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Case Report

A Case Report of a Massive Pleural Effusion as an Unusual Presentation for a Rare Condition

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Abstract

Primary mediastinal B-cell lymphoma (PMBCL) is a rare but aggressive mature B-cell lymphoma arising from thymic B cells. PMBCL represents approximately 2 to 4% of non-Hodgkin lymphoma, with a predominance among females in the third and fourth decades [2].

In this study, we report a case of a previously healthy 16-year-old female who presented to our primary health care Center with a dry cough for 6 weeks associated with unintentional weight loss and night sweats. On examination, decreased breathing sounds were noted on the right side of the chest. A chest x ray showed right sided massive pleural effusion, so patients referred to the hospital as a case of pleural effusion for investigation.

Our case is unique because of the presentation of PMBCL with massive pleural effusion as the initial clinical manifestation for the disease, which is an uncommon initial presentation. Furthermore, the diagnosis at the age of 16 is highly unusual, as PMBCL typically occurs in individuals aged 30 -39 and often presents with compressive symptoms like SVC obstruction syndrome.

Keywords: B cell lymphoma; Cough; Pleural effusion; Non-Hodgkin lymphoma

Introduction

Primary mediastinal B-cell lymphoma (PMBCL) is a rare but aggressive mature B-cell lymphoma arising from thymic B cells. PMBCL represents approximately 2 to 4% of non-Hodgkin lymphoma, with a predominance among females in the third and fourth decades. Although previously thought to be a subtype of diffuse large B-cell lymphoma (DLBCL), it exhibits clinicopathologic characteristics that have led to its recognition as a distinct entity by the WHO in 2016 [1].

Clinically, they are characterized by rapidly increasing anterior mediastinal masses, which can cause compression of the surrounding tissues. The diagnosis of PMBCL mainly depends on the pathological features, imaging examination, and clinical features.

In this study, we report a case of a young female, 16 years old, previously healthy, who presented to our primary care health Center with a dry cough for 6 weeks associated with unintentional weight loss and night sweats. Physical examination showed decreased breath sounds in the right side of the lung. Chest x-ray done showed right side massive pleural effusion (Figure 1). Accordingly, the patient was referred to the Emergency department and admitted as a case of pleural effusion for further investigations. Pleural fluid analysis showed exudative effusion as per the light's criteria, and cytology showed Exudative Lymphocytic effusion. Moreover, a Tuberculosis workup was done, and it was negative. The patient was empirically treated as a case of community-acquired pneumonia with antibiotics, but her condition did not respond. Then, PAN CT showed a large mediastinal mass with significant mediastinal shift and total occlusion of the Superior vena cava with pulmonary artery compression and left internal jugular vein thrombosis. Mediastinal CT-guided biopsy showed large B-cell lymphoma. Currently, the patient receives treatment in NCCCR

(The National Center for Cancer Care and Research) under the haematology team.

Our case (PMBCL) is unique because it presented with massive pleural effusion as primary clinical manifestation for the disease which is a rare complication in addition to that our case is detected early at the age of 16 years old which unusual age group as most of these cases detected at the age between 30 to 39 with compressive symptoms like SVC obstruction syndrome. Furthermore, a chronic cough at such a young age can be a very subtle diagnosis that can easily be overlooked.

We shared this case due to its rarity. As mentioned in the literature, its annual incidence is 0.4 per million. Also, our patient was a 16-year-old female, which is a younger age group than the usual presentation of the disease. Our patient had the classical presentation of cough, which could have been missed early in the course of the disease and progressed to further complications, including central airway obstruction and respiratory failure, given the aggressive nature of this type of lymphoma.

The initial imaging done was a chest x-ray, which showed pleural effusion. The patient was then sent to the Emergency department for further investigation, including pleural tap and CT scan (Figure 2).

There is a wide range of differential diagnoses of the case that was considered, which included parapneumonic effusion, empyema, tuberculosis, heart failure, autoimmune causes such as lupus pleuritis, and malignancy (Figure 3).

Case Presentation

A 16-year-old female who is previously healthy presented to our primary health care center with a 6-week history of cough. It was productive of white/yellow sputum and associated with bloody strikes and sometimes vomiting after forceful coughing (post-tussive vomiting). She also reported unintentional weight loss (8 kg: 67 > 59 kg), decreased appetite, and night sweats, but no fever. No sick contacts. No known exposure to Tuberculosis. No animal exposure. No history of previous irradiation. **Review of systems**: She denied noticing any lumps. No abdominal or urinary symptoms. No skin rash. **Past medical history**: No history of chronic illness. Not on chronic medications. **Allergy**: No known allergies. **Family and social history**: She is a student who came to Qatar in 2016. She has 5 brothers and 4 sisters. No family history of malignancies or other chronic medical conditions. **Surgical History**: No previous surgeries. **Immunizations**: Up to date; last vaccines were 2 doses of the COVID vaccine in 2021.

On Examination: **Vital Signs**: Temp Axillary 36.9 C, Respiratory Rate 16 breaths/min, Blood Pressure 110/83, Oxygen Saturation 99%. **Heart**: JVP not raised, Normal heart sounds with no murmurs appreciated, but she was tachycardic. **Chest**: Percussion note dull with decreased right lung breathing sounds. **Abdomen**: Not distended, soft and non-tender, no hepatosplenomegaly. **CNS (brief)**: Alert, Glasgow coma scale 15/15, No focal neurological deficit. **Musculoskeletal / Spin**e: No joint swelling. No pedal oedema. No signs of DVT.

Investigations

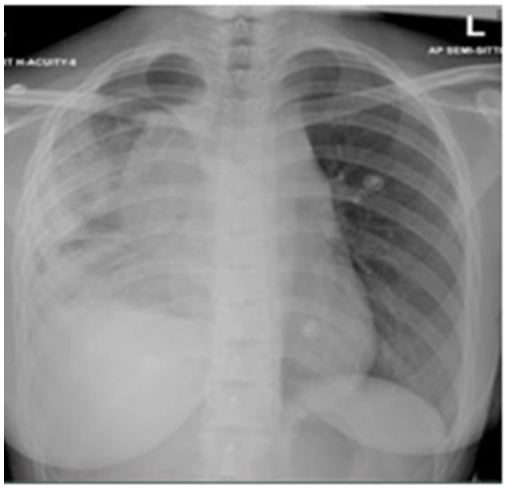

Figure 1: Chest x-ray: Widening of the mediastinum with predominant peripheral patch consolidation of the right lung.

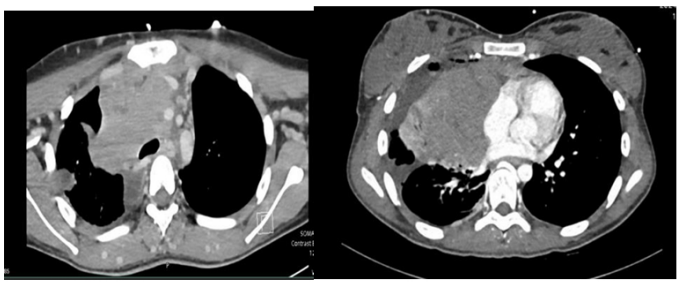

Figure 2: CT scan shows: Mediastinum mass. Centred on the right side and partially pushes and encases the great vessels in the upper mediastinum and the heart in the lower mediastinum.

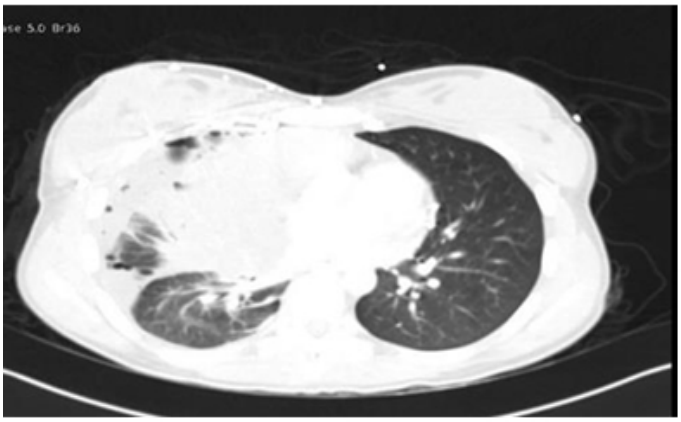

Figure 3: The right lung shows peripheral changes in the form of patchy consolidation and cystic changes with volume reduction.

Discussion

Primary mediastinal B-cell lymphoma (PMBCL) is a rare but aggressive mature B-cell lymphoma arising from thymic B cells. Although previously thought to be a subtype of diffuse large B-cell lymphoma (DLBCL), it exhibits clinicopathologic characteristics that have led to its recognition as a distinct entity by the WHO [2].

PMBCL represents approximately 2 to 4% of non-Hodgkin lymphoma, with a predominance among females. The annual incidence of PMBCL is 0.4 per million, with a 5-year survival rate exceeding 70%, with improving supportive care and genetic characterization of the disease [3].

The median age of diagnosis in females is 35 years [4].

The characteristic clinical manifestation of PMBCL is a fast-growing mass in the anterior mediastinum, which leads to local respiratory symptoms due to compression, including superior vena cava syndrome, pleural or pericardial effusion. Common symptoms include cough, dyspnoea, hoarseness, dysphagia, airway or vascular damage, and B symptoms (fever, night sweats, and weight loss). Lactate dehydrogenase (LDH) is also elevated. The involvement of distant lymph nodes and bone marrow is rare. Symptoms develop rapidly, usually within a few weeks of disease onset.

We shared this case due to its rarity. As mentioned in the literature, its annual incidence is 0.4 per million [5].

Also, our patient was a 16-year-old female, which is a younger age group than the usual presentation of the disease.

Our patient had the classical presentation of cough, which could have been missed early in the course of the disease, and progressed to further complications, including central airway obstruction and respiratory failure, given the aggressive nature of this type of lymphoma.

The initial imaging done was a chest x-ray, which showed pleural effusion. The patient was then sent to the Emergency department for further investigation, including pleural tap and CT scan. There is a wide range of differential diagnoses of the case that was considered, which included parapneumonic effusion, empyema, tuberculosis, heart failure, autoimmune causes such as lupus pleuritis, and malignancy.

Following Pleural tap and imaging, the diagnosis was confirmed by mediastinal biopsy guided by CT scan. Staging was done, and the patient was started on the Dose Adjusted -EPOCH-R chemotherapy regimen, which includes the following drugs: etoposide phosphate, prednisone, vincristine sulphate (Oncovin), cyclophosphamide, doxorubicin hydrochloride (hydroxydaunorubicin), and rituximab.[5]

Declarations

Contributors: All authors contributed to planning, literature review, and conduct of the review article. All authors have reviewed and agreed on the final manuscript.

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