Case Report

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Portal Hypertension in Langerhans Cell Histiocytosis

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Abstract

Langerhans Cell Histiocytosis is a rare non malignant disease characterized by a clonal proliferation of pathologic cells with the characteristic Langerhans cells in single or multiple sites and an unpredictable course. The clinical presentation is heterogenous ranging from single system involvement to a multisystem life threatening disease. We report a case of LCH in 2 yr old female child with clinical signs of portal hypertension and hepatic involvement .

Keywords: Langerhans Cell Histiocytosis; Portal Hypertension; Hemetemesis.

Introduction

Langherhans cell histiocytosis (LCH) formally called Histiocytosis X is one group of poorly understood disease of histiocytes. The clinical spectrum of disease ranges from the chronic localized form to an acute leukemia like disease with fatal outcome. LCH typically occurs in children and adolescents, but can develop in all age groups with male predominance. We present a case of LCH in a female child with liver involvement and Portal hypertension that could be diagnosed using radiological imaging and doppler studies .

Case Report

A 2 year old girl child diagnosed to have LCH at the age of 6 months, presented in our casualty with 3 episode of hematemesis. She had no other bleeding manifestations. Her symptoms started at 6months of age with seborrheic skin lesions over neck, back and chest, managed as seborrheic dermatitis and improved with treatment . Later at the age of 8 months, she had new lesions of similar pattern which was evaluated in detail by skin biopsy , skeletal survey and bone marrow (BM) aspiration which demonstrate CDIa positivity. She was started on regular chemotherapy with daily prednisolone plus weekly vinblastine.

On physical examination child was sick looking, tachypoenic with grossly distended abdomen . Her head to foot examination revealed pallor, icterus and angular stomatitis. On systemic examination had abdominal distension, hepatomegaly with liver span of 11cms and massive splenomegaly. Shifting dullness present. Other systems were clinically normal.

Her lab investigations showed Hemoglobin of 6.0 gm/dl with microcytic hypochromic picture on peripheral smear. Her total serum bilirubin was 8.9mg/dl , with direct bilirubin 6.3mg/dl. Alkaline phosphatase was 783 IU with mild elevation in transaminases. Prothrombin time was also prolonged. Viral markers for Hepatitis was negative. Urine analysis was normal. USG abdomen showed coarse nodular echotexture with enlargement of liver predominantly left lobe, enlarged spleen of 12 cm with moderate ascites. X ray skull shows lytic lesion over frontal and parietal bones. Chest x-ray was normal.

Portal vein doppler was done which showed 16 cm/s flow with 13 mm hg pressure,suggestive of portal hypertension. Child was admitted to our Pediatric ICU and received transfusion of FFP/PRBC. Therapeutic ascitic tap done and drained 1 litre of fluid. Betablockers were added to lower the portal pressure . Child improved , discharged and is now on follow up.

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Discussion

Langerhans cell histiocytosis (LCH) is a group of idiopathic disorders characterized by the presence of cells with characteristics similar to bone marrow–derived Langerhans cells juxtaposed against a backdrop of hematopoietic cells, including T-cells, macrophages, and eosinophils.The definite diagnosis of LCH is based on cytology or histology in combination with immunohistochemical tests for CD1a and S100 protein expression.

The natural history of liver LCH fits into two stages: early infiltration by histiocytes and late sclerosis of the biliary tree [1]. Clinical diagnosis mainly depends on a high degree of suspicion, as most patients who have liver involvement may have disease at other sites also. Multi-organ LCH is more common, but localized LCH forms can also occur and it can even be the first manifestation [2].

Liver involvement in children LCH typically presents with hepatomegaly, abnormal liver enzymes, or jaundice, associated with multiorgan involvement. As mentioned above, our patient also has hepatomegaly with liver dysfunction. Hepatomegaly is a key sign of liver involvement and, along with other hepatic signs and symptoms, indicates the need for further investigations . Generally, hepatomegaly was due to the direct infiltration of LCH [1,3]. The progression of damage may continue despite the regression of the LCH, and after an initial injury the fibrotic disease process can be self-perpetuating even without the presence of Langerhans cells . Although the hepatomegaly usually regresses and is not predictive of the more severe liver involvement, children with hepatomegaly is younger and has a higher overall mortality.

Unfortunately, hepatomegaly in children is a common and nonspecific clinical finding, it may also be due to Kupfer cell hypertrophy and hyperplasia secondary to a generalized immune reaction or by enlarged portal lymph nodes causing obstruction. Moreover, it is important to bear in mind that hepatomegaly and steatosis may occur as adverse effects of systemic chemotherapy for Langerhans cell histiocytosis.Liver involvement may be severe to produce oedema and ascites . Hepatic involvement may lead to prolonged prothrombin time. Inflammation and scarring of the bile ducts will lead to sclerosing cholangitis causing portal hypertension [4].

The liver lesions can be due to direct or indirect effects of LCH. Direct liver involvement by LCH are in two forms: first, one is a portal tract inflammation without LCH infiltration, due to cytokines and the second one is the infiltration of the portal tracts and bile ducts with LCH. Laboratory findings in such cases are of cholestasis which reflects the damage of large or medium sized bile ducts [3]. Indirect effects on the liver of LCH elsewhere in the body are mediated through an accompanying macrophage activation syndrome that is most likely responsible for hepatomegaly, splenomeglay and hypoalbuminemia but without direct infiltration. These indirect effects are completely reversible [4]. In these patients, the disease is usually chronic and progressive and the liver biopsy may demonstrate infiltration of the portal area and basement membrane of the bile ducts with LCH cells [3].

Liver involvement in LCH is not uncommon (from 19% to 60% of cases) and it bears a poor prognosis [5,6]. In children less than five years old, liver involvement is relatively frequent, even though it is often overlooked.

A number of studies have reported high mortality (30–50%) when high-risk organs including the liver, spleen, lungs and hematopoietic system are involved, compared with the situation when these areas are not involved (<10%). The 3-year survival rate with liver involvement is 51.8%, compared with that of 96.7% without liver involvement [7].

Only very few cases of portal hypertension in LCH were reported from our country [8]. Therefore in a child with LCH with hepatomegaly one has to keep in mind the possibility of portal hypertension and plan treatment accordingly.

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