

Secondary Amyloidosis Involving the Bone Marrow: A Rare Case Report

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Received: March 13, 2019; Accepted: March 26, 2019; Published: March 28, 2019

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Abstract

Amyloidosis encompasses a heterogeneous group of diseases caused by extracellular deposition of fibrillary protein. Amyloid deposits in AA Amyloidosis are composed mainly of serum amyloid protein, an apolipoprotein of High Density Lipoprotein (HDL) that serves as an acute phase reactant. Amyloid is usually not seen or it is very sparsely presented in a Bone Marrow aspirate or biopsy. Herein we discuss a case of 55 years old male with this rare presentation of secondary amyloidosis, involving bone marrow.

Key Words: BM- Bone Marrow; CT Scan - Computed Tomography Scan; TLC- Total Leucocyte Count; HDL- High Density Lipoprotein

Introduction

A 54 year old male patient, known case of Chronic Liver Disease being treated for Abdominal Koch's for last 4 months presented in the Medicine OPD with the complaint of persistent abdominal distention for 2 months. It was associated with a decreased oral intake, shortness of breath and a decreased urine output. On general physical examination the patient had icterus, gross ascites and hepatomegaly. There was no associated splenomegaly or lymphadenopathy. The liver span was 13cm. Contrast Enhanced CT Scan revealed hepatomegaly with acute or chronic liver disease with findings suggestive of possibility for Amyloidosis, leading to Portal Hypertension that cannot be ruled out. X-Ray Pelvis did not reveal any lytic lesions. Liver function Tests were deranged showing the following parameters: total Bilirubin raised to 2mg/dl (n= 0.2-1.2mg/dl); Alkaline Phosphatase raised to 953U/L (n= 44-147 U/L) and Gamma Glutamyl Transferase raised to 1956 U/L (n= 44-147U/L). Renal Function Tests were deranged showing the following parameters. S. Urea 139mg/dl (n= 7-20mg/dl), S. Creatinine 4mg/dl (n= 0.6-1.2mg/dl), S. Uric acid 7mg/dl (n= 3.4-7mg/dl). Complete Hemogram revealed Leucocytosis and Thrombocytopenia. Hemoglobin was 11.6g/dl (n= 13.5-17.5g/dl), TLC 15.6×10^3 /microl (n= $4-11 \times 10^3$) and platelet count 97×10^3 /microl (n= $1.5-4 \times 10^3$). Ascitic Fluid Biochemistry revealed Adenine Deaminase levels <4 U/l with 2100 mononuclear cells/mm³ suggesting availability of transudate.

Hence for confirmation a Bone Marrow Aspirate and Biopsy were done. Bone Marrow Aspirate smears showed cellular particles with normal maturation in Erythroid, Myeloid and Megakaryocyte series. There was no dysplasia seen amongst the hematopoietic cells. The myelogram was nRBC39 Myelocytes24 Metamyelocytes20 Stab07 Neutrophils02 Plasma cells 05 Lymphocytes03. However, an extensive deposition of a pink amorphous extracellular material between these hematopoietic cells was established (Figure1, Figure 2). In staining of the same material with Methyl Violet metachromatically positivity was assessed (Figure 3), which suggested Amyloid deposition.

Bone marrow trephine biopsy showed mildly hypocellular bone marrow with adequate representation of erythroid, myeloid and megakaryocyte series. In addition, there was presence of abundant extracellular eosinophilic amorphous material (probably amyloid) in the vessel wall and interstitium (Figure 4). This material was also metachromatically positive for methyl violet. Immunohistochemistry for Serum Amyloid, a Protein assay was done for confirmation, and the probe was reported as positive (Figure 5).

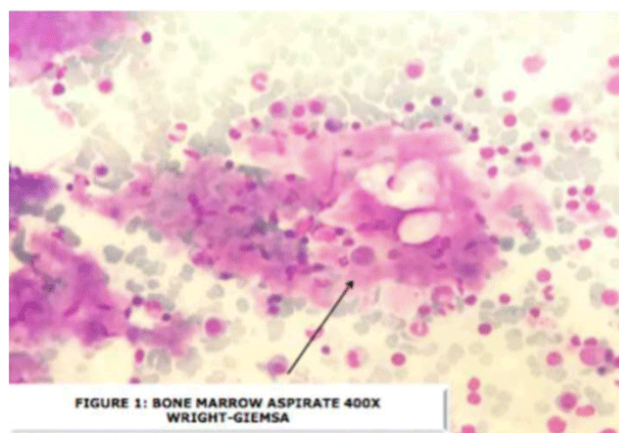


Figure 1: Bone marrow aspirate 400x Wright- Giemsa

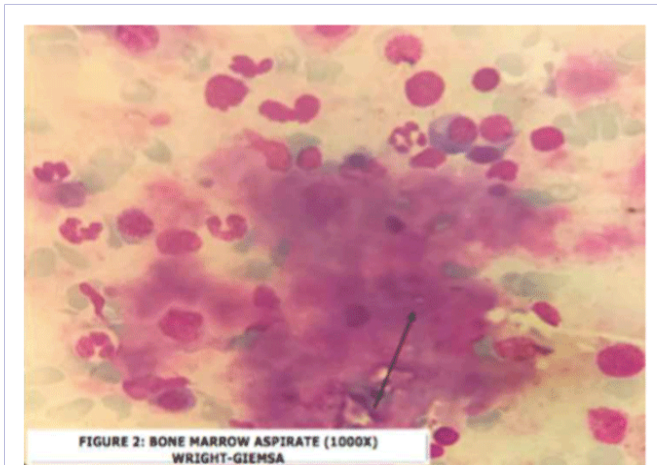


Figure 2: Bone marrow aspirate (1000x) Wright - Giemsa

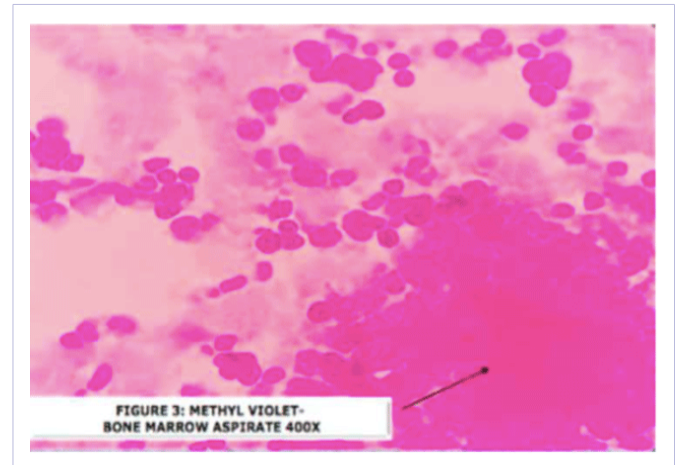


Figure 3: Methyl violet bone marrow aspirate 400x

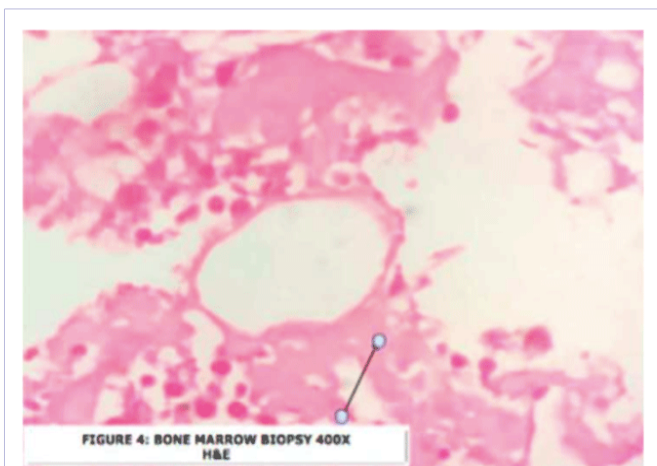


Figure 4: Bone marrow Biopsy 400x H&E

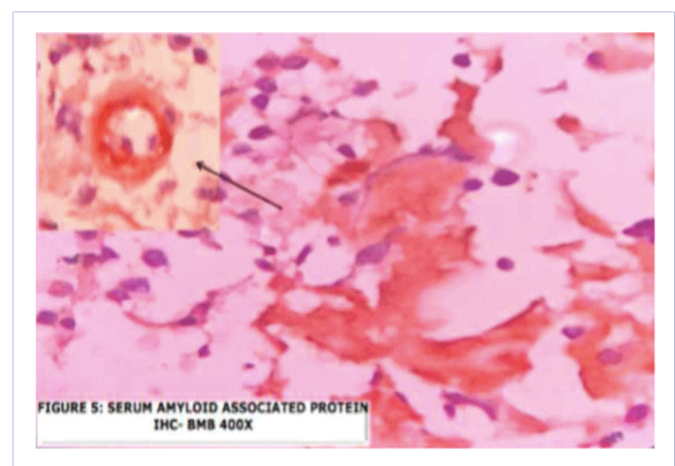


Figure 5: Serum Amyloid associated protein IHC-BMB 400x

Serum Protein Electrophoresis with immunofixation revealed a polyclonal increase in the Kappa Light chain to 79.20 mg/dl and Lambda Light chain to 93.60 mg/dl. The b2 microglobulin levels were increased to 9687ng/ml. Myeloma protein panel showed an albumin level of 1.73 g/dl. No Myeloma Band was detected, hence ruling out Multiple Myeloma.

So, a clinicopathological diagnosis of Secondary Amyloidosis involving the bone marrow was made.

Discussion

Amyloidosis encompasses a heterogeneous group of diseases, caused by the extracellular deposition of autologous fibrillar protein, which aggregates into a 3-D beta-lamina disposition that impairs normal organ function [1].

Amyloid deposits in AA Amyloidosis are composed mainly of Serum Amyloid A Protein, an apolipoprotein of HDL that serves as an acute phase reactant [2].

Amyloid is usually not seen or it is very sparsely presented in aspirate or biopsy. Much of our information regarding amyloid involving bone marrow is based on primary amyloidosis.

On extensive research we could find a single study by Sungur et al, who reported secondary amyloidosis involving bone marrow due to renal failure caused by Familial Mediterranean Fever [3]. The current study conducted bone marrow biopsy on 14 out of 39 patients. Amyloid deposition was seen in 11/14 (78.6%) of the patients.

Secondary amyloidosis involving bone marrow amyloid deposition is very unusual and is rarely reported on the literature.

References

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