CASE REPORT

UNUSUAL PRESENTATION OF AN UNUSUAL HEPATITIS

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ABSTRACT

Autoimmune hepatitis is an uncommon cause of chronic hepatitis in Pakistan. One such case is described occurring in an adult female cirrhotic diagnosed on raised serum IgG and LKM-I.

Key words: Autoimmune hepatitis, immunoglobulin G, LKM-I.

INTRODUCTION

Chronic hepatitis is common in Pakistan and most frequent causes are various viral hepatitis[1-3], while occasionally other causes like alcohol, Wilson's disease and, hemochromatosis are also seen. Autoimmune hepatitis (AIH) is an uncommon disease in Pakistan. It is characterized histologically by interface hepatitis, and serologically by the presence of non-organ and liver specific autoantibodies and increased levels of immunoglobulin G. Its onset is often ill-defined, frequently mimicking acute hepatitis[4]. An interesting case of AIH with unusual presentation is being presented in this report.

CASE REPORT

A 45 years old female presented through the out patient department of Civil Hospital Karachi with complaints of abdominal distension for 1 month and swelling over both feet since 15 days. Abdominal distension was gradual in onset, not associated with any abdominal discomfort or abdominal pain. Then 15 days back, patient developed swelling over ankle region that gradually increased with no associated shortness of breath, orthopnea, or paroxysmal nocturnal dyspnea. She had pruritis that was worse in the day time and more marked on limbs for six months. She gave history of hepatitis D & E previously, with two pints of blood transfusion 2 years back and jaundice 6 months back that resolved after 20 days.

Physical examination showed the areas of hypo-pigmentation over both shins. Scratch marks were present on her limbs. She was vitally stable, mildly anemic. Pedal edema was present bilaterally. Abdominal examination showed distended abdomen with everted umbilicus, liver span of 10 cm, spleen palpable by about two and half fingers below left costal margin and shifting dullness was positive.

Laboratory studies showed normochromic normocytic anemia with normal electrolytes, blood urea, creatinine. Bilirubin was 3.3 mg/dl; serum ALT was 116 U/L; alkaline phosphatase was 290 U/L. Prothrmobin time was 25/15 sec with INR of 1.97 and serum albumin of 1.97g/dL. HBsAg, anti-HBc (total), anti-HCV, anti-HEV were negative while anti-HAV (IgG) was positive. Ascitic fluid report showed total proteins of 490 mg/dl and SAAG was 1.83. Ultrasound of whole abdomen showed liver being normal in size with irregular border, increased echogencity and coarse parenchyma. Portal vein diameter was 1.2 cm. Spleen was enlarged measuring 17.7 cm with massive amount of free fluid present in abdomen. Upper GI endoscopy was normal. Working diagnosis of cirrhosis was made and search was started to find the cause.

Her serum ferritin, serum iron, TIBC, and serum ceruloplasmin were normal thus excluding

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Received November 21, 2006; accepted: January 29, 2007.

hemochromatosis and Wilson's disease. Her IgG was increased raising suspicion of autoimmune haepatitis. ANA, ASMA, AMA were negative but anti-LKM-1 was strongly positive. Liver Histopathology showed porto-portal fibrosis with inflammation and presence of lymphocytes mostly limited to portal and periportal areas, but some spillage was noted in hepatic lobules too. Score of the patient according to the scoring system for diagnosis of autoimmune hepatitis proposed by International Autoimmune Hepatitis Group was 17, which made it a definite case of AIH[5].

Final diagnosis was therefore cirrhosis due to Type 2 autoimmune hepatitis.

DISCUSSION

Autoimmune hepatitis is a generally progressive, chronic hepatitis of unknown cause that occurs in children and adults of all ages. Occassionally, it has a fluctuating course, with periods of increased or decreased activity. It is characterized by the presence of interface hepatitis and portal plasma cell infiltration on histologic examination, hypergammaglobulinemia, and autoantibodies[4-7]. Women are affected more than men (gender ratio, 3.4:1), and all ages and ethnic groups are susceptible[8, 9]. It remains important to distinguish autoimmune hepatitis from other forms of chronic hepatitis, because a high percentage of cases respond to anti-inflammatory or immunosuppressive therapy, or both.

The presentation of autoimmune hepatitis is heterogeneous, and the clinical course may be characterized by periods of decreased or increased activity; thus, clinical manifestations are variable. The spectrum of presentation ranges from no symptoms to debilitating symptoms and even fulminant hepatic failure[10-13]. But presentation with cirrhosis is unusual and our patient presented with this unusual presentation. Laboratory abnormalities usually include aminotransferase elevations more striking than abnormalities in bilirubin and alkaline phosphatase levels. Also there is a generalized elevation of serum globulins, in particular, gamma globulin and IgG, which are generally 1.2 to 3.0 times normal[14]. The characteristic circulating autoantibodies include ANA, smooth-muscle antibody, antiactin antibody, SLA/LP autoantibodies, pANCA, anti-LKM-1, and anti-LC-1.14.

Three types of AIH have been proposed based on differences in their immunoserologic markers[4]. They do not have distinctive etiologies or responses to corticosteroid therapy[4].

Type 1 AIH is the most common form of the disease worldwide, and it is associated with ANA and/or SMA. It affects all age groups. They have a higher frequency of treatment failure, relapse after drug withdrawal, and requirement for liver transplantation than patients with other alleles[4]. Type 2 AIH is characterized by anti-LKM-1. A distinct form of LKM- positive AIH has been recognized in association with autoimmune polyendocrinopathy-candidiasis-ectodermal dystrophy(APECED). It is characterized by ectodermal dystrophy, mucocutaneous candidiasis, multiple endocrine gland failure (parathyroids, adrenals, ovaries), autoantibody production, and AIH in various syndromatic combinations[4]. Type 3 AIH is the least established form of the disease, and it is characterized by the presence of anti-SLA/LP in serum. Patients have clinical and laboratory features that are indistinguishable from patients with type 1 AIH, and they respond well to corticosteroids[4].

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