Superior vena cava syndrome in primary mediastinal B-cell lymphoma with sclerosis

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Abstract

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

We present a case of a caucasian immunocompetent 21-year-old female who presented to the Emergency Department with acute onset SVCS (cough and swelling of the face, neck and upper limbs) for two weeks. Laboratory results were normal. A chest radiograph revealed marked widening of the mediastinum. Chest computerized tomography (CT) disclosed an anterior mediastinal mass (18 x 14 cm). She was admitted in Internal Medicine Department for etiological study. Thoraco-abdominopelvic magnetic ressonance was performed in order to differentiate compression versus mass invasion. A gross anterior mediastinal mass (109x60x105 mm) occupying the totality of the prevascular space was found, extending from the sternal furcula to pericardium, totally embedding the superior vena cava.

A CT guided biopsy was done and histopathological examination of the mass revealed PMLBL (CD20 histochemical positivity). Initial medical management consisted of pain relief medication and corticotherapy with quick symptomatic relieve. Currently, the patient is undergoing chemotherapy (dose-adjusted etoposide, prednisone, vincristine, cyclophosphamid, doxorubicin, and rituximab - DA-EPOCH-R).

PMLBL predominantly affects women in the third decade of life. This entity has unique clinicopathologic aspects and an aggressive behavior. It should be considered as one of the differentials in a young patient with SVCS. Prompt recognition, a timely diagnosis, management and appropriate treatment may be crucial for prognosis.

Biography

Bruno Besteiro has completed his PhD at the age of 25 years from Faculdade de Medicina da Universidade do Porto (FMUP). He is an Internal Medicine resident of the Internal Medicine Department in Centro Hospitalar e Universitário de São João, Oporto, Portugal. He has also teaching-activities as assistant of Medicine at FMUP. He has published and presented more than 5 papers /cases in reputed journals and national and international congresses.

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