLs). No risk factors for this type have been identified but it may be related to 5533 C>A mutation in the MLL gene. Its molecular signature and clinical features resemble classical Hodgkin's lymphoma. PMBCL belongs to a group of aggressive diffuse large B-cell lymphomas. 2008 WHO classification distinguishes this lymphoma as a separate entity due to its specific clinical features and pathological features. Gene expression profile studies showed that it shares common features with classical Hodgkin's lymphoma. The optimal chemotherapy for this lymphoma subtype has not been established. Furthermore, no convincing data are supporting the use of radiotherapy. Relatively low patient numbers are the main obstacle in conducting randomised prospective trials. Hence, therapeutic decisions have been based mainly on retrospective studies.

**Keywords:** Primary mediastinal B-cell lymphoma, Non-Hodgkin's lymphoma, Positron emission tomography scan, Chemotherapy, Radiotherapy, MLL gene, Immunohistochemical examination

## INTRODUCTION

Primary mediastinal B-cell lymphoma (PMBCL) is a relatively rare lymphoma subtype affecting mainly seen in young adults with female predominance.[1-5] It constitutes approximately 2–4% of all non-Hodgkin's lymphomas (NHLs). No risk factors for this type have been identified but it may be related to 5533 C>A mutation in the MLL gene. Its molecular signature and clinical features resemble classical Hodgkin's lymphoma. PMBCL belongs to a group of aggressive diffuse large B-cell lymphomas.[2,5,6] 2008 WHO classification distinguishes this lymphoma as a separate entity due to its specific clinical features and pathological features. Gene expression profile studies showed that it shares common features with classical Hodgkin's lymphoma. The optimal chemotherapy for this lymphoma subtype has not been established. Furthermore, no convincing data are supporting the use of radiotherapy. Relatively low patient numbers are the main obstacle in conducting randomised prospective trials. Hence, therapeutic decisions have been based mainly on retrospective studies.[6-9]

## CASE REPORT

We present the case of a 19-year-old female who came to MGM Hospital with complaints of breathlessness on exertion and cough for 2-weeks. On enquiring further, she gave a history of weight loss of around 10 kg in 3 months and on and off fever. On doing a Skiagram of the chest,

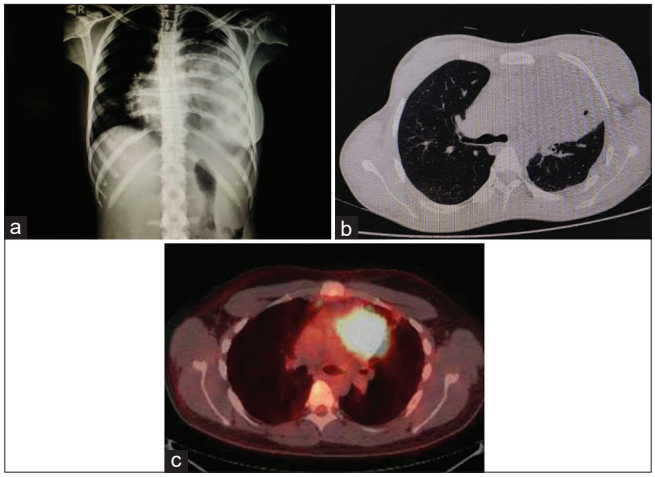


**Figure 1:** (a) Chest X-ray, (b) contrast-enhanced computed tomography chest, (c) positron emission tomography scan.

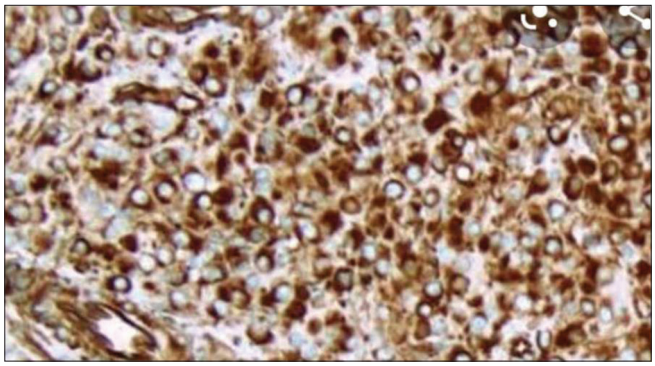

**Figure 2:** Vimentin-positive IHC biopsy specimen.

left sides shadow with blunting of costophrenic angle on the left side was seen with opacity in the left upper lobe suggestive of the left-sided pleural effusion and left upper lobe mass. An ultrasound (USG)-guided diagnostic and therapeutic pleural tap was done and around 200 ml of pleural fluid was tapped out. Infective aetiology was ruled out by running tests on the pleural fluid collected. This was followed by a contrast-enhanced computed tomography abdomen and chest for evaluation of the mass [Figure 1]. It revealed a neoplastic carcinomatous lesion of size 11.4 × 8.2 × 14.6 cm involving left upper lobe lung parenchyma with extension into the anterior mediastinum and enlarged necrotic prevascular and supraclavicular lymph nodes. The abdomen was clear suggestive of no metastasis to the abdomen. A USG-guided biopsy of the mass was performed and the sample underwent histopathological and immunohistochemical (IHC) examination which revealed a low-grade neoplasm and on running the IHC markers on it [Figure 2], the diagnosis of mediastinal B-cell NHL was made. A positron emission tomography (PET) scan was performed which showed increased uptake of FDG in the mediastinum and supraclavicular region [Figure 1]. The patient was started on an R-CHOP regimen for NHL and is on the same treatment currently with 2 cycles of chemotherapy completed. A repeat PET scan will be performed after 6 cycles of chemotherapy to check the progression of the tumour.

## CONCLUSION

Because PMBCL is uncommon, its clinical management varies across centres. There is no standard protocol for the treatment of PMBCL but chemotherapy R-CHOP and dose-adjusted E-POC regimen are shown to be beneficial. The role of radiotherapy is unclear. Recent research has brought new insight into molecular mechanisms contributing to the malignant phenotype of PMBCL and this could direct the development of targeted therapies.

### Declaration of patient consent

Patient's consent not required as patient's identity is not disclosed or compromised.

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Nil.

### Conflicts of interest

There are no conflicts of interest.

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