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Extra-cardiac compression and pericardial effusion secondary to primary mediastinal large B cell lymphoma

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PPrimary mediastinal large B cell lymphoma (PMLB) accounting for 7% of diffuse large B cell lymphomas and represent 2.4% of all non-Hodgkin lymphomas. Median age at diagnosis in the third to fourth decade. In some cases the initial presentation can be an oncological emergency with locally invasive anterior mediastinal mass arising from thymus causing external pressure on the heart and the surrounding structures. Further pressure on the trachea and the lungs can also compromise the airway or lead to superior vena cava (SVC) syndrome.

Background: PMLBL is classified as type of aggressive NHL which originating from the mediastinal thymic (medullary) B-cells, remarkably PMLBL has a discrete clinical features as well as categorically sharing some clinical and pathological features with other types of B cell lymphoma. From the most recent published studies, It was found that PMLBL has more benign outcome and excellent response to chemotherapy especially if dealt with at earlier stage.

Case presentation: A 27 years old male who admitted with pleuritic chest pain, shortness of breath and palpitations, no remarkable past medical history, Clinically he was tachycardic of heart rate 117 bpm, BP 106/60, Temp 37.4 oxygen saturation of 100% on air, No other abnormality detected on other systemic examination. The initial investigation including, FBC, renal function, LFT, TFT, clotting profile, CRP and cardiac troponin were all within the normal parameters, negative virology screening tests. ECG showed sinus tachycardia, CXR revealed widened mediastinum. Chest abdo-pelvis CT scan confirmed the presence of a large anterior mediastinal mass of 9.6cm with moderate pericardial effusion. The mass infiltrates into the mediastinum and encasing the aorta, which is causing extrinsic compression on the mediastinal structures with appearances suggestive of right sided heart strain. There was 12 mm left hilar lymph node with no evidence of pleural effusion or pleural-based disease. Echocardiography has shown extra cardiac mass anteriorly size 2.6 cm, pericardial effusion posteriorly 0.8 cm and laterally 1.3 cm, persevered LV systolic function and late systolic septal flattening, without evidence of cardiac tamponade. The patient was transferred to the local tertiary centre for pericardial effusion drainage and tissue biopsy that has confirmed the diagnosis of primary mediastinal diffuse large B Cell Lymphoma, as a result, the patient subsequently started on chemotherapy treatment (R-CHOP) under the care of haematology with excellent an outcome. Few weeks later, patient was readmitted with neutropenic sepsis and large left pleural effusion as consequences of chemotherapy treatment for which he was treated aggressively with intravenous antibiotics and made a good recovery.

Discussion: PMLBL is vigorous tumour arising in the mediastinum from the thymic (medullary) B cell. It has unique clinical and pathophysiological features In contrast with systemic diffuse large B cell lymphoma, however identifying the gray-zone lymphoma is of paramount importance as highlighted by the WHO for diagnostic and therapeutic purposes. Symptoms of cough, dyspnoea, hoarseness of voice, and dysphagia can be presentative feature of SVC syndrome and local invasion of the tumour. An adequate biopsy specimen with using immunophenotyping is the key to the diagnosis, There is disparity in clinical practice for treating PMLBL, the treatment options hugely depend on the patient and tumour characteristics. In this case the possibility of clinical SVC syndrome was excluded based on the facts that there were no characteristic signs of central venous obstruction, facial swelling or head fullness, although reported that the CT scan has shown a degree of compression on the mediastinal structures. Others potential emergencies that described in the literatures that some patients have presented with have included, cardiac tamponade, thrombosis of major neck or superior thoracic veins also hyperuricemia and tumor lysis syndrome. Patient with large mediastinal mass they may present also with cardiac or respiratory arrest at initial or during anaesthesia.

Biography

Amjed Eljaili completed MBBS on October 2010 from University of Al-Zaiem Al-Azhari, Sudan, currently practicing in UK, Wales Deanery, foundation year-2 trainee, BCUHB, emergency department. He attended several academic meetings, regionally and nationally, He has participated in various national work-shops, congress participation and membership with British Institute of Radiology, UK.

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