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Case report

Superior vena cava syndrome caused by mediastinal lymphoma: A rare clinical case ☆,☆☆

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ABSTRACT

The superior vena cava syndrome refers to a medical emergency resulting from superior vena cava compression, which develops in 2%-4% of non-Hodgkin lymphomas. Primary mediastinal (thymic) large B-cell lymphoma is an unusual and aggressive non-Hodgkin lymphoma that can present with superior vena cava syndrome.

We report the case of a Caucasian 21-year-old female, who presented with acute superior vena cava syndrome, having started 2 weeks before. Chest computerized tomography disclosed an anterior mediastinal mass (18 × 14 cm). Thoraco-abdominopelvic magnetic resonance imaging was performed in order to differentiate compression versus mass invasion. A gross anterior mediastinal mass (109 × 60 × 105 mm) occupying the totality of the prevascular space was found, extending from the sternal furcula to the pericardium, fully embedding the superior vena cava. A computerized tomography guided biopsy was performed. Histopathological and immunohistochemical analysis was consistent with Primary mediastinal (thymic) large B-cell lymphoma.

Primary mediastinal (thymic) large B-cell lymphoma has unique clinicopathologic aspects and it should be considered in a young patient with Superior vena cava syndrome. Prompt recognition, a timely diagnosis and appropriate treatment are crucial for prognosis.

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Introduction

The superior vena cava syndrome (SVCS) is a collection of clinical signs and symptoms occurring when the superior vena cava is partly or completely blocked. This entity is reported to develop in only 2% to 4% of non-Hodgkin lymphoma (NHL). Timely and effective intervention aimed at treating the malignant cause of SVCS is crucial to improve the outcome [1].

On the other hand, Primary mediastinal (thymic) large B-cell lymphoma (PMLBL)is a distinctive subtype of NHL, with specific clinicopathologic aspects and an aggressive behavior. As per literature, very few cases are described with SVCS at initial presentation [2].

We report a case of a patient who presented with acute SVCS caused by PMLBL. We also suggest a practical diagnostic and therapeutic algorithm to SVCS with an anterior mediastinal mass.

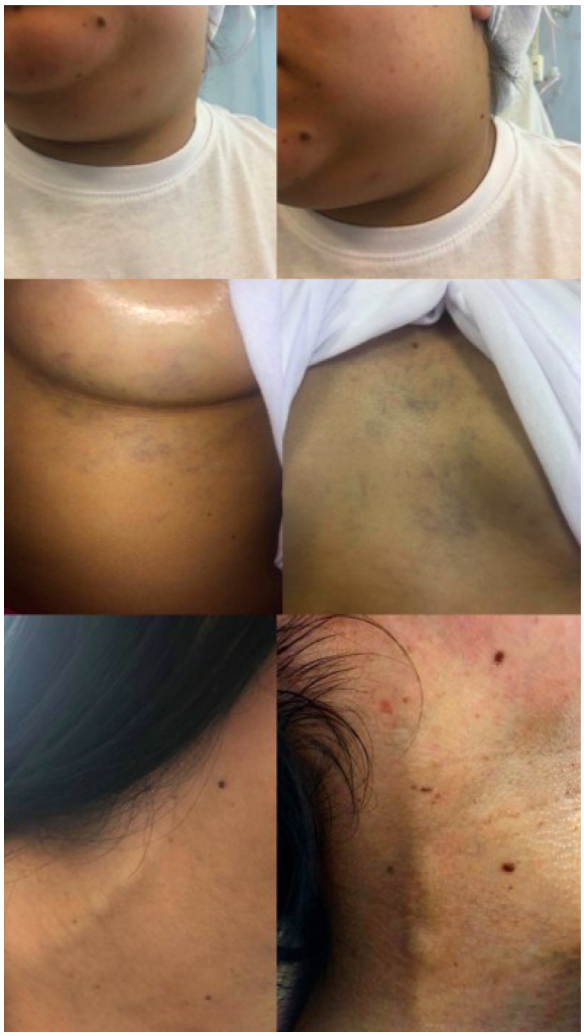

Fig. 1 – From top to bottom: Significant swelling of the face and neck. Superficial veins of the chest and left inframammary region. Jugular venous distention and ecstatic superficial veins over the chest.

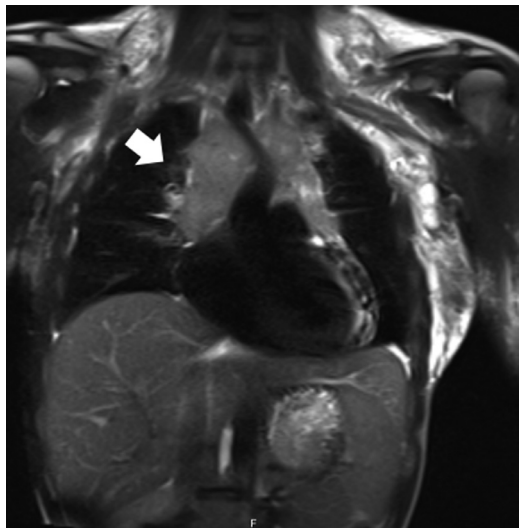

Fig. 2 – A gross mediastinal mass fully embedding the SVC (coronal T2 MRI).

Case report

We report the case of a Caucasian immunocompetent 21-year-old female with insignificant past medical history, who presented to the Emergency Department with complaints of dry cough along with swelling of the face, neck, and upper limbs for 2 weeks, worsening in dorsal decubitus position. The patient also noticed dyspnea, which intensified with orthostatic position. Physical examination revealed significant swelling of the face, neck and arms, jugular venous distention, and ecstatic superficial veins over the chest. There was no lymphadenopathy and the systemic examination was unremarkable. She had no similar prior episodes and no relevant family history was reported.

Laboratory test results revealed lymphopenia (790/mm^3), mild C-reactive protein elevation of 10.7 mg/L and a lactate dehydrogenase of 421 IU/L. A chest computerized tomography (CT) disclosed an anterior mediastinal mass which suggested compression of the superior vena cava , small left pleural effusion and small pericardial effusion.

On admission in our department, physical findings did not differ from the ones described above (Fig. 1). Further laboratory tests exhibited mild erythrocyte sedimentation rate elevation - (27 mm/1h) and a normal coagulation study. Beta-human chorionic gonadotropin (beta-hCG), alfa fetoprotein (AFP), anti-acetylcholine receptor antibodies and serologic markers for human immunodeficiency virus, hepatitis B and C virus were negative.

Thoraco-abdominopelvic magnetic resonance imaging was performed in order to differentiate compression versus mass invasion. A gross anterior mediastinal mass (109 × 60 × 105 mm) occupying the totality of the prevascular space was found, extending from the sternal furculato pericardium, fully embedding the SVC (Fig. 2,3,4).

A CT guided biopsy was performed. Histopathological analysis showed the presence of numerous medium-sized to large

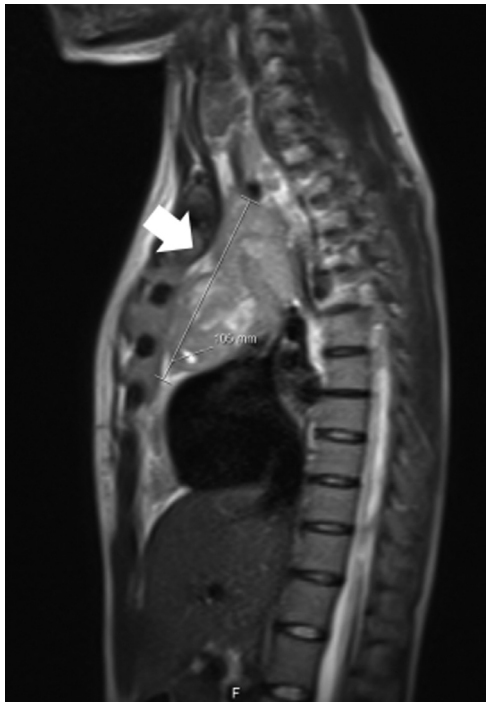

Fig. 3 – A gross mediastinal mass fully embedding the SVC (sagittal T2 MRI).

cells in a fibrous stroma. The neoplastic cells showed abundant pale cytoplasm and slightly irregular nuclei (Fig. 5). By immunohistochemistry, neoplastic cells were immunoreactive for CD20 (Fig. 6). No CD30 and CD15 expression was observed. The findings were consistent with the diagnosis of PMLBL.

Positron emission tomography revealed exclusively supradiaphragmatic ganglionic involvement.

Initial medical management consisted of pain relief medication andcorticotherapy (methylprednisone 200mg during 6 days) with quick symptomatic relieve and clinical improvement (Fig. 7).

Upon completion of the first chemotherapy cycle (dose-adjusted etoposide, prednisone, vincristine, cyclophosphamide, doxorubicin, and rituximab- DA-EPOCH-R), the patient was asymptomatic with hematologic recovery. A core biopsy showed no bone marrow involvement. The patient is currently going through a third chemotherapy cycle with a good clinical response and no complications reported.

Discussion

PMBCL is a rare entity of NHL, representing approximately 2% to 4% of all NHL and 6% of diffuse large B-cell lymphomas. It affects mainly female young adults (median age of 35) [3,4], and presents as a large, fast-growing invasive tumor in mediastinum [5].

On the other hand, the main causes of SVCS includes malignant conditions (60%–85%). Nonmalignant conditions are especially due to SVC thrombosis caused by intravascular devices and catheters [6]. Non-small-cell lung cancer is the most common malignant cause of SVCS, followed by small-cell lung cancer (50% and 25%, respectively), with lymphomas accounting for about 10% of cases [7].

In a patient presenting with malignancy-related SVCS, a detailed clinical history and physical examination, combined with CT imaging, are crucial to define the urgency of medical intervention and, to establish a definitive diagnosis [8].

In what concerns imaging features, in contrast to other malignant conditions (such as thymoma or classic Hodgkin lymphoma), PMBCL infiltrates mediastinal structures and middle mediastinal adenopathy is not a feature [4].

When a mediastinal (anterior) mass is identified a definitive diagnosis and treatment of the underlying condition will depend on microscopic evaluation. Specially in the case of lymphoma, the identification of its molecular and genomic features is critical to confirm its subtype [4,5].

Diagnosis is based on histopathological and immunohistochemical analysis, which defines not only the type of treatment that might be suitable but also the success of treatment monitoring [3]. Excisional biopsy is the optimal method for the initial diagnosis [8].

Moreover, it is not uncommon that PMBCL cells express CD30, a marker frequently seen in classic Hodgkin lymphoma [5]. In fact, PMBCL shares features with nodular sclerosis Hodgkin lymphoma, such as genetic patterns, immunological properties and a putative thymic B-cell origin. However, PMBCL cells are only rarely positive for CD15 [4].

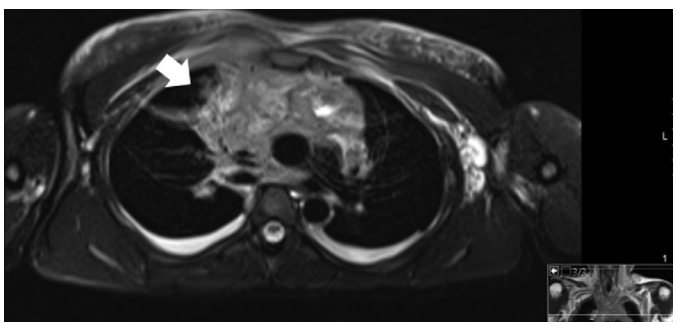

Fig. 4 – A gross mediastinal mass fully embedding the SVC (transversal T2 MRI).

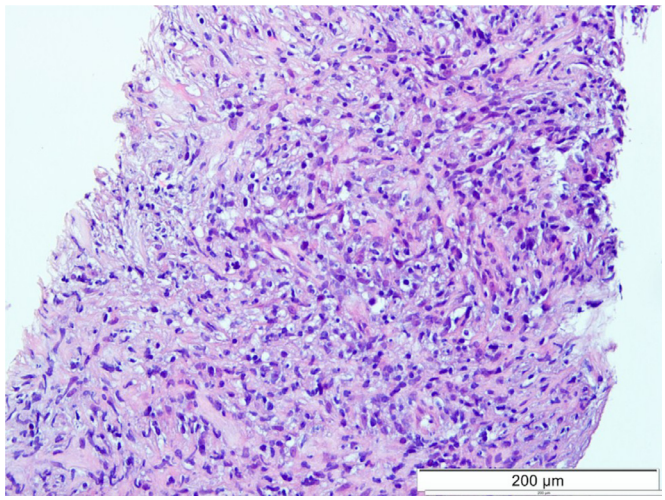

Fig. 5 – Abundant pale cytoplasm and slightly irregular nuclei.

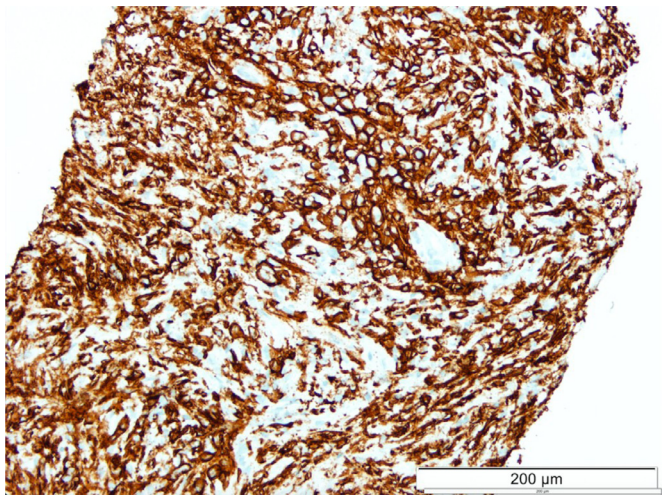

Fig. 6 – On immunohistochemistry, neoplastic cells were immunoreactive for CD20.

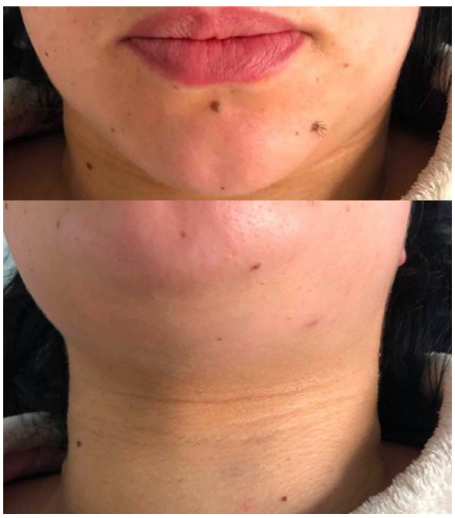

Fig. 7 – The patientś face and neck improved dramatically after methylprednisone.

As the diagnosis of PMBCL can only be established through microscopic evaluation, it is essential that pathologists are aware of and familiar with this tumor given its rarity [4].

As in this case, the positivity for CD20 makes the patient eligible for treatment with rituximab, which is effective in all B cell lymphomas due to a more avid CD20 expression.

Conclusion

This paper is significant not only because it describes a rare cause of SVCS but also gives more information about a rare reported diagnosis in the literature.

We intended to highlight the importance of differential diagnosis in young patients presenting with SVCS and the inclusion of PMBCL. It is crucial that clinicians quickly recognize and diagnose this condition so that treatment can be started, thereby improving the outcome.

Hence, the dissemination of the information gathered from this case and other similar occurrences of young patients with PMBCL will contribute to anticipate further testing and knowledge in order to attain better results.

Patient consent statement

Unfortunately, due to COVID-19 pandemic, it was not possible to obtain consent statement. However all efforts were made to preserve the identity of our patient.

Statement of ethics

The authors declare that they have no conflicts of interest.

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