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CASE REPORT

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Role of MRI in diagnosis of an unusual case of celiac disease presenting as hepatomegaly

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General Note
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ABSTRACT

Celiac disease is a gluten sensitive enteropathy usually presenting with gastrointestinal symptoms, but unusual presentations make the diagnosis of this disease sometimes challenging. Nodular regenerative hyperplasia (NRH) is a rare condition associated with connective tissue disorders, autoimmune diseases, haematological malignancy, drugs and is a cause of non cirrhotic portal hypertension. It is characterized by multiple hepatic nodules in the absence of fibrosis. The association of celiac disease with

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nodular regenerative hyperplasia had been reported rarely in literature. We describe a case of 10 year old female child who presented as hepatomegaly with portal hypertension and was diagnosed as celiac disease with nodular regenerative hyperplasia with the help of magnetic resonance imaging (MRI) abdomen. The diagnosis was confirmed by celiac serology and duodenal biopsy. MRI abdomen has sensitivity and specificity of 70 to 80 % in detecting NRH. Increased ileal folds and reversed fold patterns are specific findings for celiac disease in MRI abdomen. We also reviewed the etiopathogenesis, clinical features and diagnosis of NRH and discuss the significance of abdominal imaging especially MRI in establishing the diagnosis of celiac disease.

Key words: Celiac disease; MRI; nodular regenerative hyperplasia; non cirrhotic portal hypertension

1. INTRODUCTION

Celiac disease or gluten sensitive enteropathy is a chronic disease that affects both children and adults.[1,2] Although most patients have gastrointestinal symptoms, a wide range of extraintestinal manifestations have been described involving skin, nervous system, liver and blood.[2,3] Because of their lack of specificity, diagnosis of celiac disease sometimes is challenging. Nodular regenerative hyperplasia of liver (NRH) is an uncommon liver disease and is a cause of non cirrhotic portal hypertension.[4-7] Some case reports have described an association of NRH with celiac disease. Most patients of NRH are asymptomatic and routine radiological investigations are difficult to differentiate NRH from other nodules.[4,7] We describe here an unusual case of short stature with massive hepatomegaly who was eventually diagnosed as celiac disease with NRH and discuss the role of MRI in establishing the diagnosis of celiac disease.

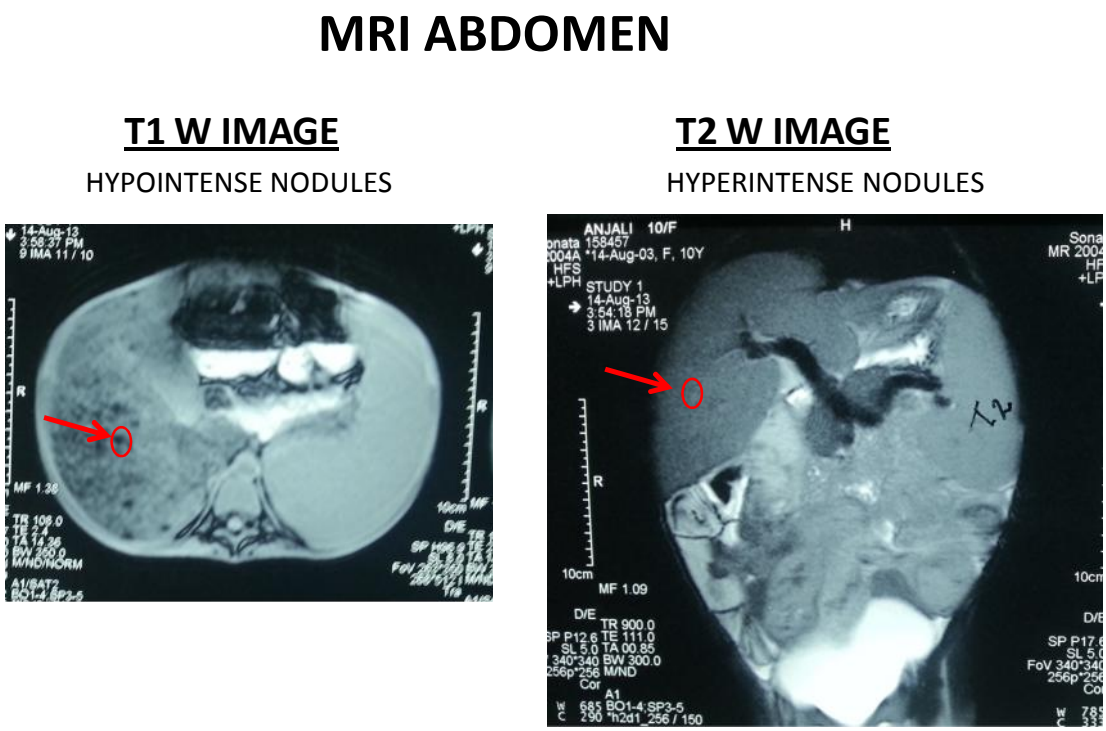

Figure 1A T1 and T2 weighted MRI images of abdomen

2. CASE REPORT

A 10 year old female child presented with complaints of abdominal distension and loss of weight and loss of appetite for the past 5 years. No history of jaundice, bleeding manifestations, altered bowel habits and family history of liver disease. Anthropometry

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revealed a weight of 16kg (<-3SD) and height of 114 cm (<-3SD). Examination revealed pallor, hepatomegaly (liver 7cm below costal margin with a span of 15cm), splenomegaly (4cm below costal margin) and ascites. Clinically, a provisional diagnosis of chronic liver disease with portal hypertension was made and child was investigated. Investigations revealed haemoglobin of 4.6 gm/dl, total leukocyte count of 2000/mm3 and peripheral smear revealed macrocytic hypochromic anaemia with leucopenia. Liver enzymes (SGOT-57U/l, SGPT-16U//L and ALP-144U/L), total bilirubin (0.4 mg/dl), total protein (6.5 gm/dl), albumin (2.2 gm/dl) and renal function tests were within normal limits. Prothrombin time was 15 (control-13) and partial thromboplastin time was 36 (control-29). Antibody testing for HIV, Hepatits B infection and HCV were negative. Serum Ceruloplasmin was 28mg/dl.

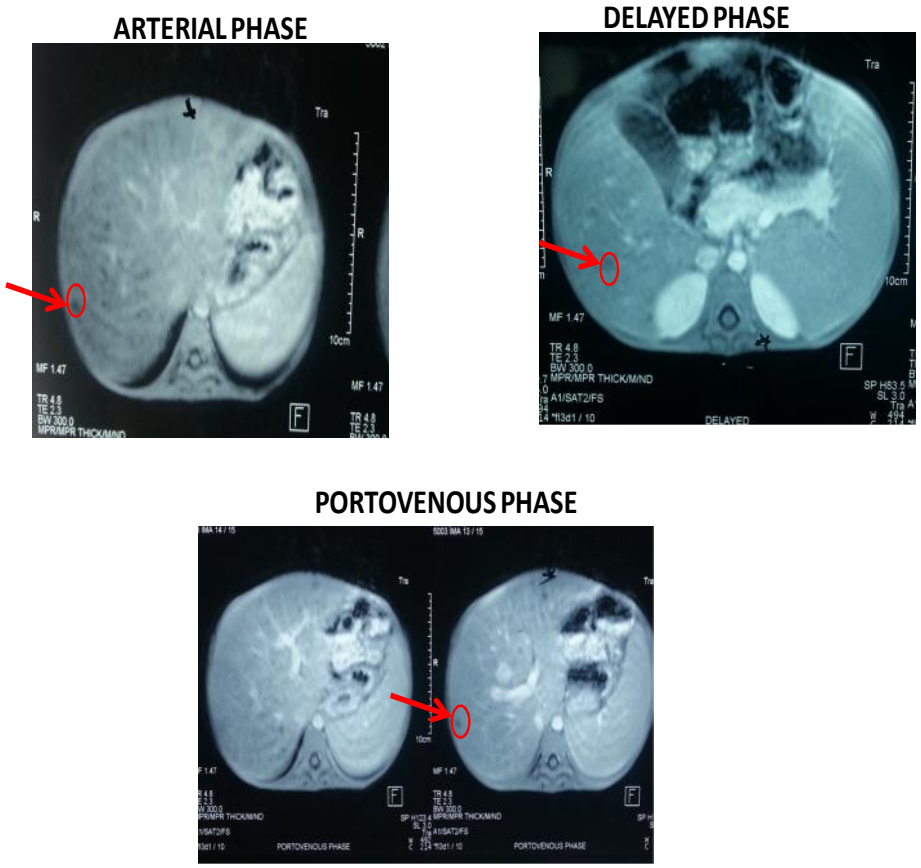

Figure 1B MRI images showing arterial and portovenous phase.

USG abdomen showed hepatosplenomegaly with coarsened liver echotexture and multiple ill defined hypoechoic lesions distributed in both lobes. Contrast enhanced CT abdomen revealed hepatosplenomegaly with non enhancing hypodense lesions diffusely distributed in both lobes of the liver and no post contrast enhancement. MRI abdomen was done to study the liver parenchyma in further detail. It revealed hepatosplenomegaly with multiple tiny nodules measuring less than 1cm, hyperintense on T1 and hypointense on T2 weighted images with no enhancement seen on the arterial phase suggestive of nodular regenerative hyperplasia. The superior mesenteric vein, splenic vein and the portal vein were dilated with the portal vein measuring 15mm at the porta. The bowel loops were dilated with fat contents within the lumen seen as hypertintense signal on T1 weighted images (fig.1A & 1B & 1C). With these findings celiac disease was suspected and levels of tissue transglutaminase antibody of IgA type in serum was determined, which was more than 800 U/l. The diagnosis of celiac disease was confirmed by duodenal biopsy which revealed complete flattening of villi with increase in the crypt villous ratio and increase in the number of intraepithelial lymphocytes [Marsh

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grade III (fig 2A & 2B)]. Child was started on gluten free diet and is under follow up. Antinuclear antibody and IgA anti cardiolipin antibody were negative. After 2 months of gluten free diet, child gained 4 kg (>10%) and liver span regressed by 1 cm.

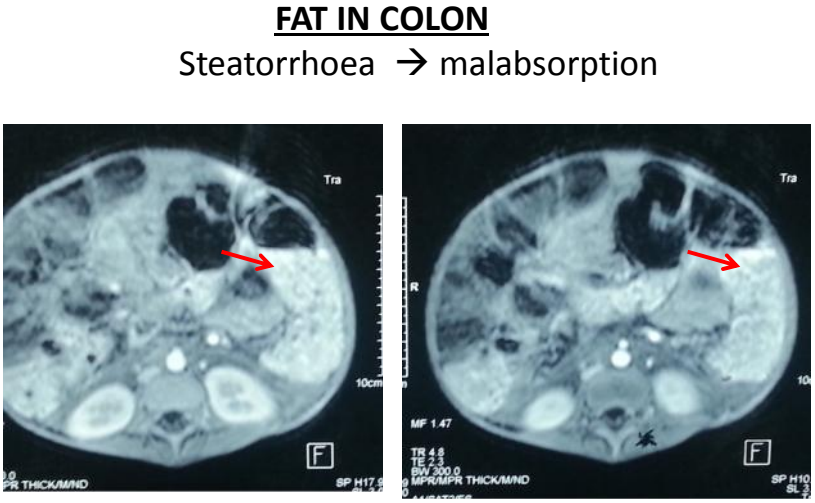

Figure 1C MRI showing intraluminal hyperintensity in T1 images

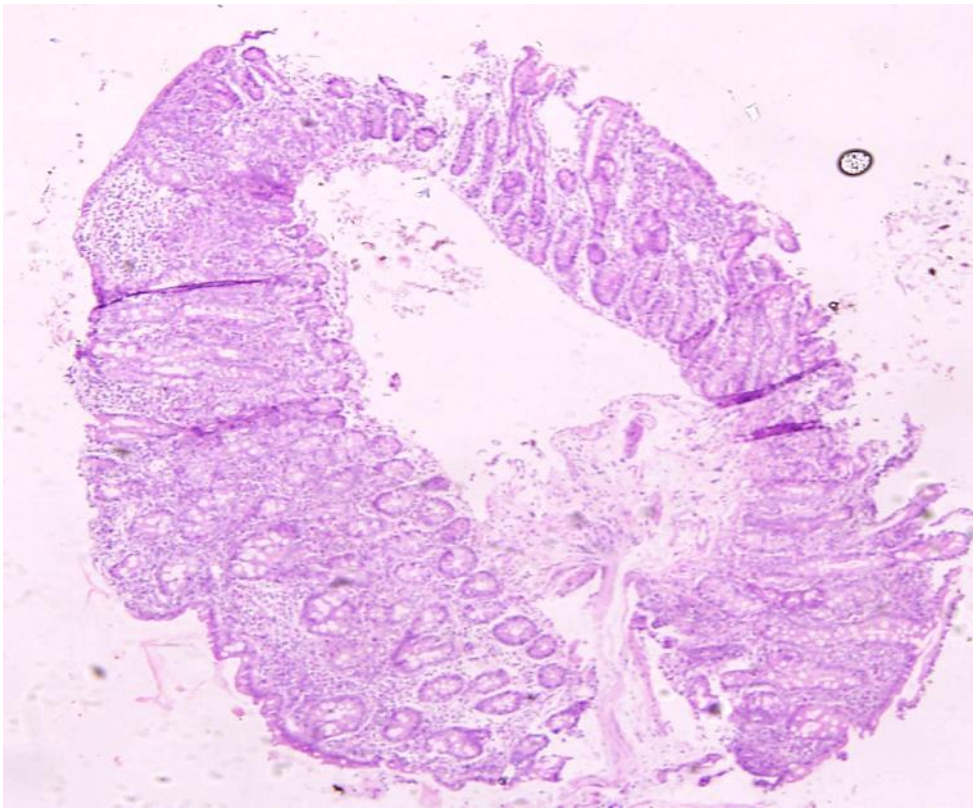

Figure 2A Dudenal mucosa with complete villous flattening, crypt hypertrohy (Haematoxylin & Eosin x 40)

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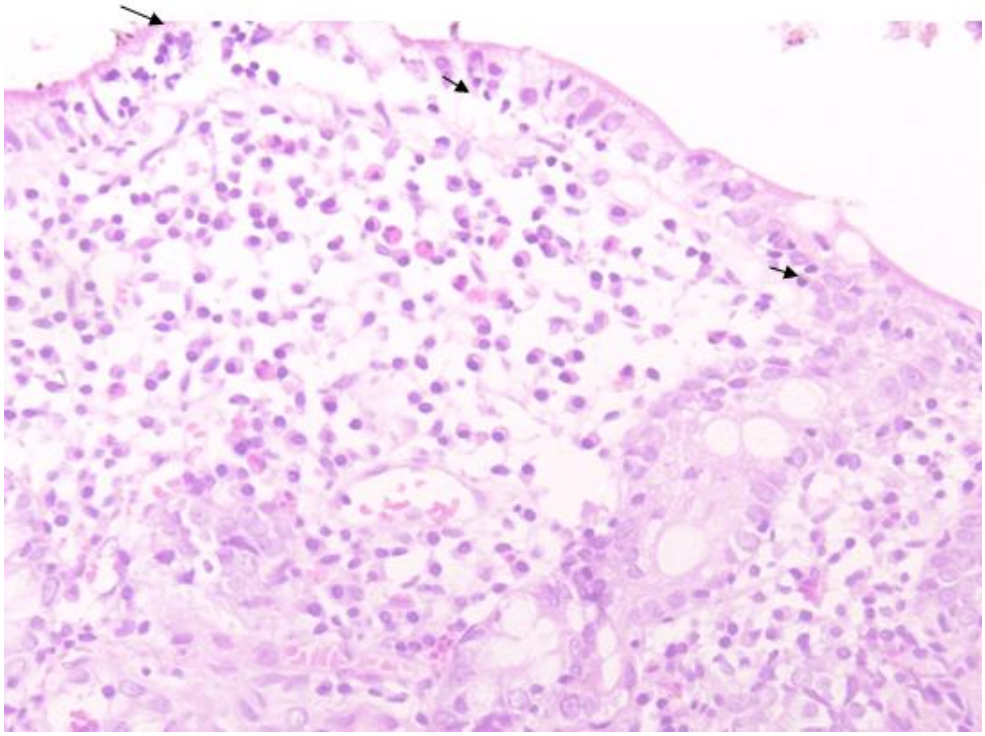

Figure 2B Prominent infiltrate of plasma cells with admixed eosinophils in lamina propria and increase number of intraepithelial lymphocytes(arrow) (Haematoxylin & Eosin x 400)

3. DISCUSSION

Though a wide spectrum of hepatobiliary diseases have been described including asymptomatic elevations of liver enzyme levels, nonspecific hepatitis, nonalcoholic fatty liver disease, and autoimmune and cholestatic liver disease,[3] NRH had been rarely described in literature especially in children with celiac disease.[4-9] Because most of these patients do not have overt gastrointestinal symptoms, a high index of suspicion is required. Nodular regenerative hyperplasia of the liver is an uncommon condition characterised by diffuse transformation of normal parenchyma into small regenerative nodules with little or no fibrosis.[4,5] It is often associated with connective tissue disorder, autoimmune diseases and haematological malignancy and is a cause of non cirrhotic portal hypertension.[4-8] Current hypothesis states that pathogenesis is based on vascular mechanism in which sinusoidal damage and/or obstruction of portal venules leads to ischemia causing atrophy of hepatocytes and subsequent regeneration in other areas.[4-7] NRH may be either asymptomatic or present with features of portal hypertension (ascites, varices & splenomegaly).[4,6] Laboratory parameters including serum aminotransferases, prothrombin time and serum bilirubin are usually normal.[4,6]

Routine radiological investigations like USG and CECT abdomen are unable to differentiate NRH from other nodular lesions.[4,8] MRI is superior to CT in diagnosing NRH,[4] other intestinal and extra intestinal features of celiac diseases,[10] because of its inherent propensity to differentiate soft tissue lesions. NRH lesion appears hyperintense on T1 weighted image and iso/hypo intense on T2 weighted images, with a sensitivity and specificity of 70%-80% when using gadolinium contrast.[4,8] MRI studies done by Tomei et al in 31 adults with celiac disease, showed bowel dilatation in 61.3% , increased number of ileal folds in 48.4%, reversed fold pattern abnormalities in 38.7%, increased wall thickness in 16.1%, duodenal stenosis in 6.5%, mesenteric lymphadenopathy in 41.9%, mesenteric vascular changes in 22.6%, ascites in 6.5% and no abnormalities in 12.9%.[10] MRI demonstrated a specificity and accuracy of 100%, and sensitivity of 79% and 75% for increased number of ileal folds and reversed fold pattern abnormalities respectively.[10] The paucity of literature suggests that there is insufficient awareness of potential role of abdominal MRI in celiac disease. In our case MRI findings of abdomen lead us to the diagnosis of celiac disease.

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Hence, in children with unexplained abdominal symptoms and inconclusive findings in USG and CT, MR imaging of abdomen has a potential role in establishing the diagnosis of celiac disease in unusual cases.

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