

Impaired Splenic Function in Systemic Amyloidosis: Diagnostic Importance of Peripheral Blood Film

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Introduction

We describe case of a 40 year old male, who went on to developed systemic amyloidosis 3 years after the diagnosis of chronic renal failure. The diagnosis of systemic amyloidosis was suspected upon a routine examination of periheral blood film showing features of hyposplenism. We would like to highlight the importance of examination of pheripheral blood film in patients with renal failure for the diagnosis of extensive systemic amyloidosis which occasionally leads to functional hyposplenism, recognized by the presence of abnormal red cell morphology and reduced splenic uptake on isotope scan¹.

Case Report

A 40 years old male whose co-morbid included a long history of hypertension also suffered from pulmonary tuberculosis at the age of 18 years. Three years back he presented at a hospital in United States of America with complaints of fever, vomiting and lethargy of a months duration. Laboratory investigations revealed deranged renal functions and an ultrasound abdomen showed presence of shrunken kidneys. Hence a diagnosis of chronic renal failure was made. He was then started on hernodylasis.

He subsequently required hospital admissions in December 1997 for the management of urinary tract infection and later on in January 1998 with bilateral pleural effusions. Finally in view of his deteriorating renal functions and henceforth its various complications he was offered the option of renal transplantation.

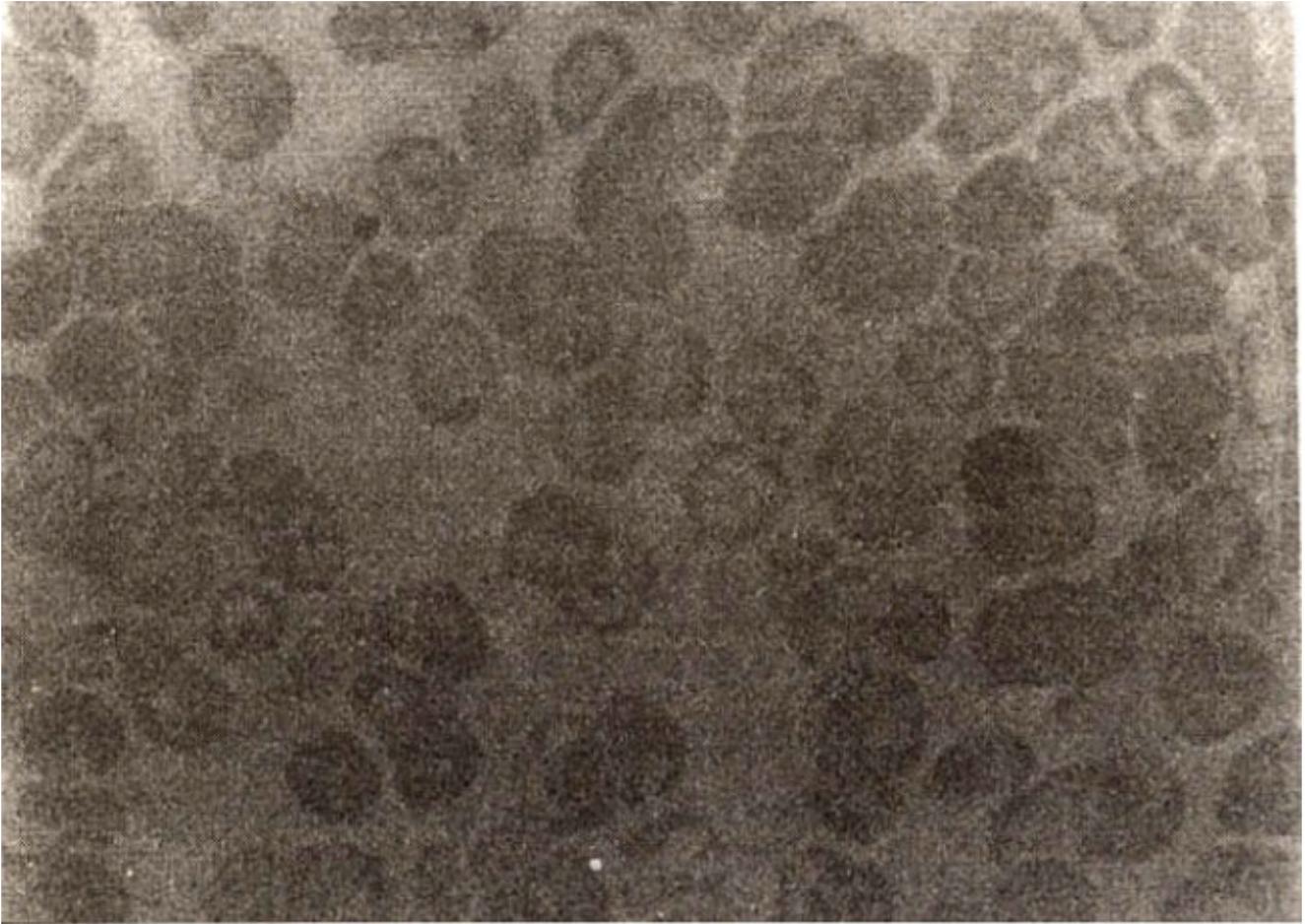


Figure 1. Peripheral blood film shows features of hyposplenism: anisocytosis, poikilocytosis, target cells, nucleated red cell and howell jolly bodies. Also seen in this blood film are  burr cells characteristic of renal damage (Romanowsky stain x 100 magnification).

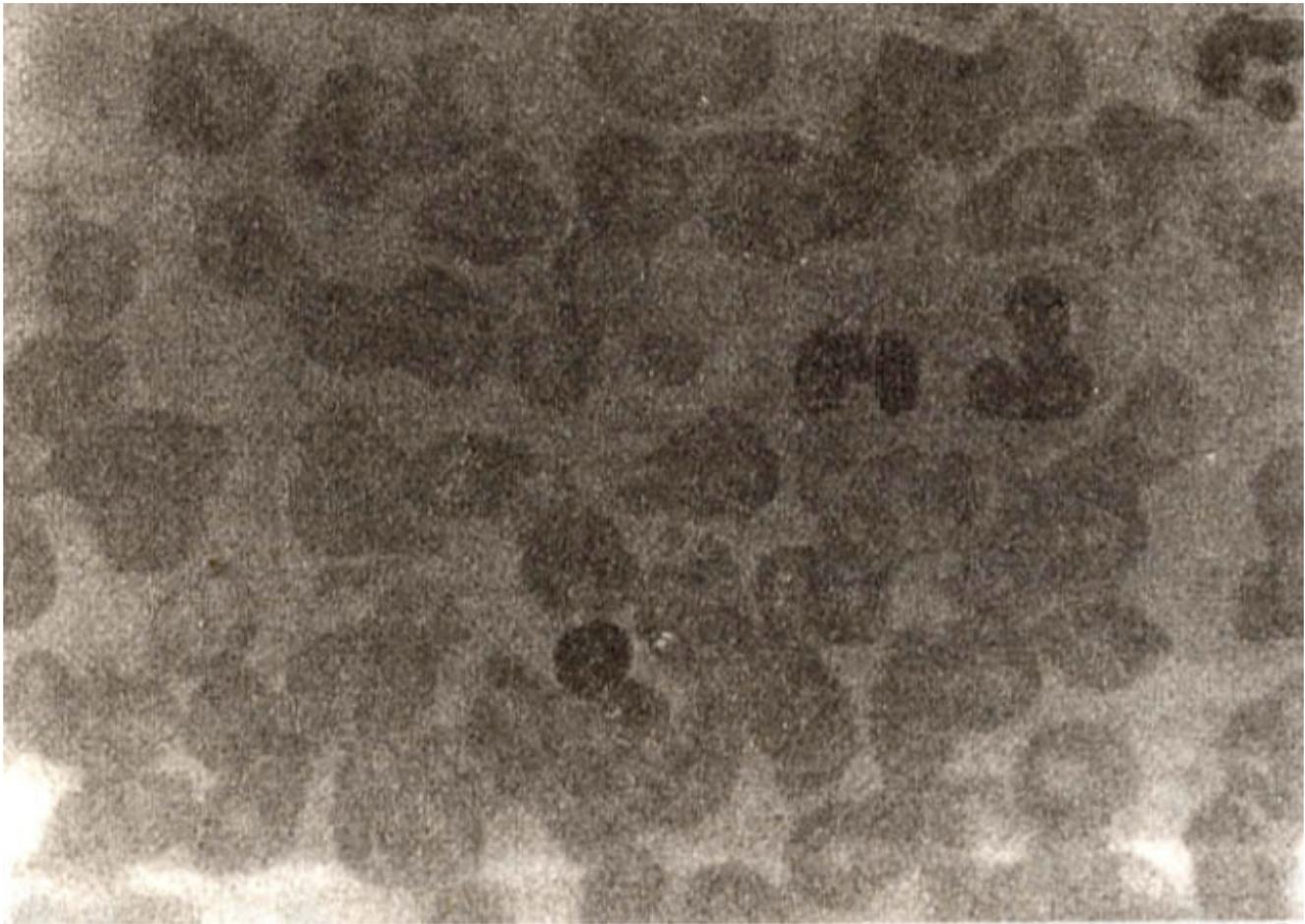


Figure 2. Peripheral blood film reveals features of hyposplenism along with the presence of neutrophils and large platelets (Romanowsky stain x 100 magnification).

He underwent renal transplantation in November 1998. In the following month he suffered from gastrointestinal infection as a result of post transplant immunosuppression.

He remained symptom free till May 2000 and then went on to develop perinephric collection of infected fluid which was drained through a nephrostomy tube. As events unfolded, he was finally hospitalized again in October 2000 with complaints of loose motions, vomiting and generalized abdominal pain. On examination he had pallor and pedal edema. Rest of the examination was unremarkable. At the time of admission his relevant laboratory investigations were as follows: serum creatinine 7.1 mg/dl (normal range INRI 0.85-1.35), blood urea nitrogen 81 mg/dl (6-16), total proteins 6.1 gm/dl (5.7-7.75), serum albumin 2.3 g/dl (3.5-5.0), serum globulin 3.8 g/dl (1.8-3.2), albumin/globulin ratio (AIG Ratio) 0.6 and creatinine clearance: 44.84 ml/min (97-137).

His hemostatic profile showed a normal prothrombin (PT) and activated partial prothrombin time (APTT). Peripheral blood counts were: hemoglobin 9.3 gm/dl (13.7-16.3), hematocrit 28.3% (41.9-48.7%), corrected white blood cells count $20.6 \times 10^9/L$, platelet count 166,000 with normal red cell indices. Peripheral blood film revealed anisocytosis, macrocytes, hypochromia, poikilocytosis, dimorphic red cell population, target cells, burr cells, fragmented red cells, nucleated red cells, polychromasia and howell jolly bodies. Occasional left shift neutrophils were seen along with large platelets. On the basis of peripheral blood film findings a strong suspicion of hyposplenism was made and various causes of impaired splenic function were considered including systemic amyloidosis. Subsequently a definitive diagnosis of systemic amyloidosis was made based on rectal and renal biopsy. A bone marrow trephine also showed congo red stained amyloid deposits. ^{99m}Tc -sulfur colloid or microlite scans isotope scan of the spleen however was not done.

Patient later on developed laceration of the lower pole of the kidney and ultimately died of overwhelming sepsis and disseminated intravascular coagulation.

Discussion

Amyloidosis is a rare monoclonal plasma cell disorder. The evaluation procedures include clinical signs and symptoms - and immunoelectrophoresis and immunofixation of serum and urine. The findings of a monoclonal protein necessitates a biopsy to confirm the diagnosis. Biopsy of the affected organ confirms the diagnosis but congo red staining of bone marrow specimen or subcutaneous fat aspirate are non-invasive, sensitive and less expensive, procedures². Recently attention has been drawn to hyposplenism in systemic amyloidosis^{3,4}. Infact poor splenic function is now an accepted finding in a wide range of systemic disorders including coeliac disease, ulcerative colitis, Crohn's disease, sickle cell disease, primary thrombocythemia and systemic lupus erythmatosus⁵. Involvement of spleen in systemic amyloidosis affects the reticulum of the pulp and venous sinuses which in turn alters one the most important functions of spleen i.e., phagocytosis and sequestration. This functional hyposplenism results in the presence of intraerthrocytic inclusions (i.e., howell jolly bodies) and abnormally shaped erythrocytes (i.e. poikilocytes). This indeed has been previously reported in the literature^{6,7}. In this case, findings on the peripheral blood film strongly suggested impaired splenic function which, later on helped in establishing a diagnosis of systemic amyloidosis through rectal and bone marrow biopsy.

In conclusion, this case further highlights and strengthens the importance of peripheral blood film in the diagnosis of functional hyposplenism secondary to systemic amyloidosis.

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