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Original Article

A Rare Case of Waldenstrom Macroglobulinemia Presenting as Pancytopenia

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ABSTRACT

Monoclonal gammopathy of any concentration. It's a variant of Lymphoplasmacytic lymphoma with Bone marrow involvement & IgM monoclonal gammopathy. Waldenstrom macroglobulinemia constitute around 2% of hematological malignancy. It's rare slowly progressive disorder with clinical features of Anemia, Thrombocytopenia malignancy, hepatosplenomegaly & lymphadenopathy. The case is remarkable because patient didn't show any common clinical features of Waldenstrom macroglobulinemia.

Key Words: *Waldenstrom; Macroglobulinemia; Pancytopenia; Anemia*

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INTRODUCTION

Waldenstrom macroglobulinemia is a rare malignancy of mature B cell characterized by monclonal IgM in the blood & presence of small lymphocytes, lymphoplasmacytic cells & plasma cells [1]. WM is a variant of lymphoplasmacytic lymphoma with high concentrations of monoclonal immunoglobulin protein in blood. The clinical features due to marrow infiltration include cytopenia, anaemia which manifests as fatigue and weariness along with other symptoms such as night sweats or weight loss and fever. Lymphadenopathy, hepatosplenomegaly is observed due to infiltration of peripheral tissue which is seen in 20-30% cases [2]. Due to increase IgM paraproteins in the circulation, it manifests as symptoms of hyper viscosity, mainly blurring of vision, headache & stroke. Neuropathies occur because of reactivity of IgM paraprotein with myelin sheath. Igm may deposit in gastrointestinal track can cause diarrhoea. Coagulopathy may occur because Of binding of IgM to clotting factors. The skeletal involvement in most the cases is absent [3]. The L265P mutation in MYD 88 is detectable in 90% cases. Here we report a case in a patient who presented with pancytopenia along with neurological symptoms. proper haematological examination is necessary for early diagnosis & preventing major complications [4].

Case Presentation

68 years old male patient presented with complaint of weight loss, generalized weakness and decreased appetite last since 3 months. Patient was conscious but disoriented. Patient had no icterus, lymphadenopathy or hepatomegaly. USG reveled mild splenomegaly. Patient's relative gave history of his forgetfulness & repetition of commands. The hemogram show Hb-2gm/dl, TLC- 3800/cubic mm, platelets count -54000/cubic mm, RBCs count of 1.1 million /cubic mm. Peripheral blood smear reveled rouleaux formation (Figure *1*). DLC showed 30% polymorphs, 60% lymphocytes, 6 % monocytes & 4% eosinophils. ESR was 58 mm/hr. Blood group could not be determined because of auto agglutination. M band was seen on protein electrophoresis. Total protein 9.8g/dl & albumin 2.5g/dl. HCV was not nonreactive, osteo-skeletal survey was normal. KFT was within normal limits. Serum calcium was 9.9mg/dl. In view of pancytopenia bone marrow aspiration was performed but was failure. So, bone marrow biopsy was performed, bone marrow imprint and biopsy were studied. Biopsy imprint reveled plasmocytic lymphocytes, plasma cells and atypical lymphocytes along with increased number of mast cells (Figure *2*). Bone marrow biopsy reveled total effacement of architecture by diffuse population of lymphoid cells and plasma cells (Figure *3*) (Figure *4*) (Figure *5*).

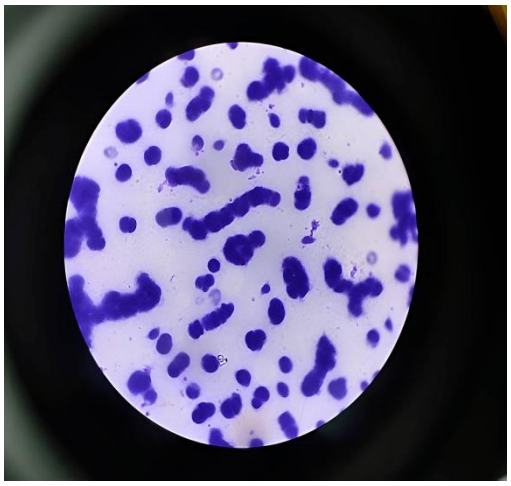


Figure 1: Peripheral smear showing rouleaux formation.

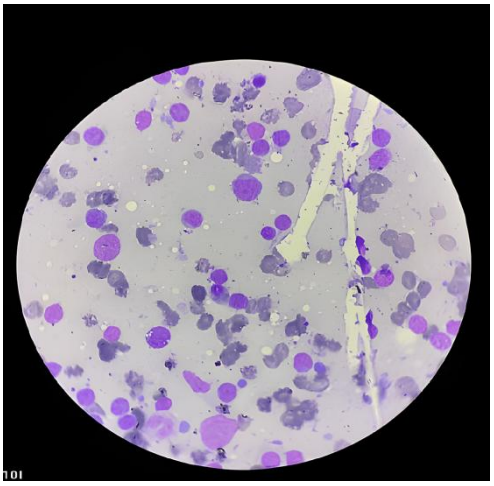


Figure 2: Bone marrow showing atypical lymphocytes

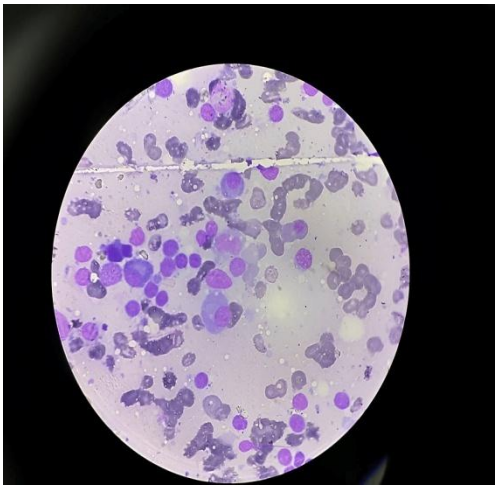


Figure 3: Bone marrow imprint showing plasma cells.

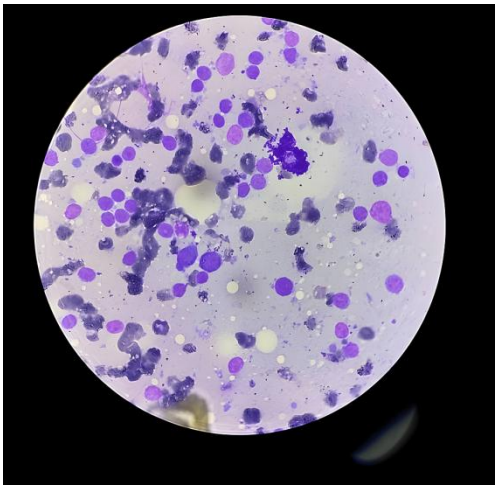


Figure 4: Bone marrow showing mast cells

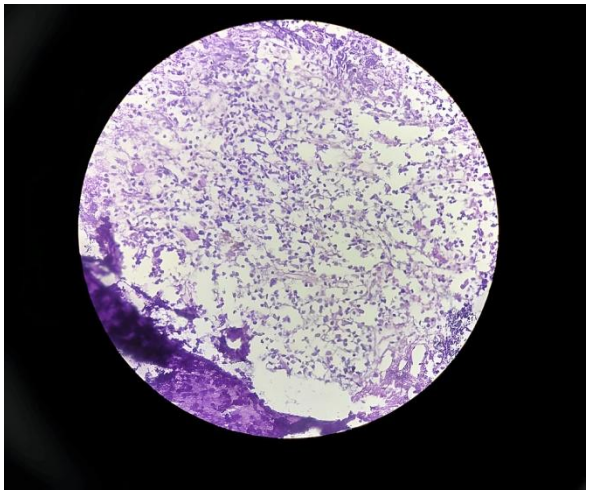


Figure 5: Bone marrow biopsy showing effacement of architecture

Immunohistochemistry was done as flowcytometry could not be performed. IHC reveled lymphoid cells immuno-positive for LCA (CD-45) (Figure *6*) CD-20 (Figure *7*)/ Pax-5 (Figure *8*) & immuno-negative for CD-138/ CD-38/MUM-1/BCL- 6/ Cyclin D1/Annexin A-1/MPO. A small lymphoid cell exclusively express Kappa light chain (Figure *9*) & immuno-negative for Lambda light chain.

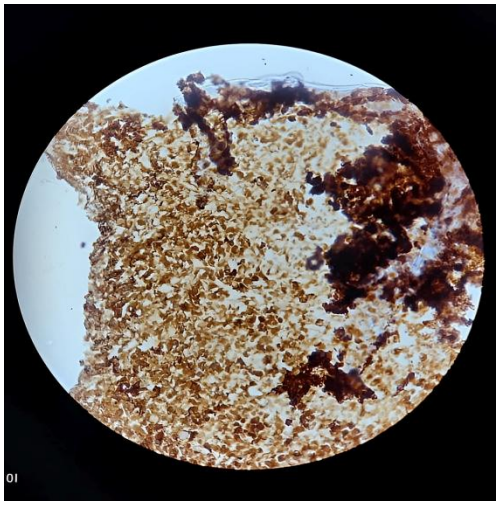
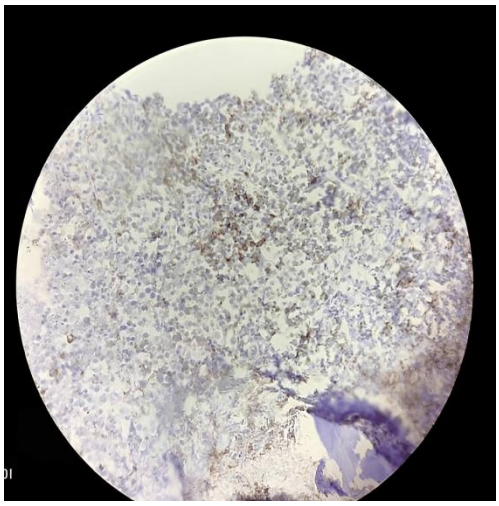


Figure 6: IHC-LCA positive Figure



7: IHC-CD20 positive

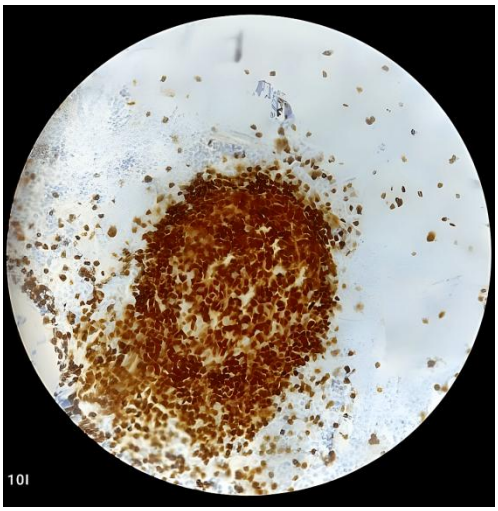
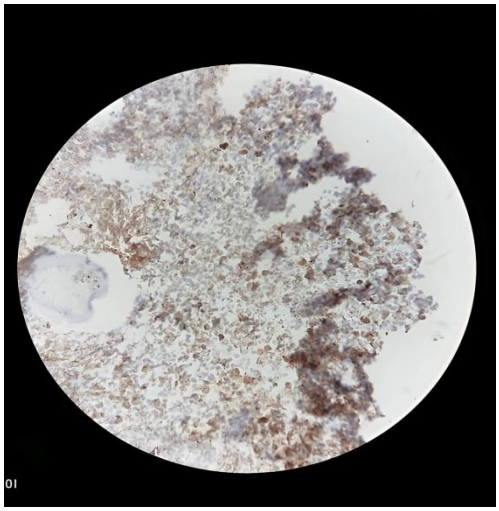


Figure 8: IHC pax-5 positive Figure



9: IHC. -kappa light chain positive

DISCUSSION

Jan G. Waldenstrom, a Swedish physician was the first to document Waldenstrom macroglobulinemia. He had reported six cases that had the features of bleeding from the nose and mouth, anaemia, enlarged lymph nodes and also found decreased fibrinogen on bloodwork [5]. Upon further investigating, he found the cause of the blood being sticky being the macroglobulin. He also found the bone to have proliferating plasma cells [6]. According to WHO, Waldenstrom Macroglobulinemia is a neoplasm of small B lymphocytes, plasmacytoid lymphocytes and plasma cells involving bone marrow & sometimes Lymph nodes & spleen [7]. Excluding conditions that do not meet the criteria of small B cell lymphoid neoplasm having plasmacytoid differentiation, a staggering majority of LPL> 90% have MYD88L265P mutation [8].

WM accounts for 1-2% of haematological malignancy. It occurs in adults with median age in seventh decade with slight male preponderance. Some cases of LPL are associated with hepatitis C virus infection which responds well to antiviral therapy.

WM is rare tumour accounting for 5 per million cases per year. Our patient was a 68 year old who presented with chief complaints of weight loss, generalised weakness and decreased appetite for 3 months. Patient was conscious but disoriented. This patient had no lymphadenopathy but was found to have mild splenomegaly on USG. CBC & PS findings were of pancytopenia Haemoglobin 2gm%, TLC 3000/cumm, platelet 54000/cumm. Rouleux formation was seen along with bluish tinge to the peripheral smear in background, Bone marrow aspiration was dry tap. Bone marrow biopsy was performed along with imprint smears. Serum Protein electrophoresis show M spike. Imprint smear revealed lymphocytes, plasmcytoid lymphocytes, plasma cells & mast cells. Erythropoiesis, myelopoiesis & megakaryopoesis was suppressed. Immunohistochemistry was performed. The lymphoid cells were immune positive for LCA (CD45)/CD -20/Pax 5 & immune negative for CD 38/138/MUM-1/Bcl6/cyclin D1/Annexin A1 & MPO. These lymphoid cells were immune negative for Lambda light chain but expressed Kappa light chain.

The 2016 WHO criteria has classified mature B cells neoplasms including lymphoplasmacytic lymphoma as a subcategory Even though lymphoplasmacytic lymphoma is often seen in association with an IgM paraprotein, this is not essential criteria for diagnosis [9]. Although Waldenstrom macroglobulinemia is seen in a considerable category of patient with lymphoplasmacytic lymphoma yet it is not the same as WM; it is better defined as Lymphoplasmacytic lymphoma with involvement of bone marrow and any concentration of IgM monoclonal gammopathy [10]. It has to be differentiated from other small B -Lymphocytes neoplasm showing plasma cell differentiation. A vast population (> 90%) of LPL have MYDBB L265P mutation.

CONCLUSIONS

A diagnosis of Waldenstrom macroglobulinemia was offered on the basis of morphology & Immunohistochemistry of bone marrow. Waldenstrom macroglobulinemia is rarely reported in the Indian clinical setting, usually present with generalized weakness and seen with a median age in seventh decade of life with male preponderance. this case is unusual because the patient didn't exhibit the expected features of Waldenstrom macroglobulinemia except for pancytopenia. A thorough blood work including bone marrow studies, immunohistochemistry along with serum electrophoresis are essential in distinguishing Waldenstrom macroglobulinemia from other lymphomas.

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