

Constrictive pericarditis as the first presentation of untreated Angioimmunoblastic Peripheral T-cell Lymphoma in a young patient

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DOI: 10.5530/jcdr.2015.4.4

Primary and secondary cardiac lymphoma mainly involves pericardium as pericardial effusion or cardiac tamponade, and when it does so, already the disease is in an advanced stage with many other viscera already affected.¹ Lymphoma leading to cardiac constriction is very rare, and the manifestations of constriction are predominantly subclinical or insidious or autopsy diagnosed. Hence a symptomatic clinical presentation of this rare sequel, like in our case, needs high index of suspicion for an underlying malignant disease.² Our case is further distinct with regards to the subtype of lymphoma leading to the constriction, the age of presentation of the patient, the clinical course of the disease, and the distribution of the organs involved.

The patient was a 37 year old gentleman, who developed gradually progressive distention of abdomen and swelling of both lower limbs over 3 months associated with breathlessness. He additionally noticed a painless, progressively enlarging swelling in the right groin for 3 months. His clinical examination revealed raised jugular venous pulse, with a positive Kussmaul's sign. There were enlarged lymph nodes in the right groin (firm in consistency, the largest measuring 10 cms) and left supraclavicular region (size of 2 cm). The precordial examination revealed an inconspicuous apical impulse with a pericardial knock. His lab investigation showed Hemoglobin of 10.2 gram%, Total leukocyte count of 8500/cm³, mainly polymorphs, normal liver and renal function tests. X ray chest and ultrasound abdomen had features of pleural effusion and ascites with dilated inferior vena cava. His Echocardiography showed the typical features of constriction like pericardial thickening with inspiratory augmentation of tricuspid inflow and augmented hepatic flow reversals (Figure 1C). The finding of constrictive physiology was confirmed on right heart catheterization, which showed raised, and equalization of diastolic pressures with a 'Dip and plateau sign' in the ventricular tracing (Figure 1-a,b). CECT [Contrast Enhanced Computerized Tomography] thorax and abdomen revealed multiple lymph nodes in the superior mediastinum, peripancreatic and pre-aortic location. FNAC [Fine needle Aspiration cytology] from the inguinal lymph node had features of immature round cells suggesting Non-Hodgkin's lymphoma. Excision biopsy from the left supraclavicular lymph node confirmed Non Hodgkin's lymphoma. Further Immunohistochemistry revealed positive CD 2,3,4,5,7,8, 30 & PD1⁺ markers (Figure 1a,b) which characterized it to be Angioimmunoblastic type of Peripheral T cell lymphoma. Patient has now been referred to the Oncology Department and undergoing workup for a planning of chemotherapy.

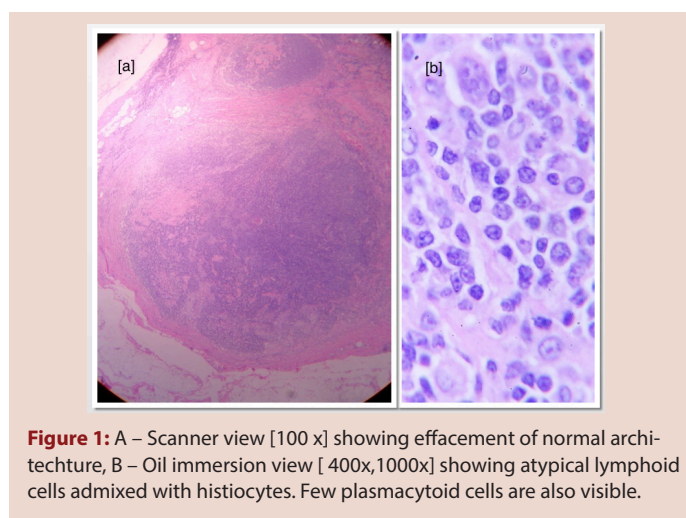


Figure 1: A – Scanner view [100 x] showing effacement of normal architecture, B – Oil immersion view [400x,1000x] showing atypical lymphoid cells admixed with histiocytes. Few plasmacytoid cells are also visible.

DISCUSSION

Our case demonstrates an uncommon or probably an inverse instance of a patient who, while being worked up for constrictive pericarditis, was found to have an undiagnosed, advanced stage Non Hodgkin's lymphoma. Constrictive pericarditis reported with lymphoma has mainly been attributed to irradiation, and have rarely had an overt clinical manifestation.^{2,3} This probably points towards a milder degree of constriction process in the usual lymphoma cases reported so far, as opposed to an aggressive symptomatic disease process in our case. Apart from the general rarity of this complication, till date this is of constrictive pericarditis reported in the literature among this subtype of Angioimmunoblastic peripheral T cell lymphoma; a rare aggressive subtype of lymphoma predominantly found in elderly patients.

Learning Points: Though rare, this subtype (Angioimmunoblastic peripheral T cell) of Non Hodgkin's Lymphoma can present in young individuals. Involvement of the heart at a relatively early stage can lead to symptomatic constrictive pericarditis, with sparing of other viscera usually involved by Non Hodgkin's lymphoma (i.e. liver, lung and spleen). We need to have strong vigilance in cases of constriction to find out the underlying etiology, sometimes completely curable.

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