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Case Study

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A PRIMARY THYMIC NON HODGKIN LYMPHOMA: A RARE MEDIASTINAL MASS

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ABSTRACT

Anterior mediastinal lymphomas may be part of systemic lymphoma and they comprise approximately 50% of mediastinal neoplasms. Primary non-Hodgkin's lymphoma of the mediastinum is a subtype with lower prevalence, representing 10% of all cases; it has two histologic subtypes that include T-cell lymphoblastic lymphoma and diffuse large B-cell lymphoma. The latter is an aggressive neoplasm that tends to infiltrate the pleura, bone structures and the lung; it originates from thymic medullary B cells. HEREBY, WE PRESENT presents a case of primary non-Hodgkin's lymphoma of the mediastinum with diffuse large B-cell subtype, its radiologic findings, pathology and a brief review of the literature. Computed tomography scan of the thorax revealed a mass in the anterior mediastinum. The suggested diagnosis was thymoma. The definitive histologic study revealed a primary thymic B-cell lymphoma.

KEYWORDS: Diffuse large B-cell lymphoma, thymus, thymoma.

INTRODUCTION

The thymus is a primary lymphoid organ which establishes and maintains the pool of effector T-cells throughout the life. However, it also plays an important role in the development and maturation of B-cells. The antigen-independent B-cell development occurs in a primary lymphoid organ, the thymus, while the antigen-dependent stages in secondary lymphoid organs such as spleen, Peyer's patches, and lymph nodes.[1]

Anterior mediastinal lymphoma type lesions generally make part of systemic lymphoma; however, a subtype with lower prevalence is the primary non-Hodgkin's thymus lymphoma. This last one is a low frequency entity, that is characterized by an aggressive behaviour with tendency for local invasion. The two histological subtypes of primary non-Hodgkin's mediastinal lymphoma are T-cell lymphoblastic lymphoma and diffuse large B-cell lymphoma[1]. DLBCL represents only 6-10% of non-Hodgkin's lymphomas. Only 19 cases of thymic large B‑cell lymphoma have been reported worldwide; to the best of our knowledge, most of them are solitary cases, the latest case reported from Mexico in 2013.[2]

Next we describe the case of a 24 year old patient with primary non- Hodgkin's lymphoma of the mediastinum with diffuse large B-cell subtype and its findings by images.

CASE REPORT

A 24 years old female, who consults emergency services given a clinical picture of 4 months of fever,[3] months of shortness of breath and swelling of neck and face. On general examination, the patient was of average built, poor nutrition and stable vitals. No Cervical, axillary or inguinal lymphadenopathy. On examination of the respiratory system, the patient was found to have decreased chest movement, decreased vocal fremitus, decreased vocal resonance, dull note on percussion, and decreased breath sounds on the right side infraclavicular. mammary, axillary and interscapular areas. Cardiovascular, neurological, and gastrointestinal systems were unremarkable.

In the thorax radiography (figure 1) a mediastinal engrossing was identified, for which a computerized tomography (CT) of the thorax (figure 2) was performed, which showed a well circumscribed heterogenous solid mildly enhancing mass lesion in the anterior mediastinum ? thymoma. with infiltration to the thoracic wall and with compressive effect over the superior cava and trachea.

Routine laboratory investigations revealed mild anemia, raised erythrocyte sedimentation rate, decreased leukocyte count, raised lactate dehydrogenase (LDH), normal liver enzymes (alanine transaminase/aspartate transaminase), and alkaline phosphatase levels with normal kidney function test, blood sugar, and thyroid profile.

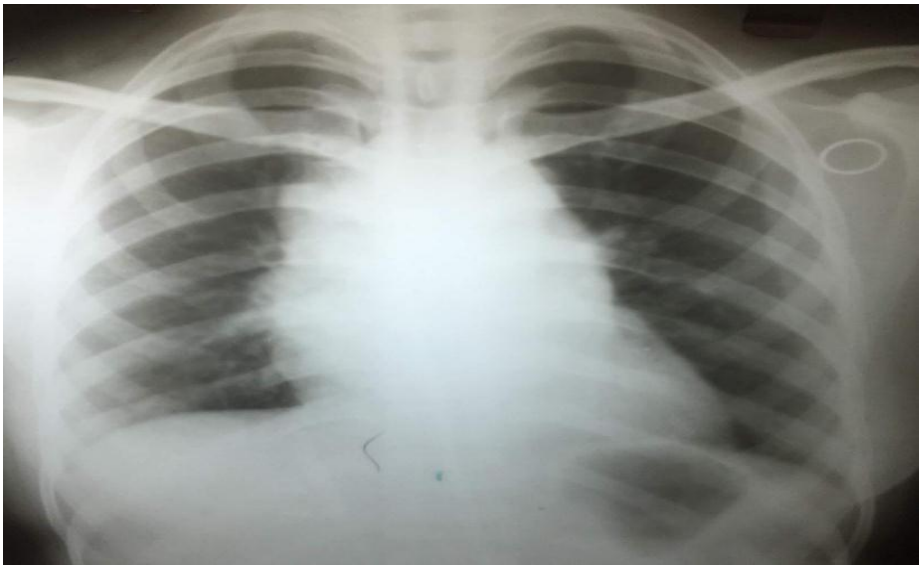


Figure 1: Mediastinal widening due to anterior mediastinal mass.

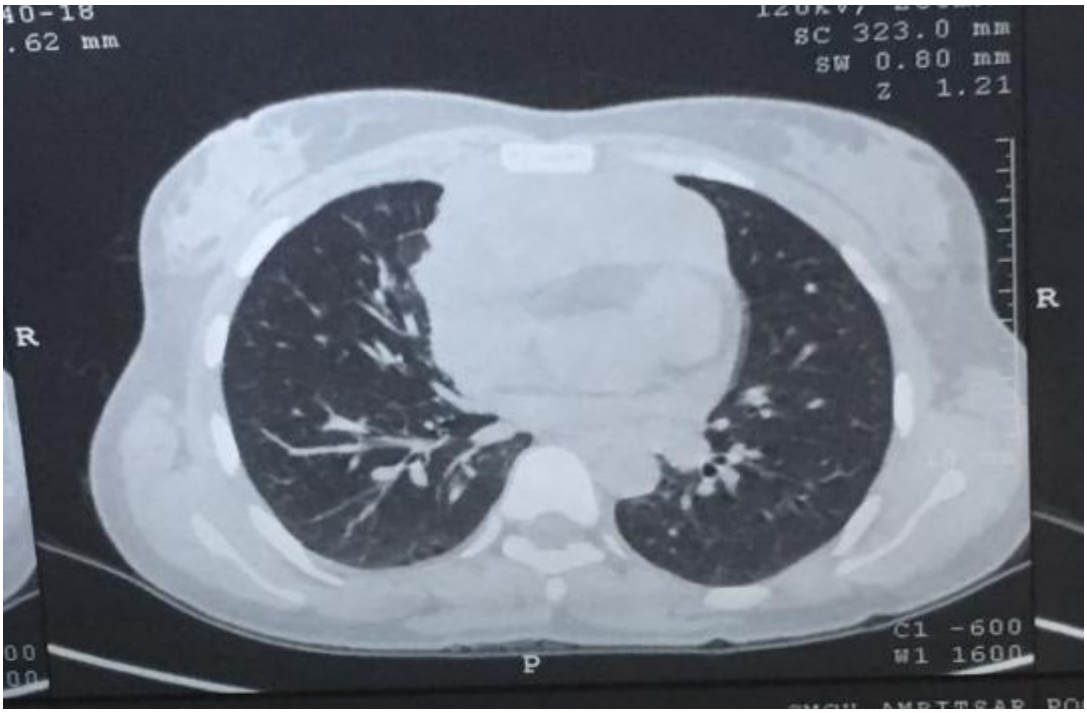
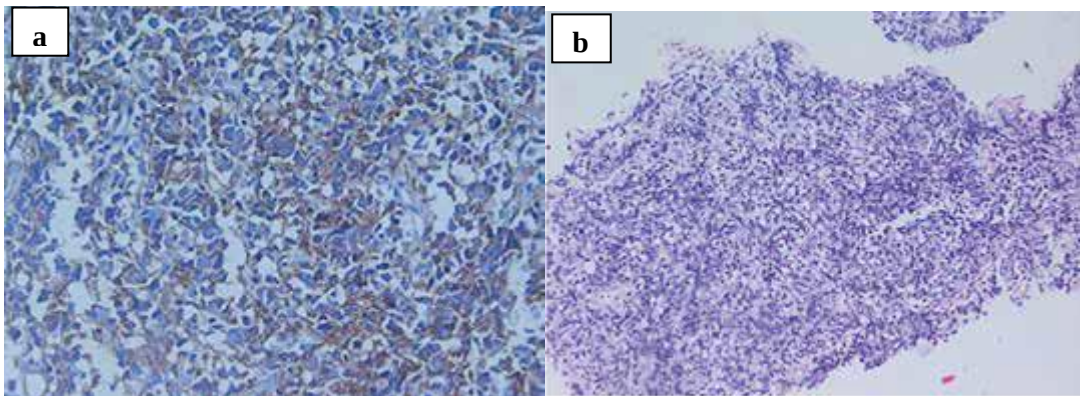


Figure 2: Mass with soft tissue density and scarce enhancement of the contrast medium, localized in the anterior mediastinum.



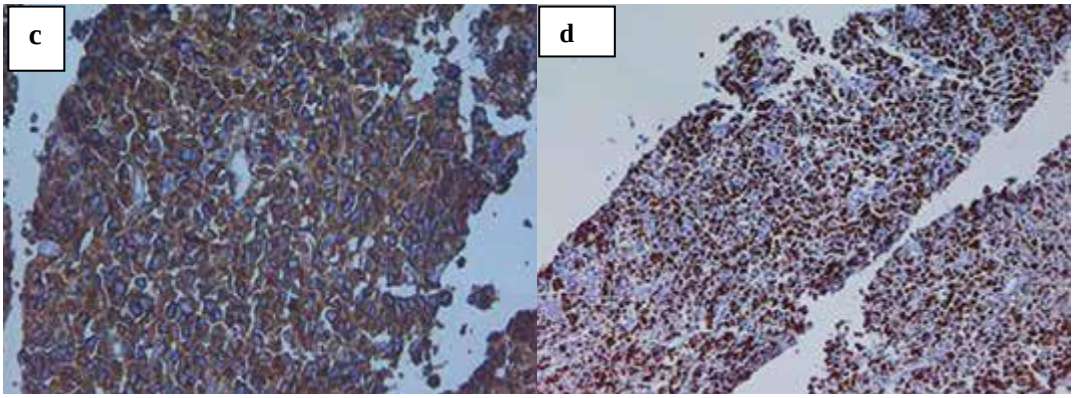


Figure 3: a). Eosin hematotoxyline coloration. b) Cellular immunoreactivity (membranous and cytoplasmatic) to CD20. c) Cellular immunoreactivity (membranous and cytoplasmatic) to CD10. d) Cellular proliferation index marked with KI67.

A USG guided FNAC from mediastinal mass show cytological smears prepared show few lymphoid cells along with few histiocytes. Possibility of lymphoblastic lymphoma to be ruled out. A USG guided trucut biopsy was performed which is provisionally suggestive of high grade B cell NHL. In the immunohistochemistry studies reactivity of the tumour cells was identified to CD20 (B lymphocyte marker) (figure 3b), CD 99 and show patchy positivity for CD30 (T lymphocyte and activated B lymphocyte marker), Bcl 2 positive in T cells, and negativity for c-myc and Tdt.

The patient was initiated on the R‑CHOP chemotherapy and is currently doing well. she has completed three cycles of chemotherapy.

DISCUSSION

Primary mediastinal diffuse large B‑cell lymphoma was first described as an entity in the early 1980s. Before then – and even since then – it has often been misdiagnosed as a thymoma or a germ cell tumor. With the advent of immunophenotyping studies, the tumor was identified as a large B‑cell lymphoma. Such tumors typically are composed of cells that resemble centroblasts (large germinal center cells); rare cases may have larger cells with abundant cytoplasm and prominent nucleoli, resembling RS cells and giving rise to a differential diagnosis with Hodgkin's lymphoma.

Tumours of the anterior mediastinum represent 50% of all mediastinal lesions; they include thymoma, teratoma, thyroid and lymphoma disease. Lymphoma represents 20% of all mediastinal neoplasias in adults and 50% in children.[4] Mediastinal infiltration due to lymphoma occurs as a systemic dissemination of the disease in the majority of patients, and in a less frequent form, as a primary tumour in upto 10% of the cases.[5] Among the group of primary mediastinal lymphomas a 15 to 25% are of the non-Hodgkin's type (NHL). Less than 3% of all NHL cases correspond to a rare subtype of lymphoma known as primary diffuse large B cell mediastinal lymphoma or thymic lymphoma.[6] The two main forms of mediastinal NHL are the T lymphoblastic lymphoma and diffuse large B cell lymphoma. The primary mediastinal diffuse large B cell lymphoma or thymic lymphoma is a neoplasm of aggressive behaviour whose origin are thymic B cells localized in the medulla of this organ. The age of diagnosis is between the third and fourth decade of life and there is predominancein the female sex.[4,7] The clinical manifestations at the momentof appearance include coughing, thoracic pain and dyspnea. It is frequent the appearance of compressive symptoms when the airway and vascular structures are invaded.[8] This tumour is usually present in the CT as a large mediastinal mass, reaching in 75% of patients more than 10 cms in diameter. With frequency it infiltrates the lungs, the thoracic wall, the pleura and the pericardium; besides, a third of patients present pleural or pericardial leakage.[8] Its borders can be smooth or lobuled and in half the cases its aspect is heterogeneous with low signal areas due to the presence of necrosis, haemorrhage or cystic degeneration.

CONCLUSION

The lymphoma is part of the neoplasias that most frequently compromise the anterior mediastinum, in the majority of cases being a Hodgkin lymphoma (50-70%). The primary mediastinal diffuse large B cell lymphoma is a low frequency entity with an atypical behavior given the tendency for local invasion and secondary invasion to the pleura, the pericardium, the thoracic wall and lung. The survival in these patients is lower when compared with other subtypes of lymphoma and its recurrence is present with higher frequency in the thorax, in the first two years of follow-up.

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