

TEACHING FILES (GRAND ROUNDS)

MASSIVE HEPATOMEGALY - CAN IT BE ACUTE LYMPHOID LEUKEMIA?

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KEYWORDS

ALL, large liver, children

ARTICLE HISTORY

Received 8 February 2020

Accepted 8 February 2020

Clinical Problem:

A 9 years old female child presented to our Pediatric Liver clinic with a loss of weight of 2.5 kgs in 6 months, abdominal distension and loss of appetite for 3 months. There was no fever, bleeding manifestations, bone or joint pain, or need for any blood transfusion. On presentation, her weight was 20 kg (between 3rd-10th percentile as per Indian Academy of Pediatrics (IAP) growth chart), height 129 cm (25th-50th percentile as per IAP growth charts). She had large hepatomegaly and a just palpable spleen. Other systems were normal. Investigations showed hemoglobin 13.5 gm/dl, white blood count 6,440 cells/cumm with polymorphs 52%, lymphocytes 39%, monocytes 8% and platelets of 2,40,000 cells/cumm. Liver function tests were normal. Abdominal ultrasound showed moderate hepatomegaly with mild splenomegaly and normal portal doppler scan. HIV, Hepatitis C Elisa and HBsAg were negative. Serum ceruloplasmin level was normal. Antinuclear antibody (ANA) was weakly positive (+), anti-liver kidney muscle (LKM) and anti-smooth muscle antibodies (ASMA) were negative. Alfa-fetoprotein (AFP) was below 0.2 ng/ml. CT Abdomen showed hepatosplenomegaly with no lesions. Suspecting storage disorder, liver biopsy was done which showed features of metastasis of hematolymphoproliferative malignancy to the liver. Hence bone marrow aspiration was performed which showed increased lymphocytes with blast-like cells. Flow cytometry (CD10 – 74%, CD19 – 41%, CD20 – 68%) confirmed the diagnosis of B cell acute lymphocytic leukemia (ALL). Child has been started with chemotherapy.

Why did the child have normal blood counts inspite of metastatic liver involvement?

Discussion:

Hepatosplenomegaly is common in leukemia and can be due to various factors such as infiltration of leukemic cells, viral hepatitis or drug-induced.¹ However, it is usually associated with other hematological parameters such

as cytopenias and fever. Isolated hepatosplenomegaly as a sole manifestation of leukemia is rare in the pediatric age group.² Liver involvement has been reported to be a frequent finding in patients with ALL, and it can present from asymptomatic hepatomegaly with infiltration of lymphocytes to hepatitis to acute liver failure.³ Extramedullary tumor infiltration into the lymph nodes, liver, spleen, testicles, and central nervous system are common.³ One-third patients with leukemia have infection or fever at presentation and one-half of the patients present with lymphadenopathy, splenomegaly and hepatomegaly.² Pathology of the liver shows diffuse infiltration of the leukemic cell as seen in our case. Liver biopsy can be helpful. Pathologically, the liver shows diffuse enlargement secondary to infiltration by leukemic lymphoblasts. In ALL and chronic leukemoid leukemia (CLL), the involvement of periportal spaces by neoplastic cells is common.² Systemic chemotherapy is the treatment of choice for hepatic infiltration with hepatic dysfunction and cytogenetic analysis is important to predict outcome and selection of therapy in ALL. Treatment should be initiated promptly to avoid fatal outcome.² Our patient had an atypical presentation of ALL. She presented with loss of weight and appetite and abdominal distension with normal blood parameters.

Compliance with ethical standards

Funding: None

Conflict of Interest: None

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