Abstract:
Autoimmune hepatitis is a disorder of unknown etiology characterized by unresolving inflammation of liver, interface hepatitis on histology, hypergammaglobulinemia, and presence of auto antibodies, strong association with other autoimmune diseases. It occurs most often in women, in second and third decades of life, may develop in either sex at any age. We present a case of 40 year old woman presented to our hospital with jaundice since one month on examination patient had jaundice, pedal edema and ascites. Investigations revealed highly elevated bilirubin and liver enzymes, serology negative for A,B,C, E with marked IgG elevation. Ultrasound abdomen showed features suggestive of cirrhosis with ascites liver biopsy and analysis of auto antibodies showing high titers suggestive of auto immune hepatitis. Patient started with steroid therapy resulting in improvement of patient condition. Elevated LFT with viral serology negative should always be investigated. Early diagnosis and treatment of autoimmune hepatitis is essential to improve prognosis. A patient of autoimmune hepatitis presenting directly in chronic decompensated cirrhotic state is very rare which happened with our case. Any autoantibody positivity should be carefully considered to avoid misdiagnosis delaying appropriate clinical management.

Key words: Autoimmune hepatitis; Cirrhosis; Steroids

Introduction
Autoimmune hepatitis is a disorder of unknown etiology characterized by unresolving inflammation of liver, interface hepatitis on histology, hypergammaglobulinemia, and presence of autoantibodies [1]. Onset is frequently insidious with nonspecific symptoms such as fatigue, nausea, abdominal pain and arthralgias at presentation, but the spectrum is wide ranging from asymptomatic presentation to an acute severe disease. Women are affected more frequently than men and the disease is seen in all ethnic groups. Incidence of autoimmune
hepatitis is 2 cases/lakh persons per year where as its point prevalence is 16.9 cases per 1,00,000 persons per year [2].

**Case Report**

A 40 year female G2P2 with 2 healthy male children presented with h/o yellowish discoloration of eyes and urine from one month and abdominal distension of 25 days along with history of easy fatiguability. No significant past history or transfusion history. Not a known smoker and alcoholic. Clinical examination showed no other findings except for icterus and abdominal distension. The case was investigated the findings were, highly elevated liver enzymes and bilirubin. The laboratory examination showed a negative serology for hepatitis B,C. Ultrasound abdomen showed cirrhosis of liver with moderate ascites and mild splenomegaly. Hepatitis A and E infections were ruled out. Upper GI endoscopy showed grade 1 esophageal varices. Analysis of autoimmune antibodies were done, which showed high titres of antinuclear antibodies (strong positive) and negative anti LKM1 antibodies. Serum copper and ceruloplasmin were within normal limits. Another important finding in this case was, a marked IgG elevation and hypergammaglobulinemia. Liver biopsy was performed showing features compatible with autoimmune hepatitis. Patient was started on steroids of 1mg/kg. Patient started showing improvement within 2 weeks.

**Investigations**

**Liver Function Tests:**

<table>
<thead>
<tr>
<th>INVESTIGATION</th>
<th>AT PRESENTATION</th>
<th>AFTER TREATMENT</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total bilirubin</td>
<td>17.4mg/dl</td>
<td>6.9mg/dl</td>
</tr>
<tr>
<td>Direct bilirubin</td>
<td>15.8mg/dl</td>
<td>5mg/dl</td>
</tr>
<tr>
<td>SGPT</td>
<td>388U/L</td>
<td>77u/l</td>
</tr>
<tr>
<td>SGOT</td>
<td>658U/L</td>
<td>66u/l</td>
</tr>
<tr>
<td>ALP</td>
<td>189U/L</td>
<td>116u/l</td>
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After 2 months of treatment bilirubin and liver enzymes dropped to baseline.

Anti nuclear antibodies - 69.55 units (strong positive) anti LKM1 antibodies – Negative
**Discussion:**

Autoimmune hepatitis is a disorder of unknown etiology. To diagnose it as autoimmune hepatitis other similar diseases should be excluded and there should be laboratory findings of immunoreactivity and interface hepatitis on histology. Most of the times autoimmune hepatitis have acute severe or fulminant presentation. Similarly manifestations of liver decompensation, such as ascites and variceal bleeding are uncommon at initial medical presentation which happened with our case.

Autoimmune hepatitis is classified into two subgroups. Type 1 autoimmune hepatitis affects young girls and middle aged women and may occur in elderly also and associated with antinuclear antibodies or smooth muscle antibodies or both [3]. Type 2 is disease of children and associated with antiLKM1 antibodies [4]. Autoimmune hepatitis in patient who also have antimitochondrial antibodies and histologic features of cholangitis constitute an overlap syndrome with primary biliary cirrhosis [5-8]. Autoimmune hepatitis is considered to be a steroid responsive disease. Appropriate management can improve quality of life, prolong survival and delay the need for liver transplantation. Glucocorticoid therapy (prednisolone 40mg /day orally) is continued until remission, treatment failure, incomplete response, or drug toxicity occurs [9]. Maintenance therapy is required for at least 2 years after LFTs have returned to normal, and withdrawal of treatment should not be considered unless a liver biopsy is also normal. Most individuals require long-term immunosuppression. Azathioprine can also be used as the sole maintenance immunosuppressive agent. Corticosteroids treat acute exacerbations but to not prevent cirrhosis; they are therefore less important in mild asymptomatic autoimmune hepatitis. Alternative treatment strategies with immunosuppressants like azathioprine, cyclosporine A tacrolimus mycophenolate mofetil, cyclophosphamide should be considered if standard treatment fails [10]. For patients refractory to medical treatment or do not reach remission within 4 years of continuous therapy, liver transplantation remains the treatment option of choice.

Autoimmune hepatitis is characterized by exacerbations and remissions, most patients develop cirrhosis and its complications. Hepatocellular carcinoma is a rare complication. About 50% of autoimmune hepatitis patients die of liver failure in five years if not treated, with treatment mortality falls to 10%.

**Conclusion:**

A patient of autoimmune hepatitis presenting directly in chronic decompensated cirrhotic state is very rare which happened with our case. Elevated LFT with viral serology negative should always be investigated. Any autoantibody positivity should be carefully considered to avoid misdiagnosis delaying appropriate clinical management. Early diagnosis and treatment of autoimmune hepatitis is essential to improve prognosis.

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**References:**

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Norwegian population. Scand J Gastroenterol 1998; 33:99-103