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ACUTE LIVER FAILURE AS INITIAL PRESENTATION OF AUTOIMMUNE HEPATITIS IN AN ELDERLY MALE.

General Medicine

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ABSTRACT

Autoimmune hepatitis is a chronic disorder characterized by continuing hepatocellular necrosis and inflammation, usually with fibrosis, which can progress to cirrhosis and liver failure. Male: female ratio is 1:4. It may present as acute or chronic hepatitis and rarely as fulminant hepatic failure.
A case of 66-year-old male who presented with jaundice, anorexia, nausea, distension of abdomen since 1month. On evaluation, the liver function test was suggestive of severe conjugated hyperbilirubinemia (D. Bilirubin 27mg/dl) with more than 60 times the normal aminotransferase levels(>2400U/L), normal alkaline phosphatase, with coagulopathy (INR 3.1) without any deranged complete blood counts. Autoimmune evaluation turned out significantly positive.

KEYWORDS

Autoimmune hepatitis in males , Acute liver failure , cirrhosis of liver .

INTRODUCTION –

Autoimmune hepatitis is a chronic disorder characterized by continuing hepatocellular necrosis and inflammation, usually with fibrosis, which can progress to cirrhosis and liver failure. It may present as either an acute or chronic disease with a fluctuating pattern, the spectrum of presentation also includes asymptomatic patients. Those patients who present with acute liver failure, jaundice, and coagulopathy, such a presentation is generally uncommon [1,2].

Autoimmune hepatitis can present at any age and in all ethnic groups and occurs predominantly in women. For type 1 autoimmune hepatitis, the female to male ratio is 4:1, but for type 2 autoimmune hepatitis, the ratio is 10:1[3]

CASE REPORT –

A 66-year-old male who presented to our hospital with complaints of jaundice, anorexia, nausea, distension of abdomen since one and half month, which was gradually progressive in nature and aggravated in the past 1 week. He was a known case of type 2 diabetes mellitus and hypertension for past six years, well controlled on oral hypoglycemics and anti hypertensives without any complications. There was no other significant drug history, he was neither an alcoholic nor did he have other addictions. On routine evaluation, the liver function test was suggestive of severe conjugated hyperbilirubinemia (D. Bilirubin 27mg/dl) with more than 60 times the normal aminotransferase levels(SGOT >2400U/L and SGPT >1137), normal alkaline phosphatase with coagulopathy (INR 3.1) complete blood counts showing hemoglobin of 12 g/dl ,WBC 8000/cumm and platelets 1,20,000 .CT abdomen showed hepatomegaly with increased echotexture, normal surface and biliary canaliculi, mild splenomegaly with moderate ascites, rest of the study was normal. Wilsons disease and infective etiology of fulminant hepatitis was ruled out.

Ascitic tap was transudative in nature, patient was suspected of autoimmune hepatitis and further evaluated, he turned out positive significantly for Anti-Nuclear Antibody, Anti Smooth Muscle Antibody along with hypergammaglobulinemia (1748 mg/dl), was diagnosed as Type 1 autoimmune hepatitis. Treatment was initiated with 60mg Prednisone corticosteroids, but within 8 days of presentation, he developed hepatorenal and hepatopulmonary syndrome and succumbed to the disease.

DISCUSSION –

The presentation of acute autoimmune hepatitis can be either acute worsening of previously undiagnosed or misdiagnosed cases [4]. Typically affects young and middle-aged females.

Type 1/classic autoimmune hepatitis, is characterized by circulating antinuclear antibodies and/or anti smooth muscle antibodies, anti SLA/LP antibodies. It is the main form of autoimmune hepatitis seen in adults and adolescents.

Type 2 autoimmune hepatitis is defined by the presence of antibodies to liver/kidney microsomes (ALKM-1) and/or to a liver cytosol antigen (ALC-1). It is typically diagnosed in infants, girls, and young women.

Autoantibody negative autoimmune hepatitis, approximately 20 percent of patients who present with all the features of autoimmune hepatitis lack circulating ANA, ASMA, or anti-LKM-1 antibodies.

In this patient it's a rare presentation of acute fulminant liver failure, in an elderly male patient with type 1 autoimmune hepatitis.

Treatment is initiated in patients who fulfill any of the following criteria:(1) Serum aminotransferase levels greater than 10-fold the upper limit of normal. (2). Serum gamma globulin level greater than twice the upper limit of normal. (3). Serum aminotransferase levels greater than twice the upper limit of normal, along with: Symptoms, An elevated gamma globulin level, even if less than twice the upper limit of normal, An elevated conjugated bilirubin level, Interface hepatitis on biopsy. (4) Histologic features of bridging necrosis or multiacinar necrosis. (5) Cirrhosis with any degree of inflammation on biopsy. (6) Children.

Treatment failure is characterized by: Sustained biochemical and histologic activity leading to the development or worsening of cirrhosis with eventual complications and death, the need for liver transplantation

CONCLUSION –

- AIH should be considered a possibility in any individual with acute or chronic hepatitis of unknown origin.
- Screening for concomitant autoimmune diseases to be considered in patients with AIH
- AIH with cholestatic features should be tested for primary biliary cholangitis
- Patients with severe disease like acute liver failure due to autoimmune hepatitis have a high short-term mortality rate.
- If they fail to show normalization of at least 1 lab parameter after starting prednisolone-based therapy or if pre-treatment hyperbilirubinemia fails to improve during 2-week treatment trial. Early liver transplantation should be considered in such individuals.

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