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Medicine

ASSOCIATION OF AUTOIMMUNE HEPATITIS WITH RENAL TUBULAR ACIDOSIS

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ABSTRACT Distal Renal Tubular Acidosis(Distal RTA) or Type 1 RTA is a classic form of renal tubular acidosis(RTA), which is commonly associated with autoimmune diseases like Sjogren's syndrome, systemic lupus erythematous, rheumatoid arthritis, hypergammaglobulinemia and autoimmune liver disease. But usually it is relatively latent not causes significant abnormalities. Here we are describing a case of recurrent Hypokalemic periodic paralysis which is diagnosed as autoimmune hepatitis with distal RTA.

KEYWORDS :

INTRODUCTION:

Autoimmune liver disease and renal tubular acidosis (RTA) are frequently associated, possibly related to their common autoimmune pathogenesis.[1] In such cases of autoimmune hepatitis (AIH), diagnosis of RTA is usually done by inability of the kidney to handle acid load.

CASE REPORT:

A 22 year old female patient presented with complaint of generalized weakness ,easy fatigablity, abdominal distension, yellowish discoloration of eyes and urine, pedal edema since 20 days.

PAST HISTORY :

Patient had history of recurrent episodes of hypokalemic periodic paralysis since 2010 and diagnosed as renal tubular acidosis .

GENERAL EXAMINATION:

On Examination had pallor, icterus, pedal edema, no cyanosis, clubbing, lymphadenopathy. vitals are stable.

SYSTEMIC EXAMINATION:

On per abdomen examination ,abdomen distended, umblicus everted, engorged veins are visible over the abdomen. On palpation splenomegaly present. on percussion shifting dullness present.

INVESTIGATIONS :

HB:8.7mg/dl,WBC:3500cells/cumm,PLT:79000/cumm,BILIRUBIN :3.6mg/dl,SGPT:14u/lt,SGOT:29u/lt,ALP:86u/lt. Total proteins3.2gm/dl.

SR. Electrolytes:sodium:133mmol/l, potassium:3.8mmol/l, chloride:100mmol/l.

Thyroid profile normal,PT:23sec, INR:1.46.

ANA Screening positive, Anti LKM antibodies positive.

On ultrasonography altered echogenicity of liver with irregular surfaces suggestive of cirrhosis of liver.

DISCUSSION

Distal RTA or type 1 RTA is the classical form of RTA characterized by a failure of acid secretion by the alpha intercalated cells of the cortical collecting duct leading to inability to acidify the urine which may be hereditary or may be triggered by an autoimmune disorder. Distal RTA is commonly associated with autoimmune diseases like Sjogren's syndrome, systemic lupus erythematous, rheumatoid arthritis, hypergammaglobulinemia and autoimmune liver disease. Previous study found coexistence of RTA in 60% of the patients with primary biliary cirrhosis and in 30% with AIH. In such cases RTA is frequently latent.

AIH is of three types. Type 1 is smooth muscle antibody (SMA) and/or antinuclear antibody (ANA) positive, and type 2 is positive for antibodies to liver-kidney microsome type 1 (anti LKM1) with 80% female predominance in both.[5] Type 2 AIH usually presents more acutely and possible in association with autoimmune disorders, family history of autoimmunity, and IgA deficiency. As in our case, a young girl had two features which possibly suggest type 2 AIH; (1) acute onset of symptoms and (2) prior history of RTA possibly of autoimmune origin.

Treatment with steroids and azathioprine has response in 80% of cases, however, relapses are also common. She was treated with prednisolone (2 mg/kg) and azathioprine (1 mg/kg) and her symptoms improved over 2 months.

CONCLUSION :

Hypokalemic periodic paralysis is a common manifestation of distal RTA, which is frequently associated with an Autoimmune hepatitis. So in case of hypokalemic periodic paralysis if associated with liver cell failure signs thought of Autoimmune hepatitis.

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