A case of Castleman disease with pulmonary hypertension

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Introduction

Castleman disease, also known as Giant lymph node hyperplasia or Angiofollicular lymph node hyperplasia, is a disease of lymph nodes and related tissues. It is not considered as a malignancy but one variety of it acts like lymphoma.

Case report

A 52 year old male was admitted to Teaching Hospital Karapitiya with loss of weight for 1 year, abdominal distension for 6 months and loss of appetite for 1 month. There was no history of fever. He had stable angina and was on anti-anginals, aspirin and verapamil.

On examination he was emaciated, pale and had large, discrete, firm and non-tender lymphadenopathy in the cervical, axillary and inguinal regions. Clinically he had cardiomegaly, a grade 4 pansystolic murmur best heard in the lower sternum and radiating to the axilla and a loud pulmonary second heart sound. Respiratory system was normal except for few bilateral fine basal crepitations. Abdominal examination showed a 5 cm hepatomegaly and splenomegaly.

His haemoglobin was 9.9 g/dL while white cell and platelet counts were normal. Blood picture showed hypochromic red cells while bone marrow examination showed normal active marrow. ESR was elevated at 80 mm/1st hour. Liver enzymes were normal and Mantoux test was negative. Ultrasound scan of the abdomen showed hepatomegaly with normal echo texture and no focal lesions and splenomegaly. There was no intra-abdominal lymphadenopathy or ascites. ECG showed right axis deviation and evidence of right atrial enlargement. Chest radiograph showed gross cardiomegaly and oligaemic lung fields. 2D Echo found severe tricuspid regurgitation and dilated right ventricle and atrium indicative of severe pulmonary hypertension. There was also a small pericardial effusion. Lung function tests were compatible with restrictive lung disease. Biopsy from an axillary lymph node showed features suggestive of Castleman disease. HIV antibody was negative.

Discussion

Castleman disease usually presents with fever, weight loss, fatigue, night sweats, infections and anaemia. Pulmonary hypertension has been rarely reported in association with multicentric Castleman disease. Only three such reported cases were found during the literature search. Of these, two had tested negative for HIV while the other had tested positive. The proposed mechanism for pulmonary hypertension has been promotion of angiogenesis by Interleukin-6 produced in the germinal centres of hyperplastic lymph nodes.

Castleman disease may present in a localized form or a multicentric form. The localized form only affects a single lymph node group, most often in the chest and abdomen. The multicentric form affects more than one group of lymph nodes and also other organs containing

lymphoid tissue. The multicentric form sometimes occur in HIV positive patients.

Histologically it is divided into a Hyaline vascular type and a Plasma cell type. The Hyaline vascular type is usually clinically localized while the Plasma cell type is usually multicentric.

Treatment decisions depend on the clinical subtypes and not on microscopic subtypes. Localized Castleman disease is treated with surgery and external beam radiation for cases not amenable to surgery. Chemotherapy either alone or in combination with radiotherapy or steroids is used in multicentric Castleman disease.

Monoclonal antibodies like rituximab or tocilizumab can neutralize the targets for IL-6 on cell surfaces.

References

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