

## A Three Year Old Girl with WaldenstromMacroglobulinemia- A Rare Case Study

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**Abstract:** Waldenstrommacroglobulinemia a chronic beta cell lymphoproliferative disorder characterised by IgM monoclonal gammopathy, bone marrow infiltration by lymphocyte, lymphocytoid cells & plasma cells. It accounts for approx. 2% of all haematological malignancies. It is rarely seen below 50 yrs. of age. A small child (3 yrs. old girl) having monoclonal gammopathy, bone marrow lymphoplasmacytosis, organomegaly & lymphadenopathy was diagnosed as WALDENSTROM MACROGLOBULINEMIA. It is a rare disease and rarest at this age

**KeyWords:** Waldenstrommacroglobulinemia, child, monoclonal gammopathy, lymphoplasmacytosis, organomegaly,

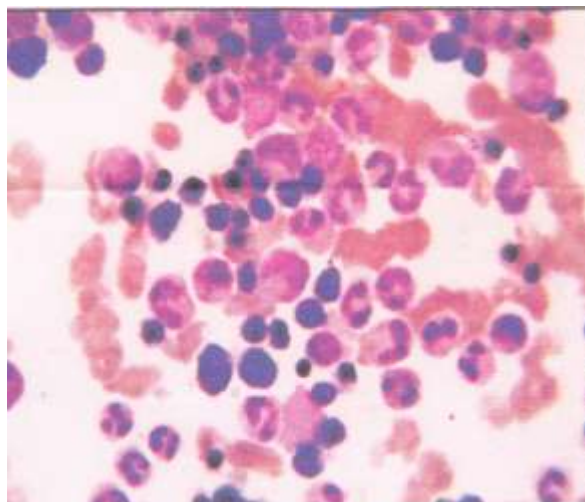
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### I. Introduction

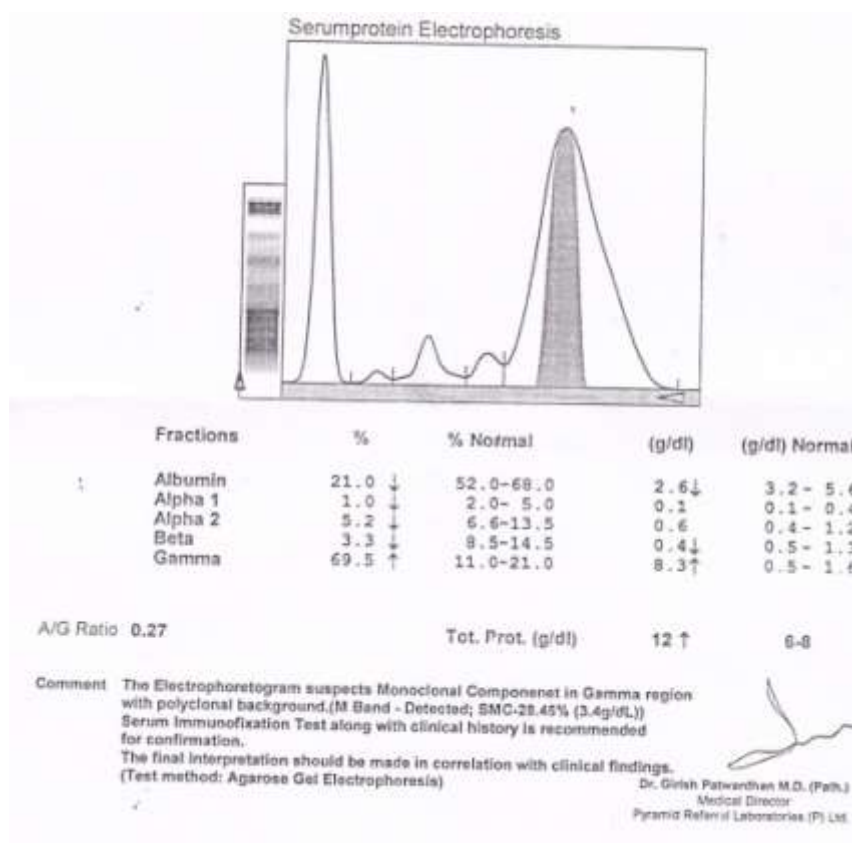
Waldenströmmacroglobulinemia (WM) is a chronic B- cell lymphoproliferative disorder characterized by IgM monoclonal gammopathy and bone marrow infiltration by lymphocytes, lymphoplasmacytoid cells, and plasma cells. It accounts for approximately 2% of all hematologic malignant neoplasms and has an annual incidence of 6 per 1 million in white males, in whom the incidence is highest. The clinical features are highly variable; many patients are asymptomatic, while others have advanced lymphoma. The IgMparaproteinemia may result in hyperviscosity syndrome and cryoglobulinemia, features considered to be highly characteristic of the disorder. The IgMparaprotein also may have autoantibody activity, which may result in peripheral neuropathy, cold agglutinin hemolysis, immune thrombocytopenia, mixed cryoglobulinemia, and acquired von Willebrands disease. There are no accepted criteria for the diagnosis of WM. The majority of published clinical trials have accepted the presence of IgM monoclonal gammopathy in the context of an apparently indolent lymphoproliferative disorder as sufficient evidence for a definitive diagnosis of WM. IgMpara proteins are, however, demonstrable in all subtypes of peripheral (mature) B-cell lymphoproliferative disorders. It has been suggested that paraprotein concentration is useful for differentiating WM from other lymphoproliferative disorders. The French-American-British cooperative group suggested that a concentration of 20 g/L was diagnostic of WM, while Kyle andGarton chose 30 g/L to define WM in a clinico pathologic review of 430 patients with IgM monoclonal gammopathy. The Revised European-American classification of lymphoid neoplasms (REAL) and World Health Organization (WHO) classification criteria consider WM to be a clinical syndrome occurring in most patients with lymphoplasmacytic lymphoma, which is defined as a disorder of small lymphoid cells that show maturation to plasma cells without the features of other lymphoma types.

### II. Case History

A 3yr old girl presented to the clinic for papular rash, abdominal swelling, fever & loss of appetite. Rash, fever for one week & abdominal swelling, anorexia for one month. All thought to be due to hepatic cause so was investigated in the line of hepatic disease. Investigation showed normal SGPT, SGOT, serum bilirubin and alkaline phosphatase. Serum protein was high with high globulin level and A/G ratio was reversed PT was 28 secs. Ultrasound of abdomen showed hepatomegaly, splenomegaly and abdominal lymphadenopathy. Patient had anaemia, high ESR Urinary B/S, B/P absent urobilinogen normal, urinary protein ++, microscopic haematuria. So at this stage it was inferred that the disease is not due to hepatic cause and it is a case of hyperglobulinaemia. So in next stage serum protein electrophoresis, skull Xray, and bone marrow aspiration cytology was done Serum protein electrophoresis showed monoclonal "M" band, this monoclonal "M" band was in the background of polyclonal "G" band, it was more than 3.4gm% total protein was 12 gm%



PHOTOMICROGRAPH of bonemarrow showing lymphoplasmacytosis



Immunofixation test was not done to confirm the “M” bandSkull x ray showed no osteolytic lesion, sr. calcium was normalBone marrow aspiration cytology showed lymphoplasmacytoid cells

### References

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