

Diagnostic Challenge of Massive Splenomegaly

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Introduction

Poorly differentiated malignant neoplasms confined to the spleen are rare. Their rarity and broad differential diagnosis in the setting of splenomegaly may create a diagnostic dilemma, in this case approached with splenectomy [1,2].

Case Report

A 61 year old Hispanic male with DM II and HTN presented with a several months history of abdominal pain, night sweats, fatigue and early satiety. Physical examination was significant for splenomegaly. Laboratory studies revealed microcytic anemia (Hb 10.2 g/dL, MCV 79) and elevated CRP and ESR (61 and 51, respectively). CBC with differential was otherwise normal. PT, PTT, BMP, LFTs, ferritin were normal. A non contrasted CT scan of the abdomen revealed a massively enlarged spleen with heterogenous appearance. The main differential diagnoses included infections, infiltrative diseases and hematologic malignancies.

RF, LDH, Hb electrophoresis were normal. Serology for brucella, HIV, and histoplasma were negative. EBV serology was compatible with past infection. Tb skin test, blood cultures, and thick and thin blood smear for malaria were negative. Bone marrow biopsy was normal with unremarkable bone marrow flow cytometry. Urine immune fixation electrophoresis showed an increase in free kappa light chains. SPEP showed elevated gamma globulin, with no monoclonal protein identified on immune fixation. IgG level was elevated. Polyclonal increase of gamma globulin fraction can be seen in autoimmune disease (SLE, RA and sarcoidosis) and chronic infections.

Whole body PET CT showed splenomegaly with heterogenous FDG uptake. Multiple hyper densities were seen within the spleen on CT. At that point, the decision was made to proceed with open splenectomy. There was no intraperitoneal evidence of tumor. Spleen weight was 1.6 kg.

Pathologic examination showed an undifferentiated malignant neoplasm, in which lineage was entirely obscure. The tumor cells were obscured by the dense mixed lymphohistiocytic infiltrate. Immunohistochemistry stains for lymphoid, histiocytic, epithelial, myofibroblastic and endothelial origin were all negative. Flow cytometry showed a normal distribution of T cell subsets with polytypic B-cells. An abnormal blast population was not detected. Electron microscopy showed undifferentiated cells without specific cytoplasmic features, but with accentuated features of abortive vascular spaces. Given the fact that the imaging studies prior to splenectomy did not suggest disseminated disease, watchful waiting was chosen.

Discussion

In a patient with massive splenomegaly, differential diagnoses include lymphoma, leukemia, storage disorders, or myelofibrosis [2]. Infections rarely cause a massive spleen enlargement.

Poorly differentiated malignant neoplasms confined to the spleen are rare. The systemic inflammation induced by this tumor and the mimicking a lymphoid neoplasm created an additional diagnostic challenge [3]. Their rarity and broad differential diagnosis in the setting of splenomegaly may create a diagnostic dilemma, in this case approached with splenectomy [4].

References

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