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Waldenstroms Macroglobulinemia with Amyloid Cardiomyopathy - A Case Report

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Abstract: ***Waldenstroms macroglobulinemia is a malignant disease of B cells that secrete IgM paraprotein similar to multiple myeloma and non - hodgkins lymphoma. WM cells have features of both plasma cells and lymphocytes and are called lymphoplasmocytoid. Secretion of the IgM paraprotein leads to hyperviscosity and vascular complications and is associated with MYD88 mutation. This extremely rare neoplasm has an annual incidence of 4 cases per million people contributing approximately 2% of all hematologic malignancies. The diagnosis is usually challenging due to a lack of specific morphologic, immunophenotypic or chromosomal changes.***

Keywords: Waldenstroms macroglobulinemia, IgM paraprotein, hyperviscosity, MYD88 mutation, hematologic malignancies

1. Case

A 55 - year - old male was admitted to our hospital with exertional dyspnea for 3 months followed by recurrent headache for 3 weeks. During the course in the hospital, he had swelling of both legs on prolonged standing. General examination revealed pallor and elevated Jugular venous pressure with dependant pedal edema. Vitals revealed a postural fall in Blood pressure.

2. Observation

Routine blood investigations were within normal limits except for anemia and high ESR (100mm/hr). additional investigations like s. calcium and renal function tests within normal limits. skeletal survey doesn't reveal osteolytic lesions. ECG showed **no** chamber hypertrophy and Echo suggestive of LV dysfunction, LVH, and Global hypokinesia. Further investigations showed Plasmacytoid lymphocytes in peripheral blood smear and bonemarrow biopsy. Serum protein electrophoresis revealed IgM monoclonal gammopathy. On immunohistochemistry, CD20 and CD 138 positive. During the course in the hospital, she was counselled and treated with plasmapharesis thus showing clinical improvement on follow - up.

3. Clinical Significance

Physicians should have a high index of suspicion for Waldenstroms macroglobulinemia while evaluating patient with features of cardiac failure in the setting of recurrent headache. Secretion of the IgM paraprotein leads to **hyperviscosity** and cardiac failure is due to deposition of amyloid which leads to Amyloid cardiomyopathy.

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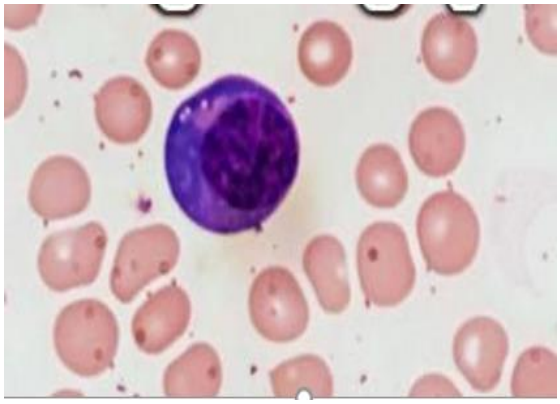

Figure 1: Plamacytoid lymphocyte in peripheral blood smear

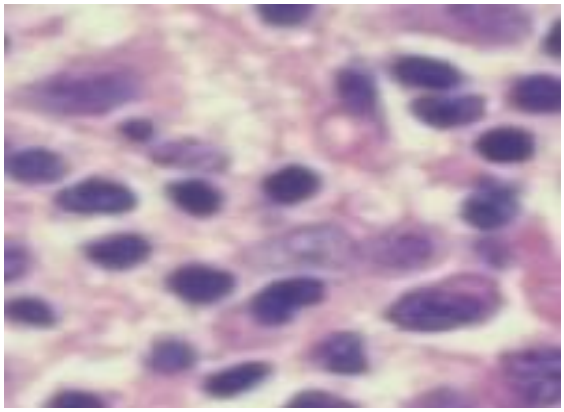

Figure 2: Plamacytosis and mononuclear lymphocytes in bonemarrow biopsy

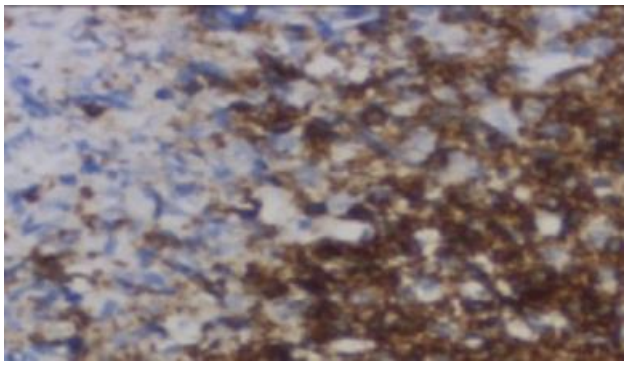

Figure 3: CD 20 positive in immunohistochemistry

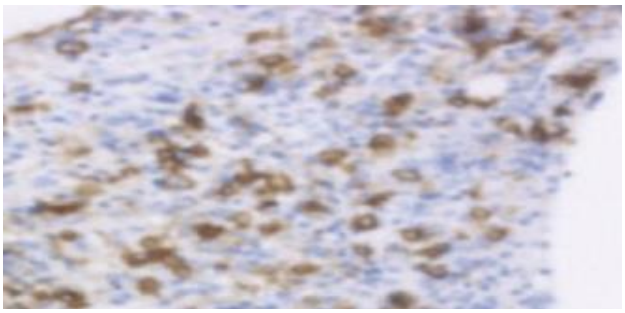

Figure 4: CD 128 positive in immunohistochemistry