

Background

Myelofibrosis, a cytokine mediated process of bone marrow stroma and its replacement with fibrous connective tissue, has been associated with neoplastic conditions, autoimmune disorders, and granulomatous diseases among other etiologies. Although Epstein-Barr virus (EBV) infection has been associated with pancytopenia, its rare association with myelofibrosis has only been reported in few pediatric patients.

A recent case of a 21 year old female who presented with 3 week history of fever and anorexia and was found to have pancytopenia and acute EBV infection, and upon examination of the bone marrow, reticulin fibrosis and gelatinous atrophy of the bone marrow is reported. Her symptomatology and blood counts normalized upon institution of antiviral treatment.

Materials and Methods

Wright-stained slides of the peripheral smear, bone marrow aspirate, in addition to the Hematoxylin and Eosin-stained slides of the bone marrow biopsy and clot sections were evaluated. Iron, Acid fast bacilli (AFB), Gomori Methenamine Silver (GMS), and reticulin stains of the bone marrow biopsy and the clot sections were examined.

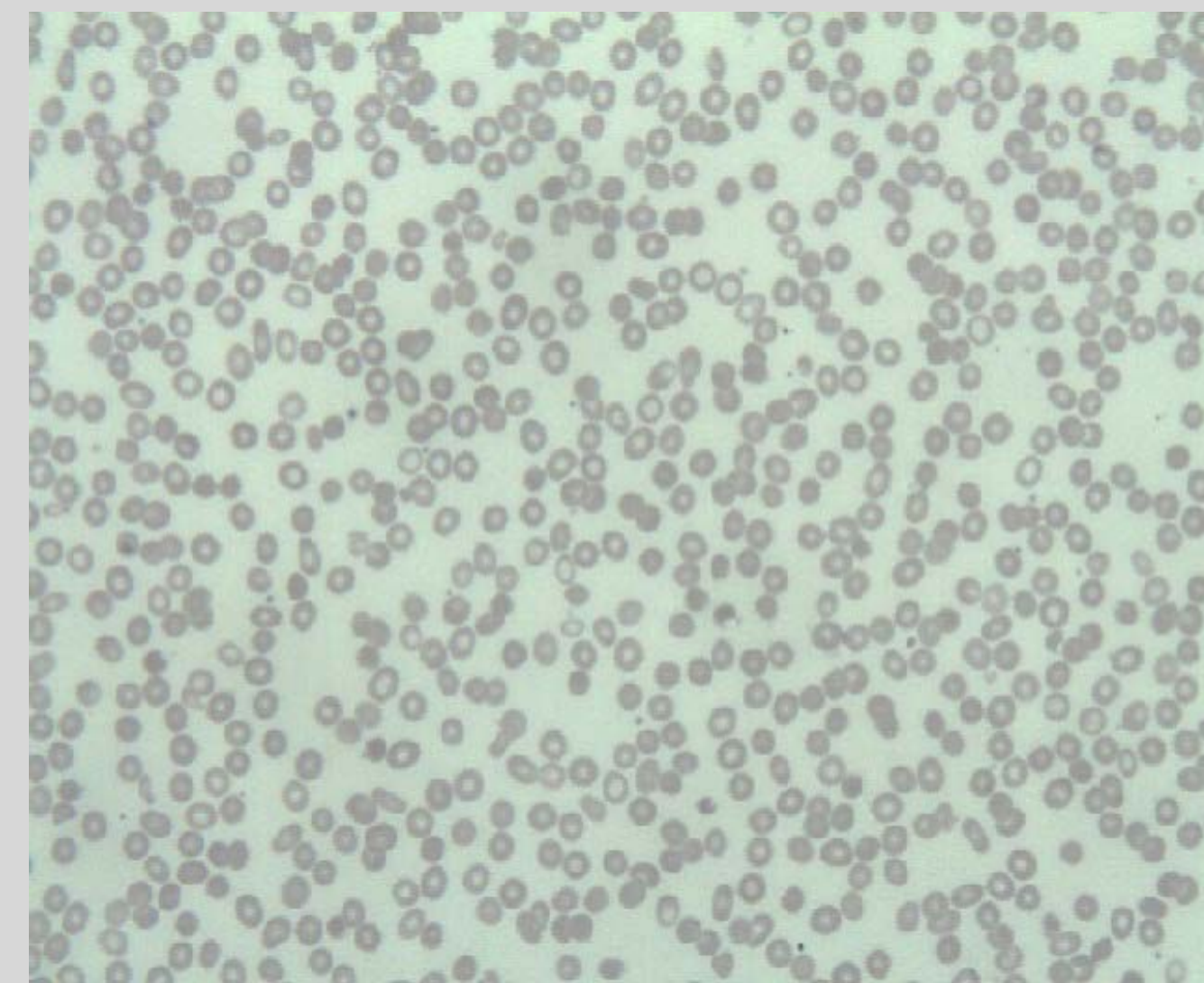


Figure 1. Peripheral blood smear showed hypochromic anemia with slight polychromasia, a few microspherocytes, and no increase in schistocytes. WBCs and platelets were normal in morphology. Mag. 100X

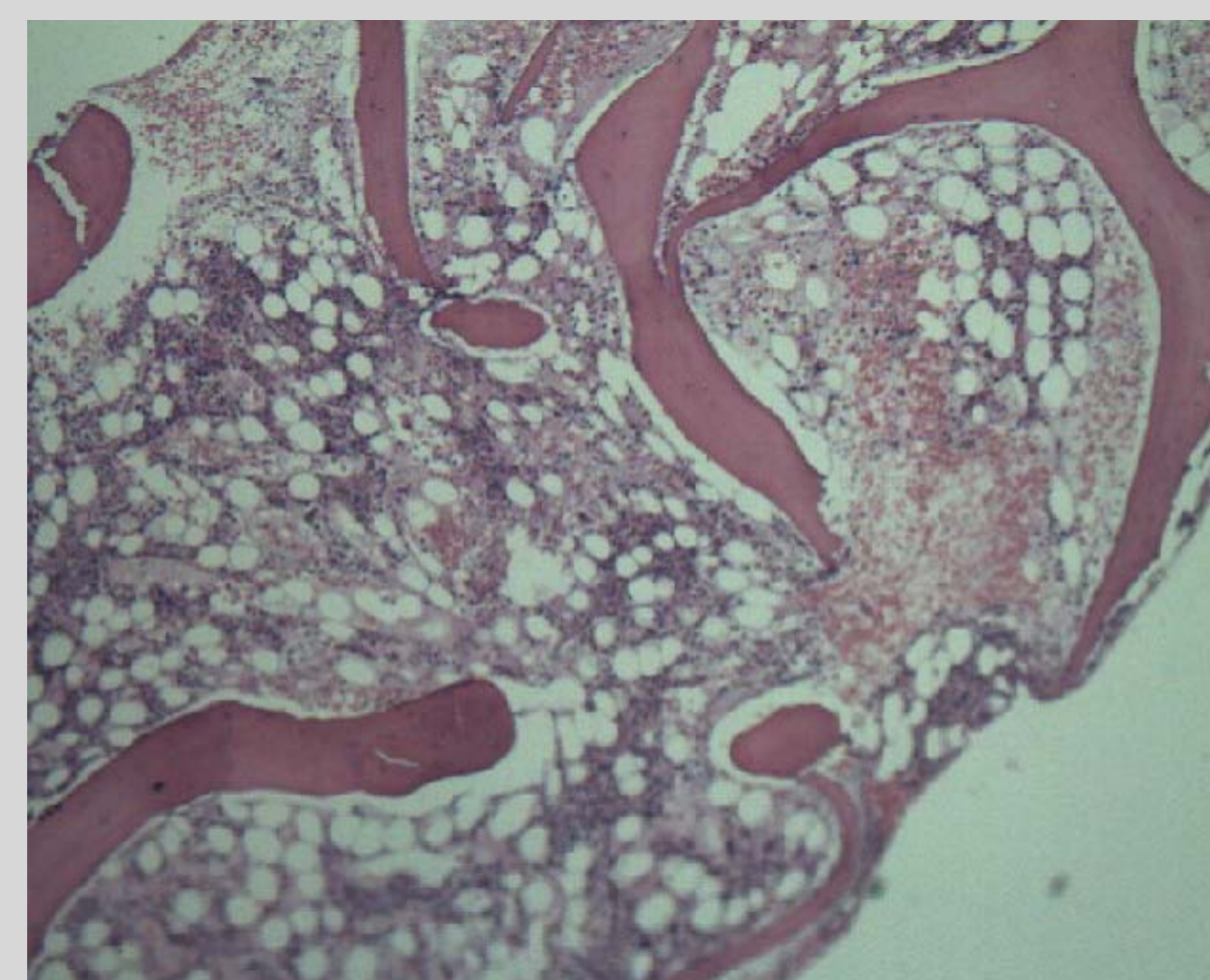


Figure 2/ 3: Evaluation of bone marrow biopsy was remarkable for hypocellularity. Estimated cellularity was ~40%, while expected for the age of the patient was ~80%. Mag. 40X.

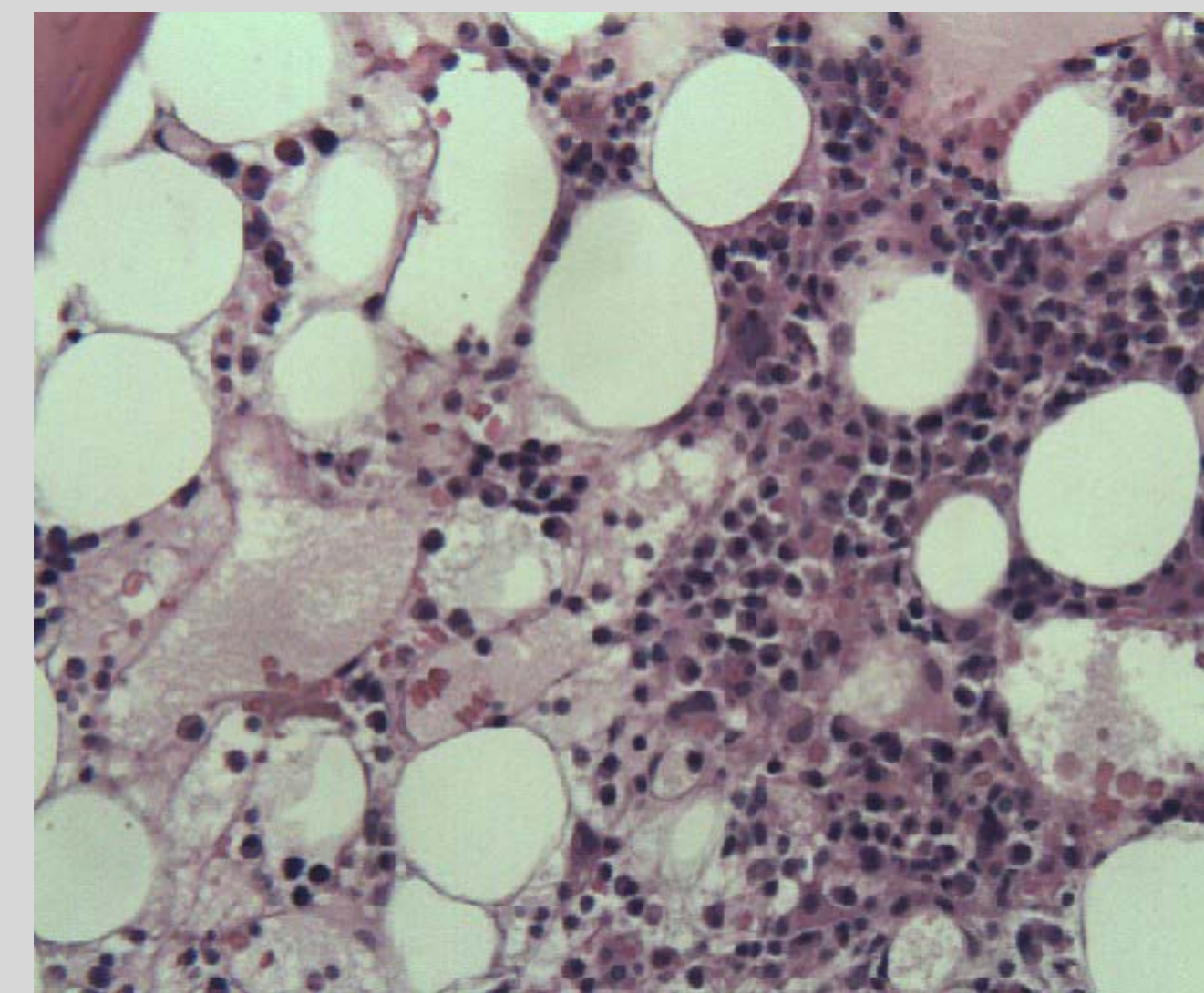


Figure 4. Bone marrow biopsy showed scattered foci of serous/gelatinous atrophy, characterized by accumulation of lightly staining eosinophilic serous fluid (mucopolysaccharides rich in hyaluronic acid) in the interstitium. This condition has been associated with severe malnutrition/anorexia, febrile illness, and malignancies among others. Mag. 100X.

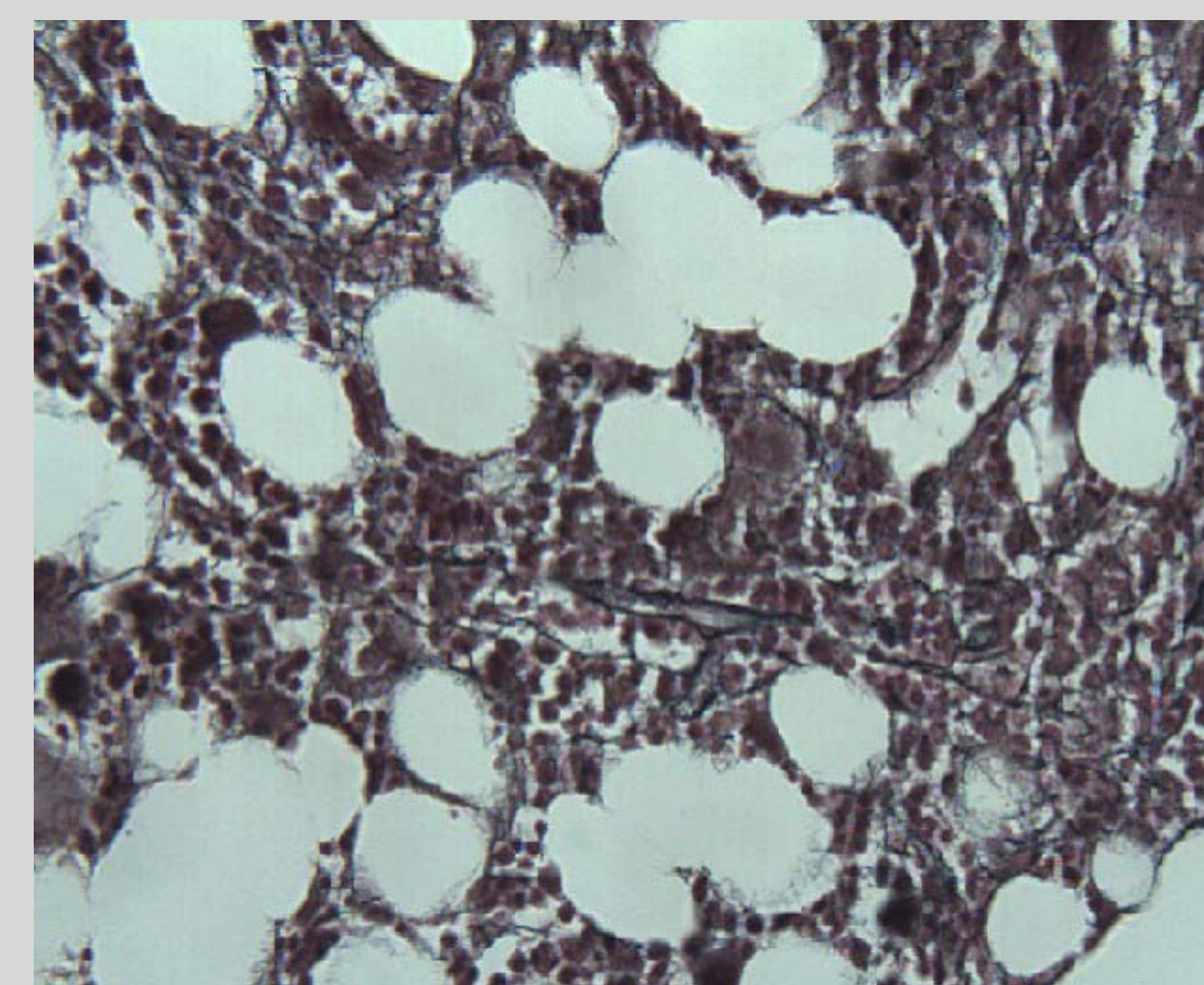


Figure 5. Bone marrow biopsy, silver stain, showed grade 2 reticulin fibrosis with diffuse fine and scattered coarser fibers. Mag. 100X.

Results

The peripheral smear showed hypochromic anemia with slight polychromasia. White blood cells and platelets were normal in morphology but decreased in number. There was no increase in schistocytes. Bone marrow aspirate and touch preparations showed normal trilineage maturation and no increase in blasts. The bone marrow was hypocellular for age (40%), with no evidence of granuloma or malignancy, but with focal areas of serous atrophy. Iron, Acid fast bacilli (AFB), Gomori Methenamine Silver (GMS), and reticulin stains showed increased iron stores with no ringed-sideroblasts. AFB and GMS stains were negative. However, the reticulin stain showed moderate reticulin fibrosis with diffuse increase in reticulin fibers, occasional denser strands, and with many intersections, most consistent with grade 2 myelofibrosis.

Discussion

The patient's pancytopenia was most likely secondary to bone marrow hypocellularity with reticulin fibrosis. Etiologies of hypocellularity in this patient were considered to include viral infections, immune disorders, and medications, which remained uncompensated for due to failure of bone marrow. Subsequent serologic testing confirmed the diagnosis of acute EBV infection. The patient was started on valacyclovir, with resolution of her symptomatology and improvement of her complete blood count after 5 days of treatment. To the best of our knowledge, this presentation of acute EBV infection has not been previously described in adult patients.

References

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- Thiele J, Kvasnicka HM, Facchetti F, et. al. European consensus on grading bone marrow fibrosis and assessment of cellularity. *Haematologica* 2005; 90: 1128-1132.