INTRODUCTION
Celiac Disease (CD) is a malabsorption syndrome that usually presents in childhood. It affects around 1% of the general population, and has intestinal and extra-intestinal manifestations. Some patients of CD become refractory to Gluten-Free Diet (GFD) with persistent malabsorption and intestinal villous atrophy. This subset of patients is considered to have Refractory Celiac Disease (RCD). Celiac crisis is a life-threatening complication of CD in which patients have profuse diarrhea and severe metabolic disturbances. The prevalence of RCD is 0.31% among diagnosed CD patients and 0.002% in the general population. Neurological involvement occurs in 8 - 10% of adults and less frequently in children. Herein we present a case of RCD in patients presenting in celiac crisis and showing no improvement on GFD.

CASE REPORT
A 13-year boy presented with complaints of fever, diarrhea and significant weight loss of 20 kg during two and half months. Diarrhea was associated with food intake. He had a history of nystagmus since birth and generalized seizures one and half years back. He had no history of pruritus, abdominal distension or tuberculosis.

On examination, he was a young boy, conscious, oriented but emaciated with Body Mass Index (BMI) of 9.4 kg/m², dehydrated, with icterus and enlarged cervical and inguinal lymph nodes, largest one, 1 cm, on the right side of neck. Respiratory, abdominal and cardiovascular systems were unremarkable. The Central Nervous System (CNS) examination revealed horizontal nystagmus, non-fatigable, with no change with position. Motor examination revealed reduced power in all four limbs with normal tone and sensations.

On laboratory investigation, hemogram showed hemoglobin of 11.2 g/dL, Mean Corpuscular Volume (MCV) of 86 fL; total leukocyte count of 7,000/cmm; platelets at 265,000/cmm; and erythrocyte sedimentation rate of 112 mm after first hour. Biochemistry showed serum sodium, at 121 mEq/L; potassium, at 2.7 mEq/L; total proteins, at 8.5 g/dL; albumin, at 2.3 g/dL; creatinine, at 0.6 mg/dL. Liver Function Tests (LFTs) showed total bilirubin level of 9.07 mg/dL; direct bilirubin at 6.64 mg/dL; Alkaline Phosphatase (ALP) at 1003 IU/L; Gamma Glutamyl Transferase (GGT) at 732 IU/L; Alanine Aminotransferase (ALT) at 77 IU/L; and Aspartate Aminotransferase (AST) at 138 IU/L. Serum IgG was 41.3 g/L while IgA levels were decreased. His blood culture grew Methicillin-Resistant Staphylococcus aureus (MRSA) and was treated with intravenous (IV) vancomycin. Fine Needle Aspiration Cytology (FNAC) of cervical lymph nodes showed chronic, non-specific inflammation. Endoscopic biopsy of the duodenum and jejunum showed moderate to severe blunting of villi, crypt hyperplasia and increased Intraepithelial Lymphocytes (IELs) consistent with modified Marsh stage IIIb (Figure 1). Celiac serology showed negative anti-tissue transglutaminase (TG) IgA and IgG while Deamidated Gliadin Peptide (DGP) antibodies were strongly positive (45.05 IU/ml). His Human Leucocyte Antigen (HLA) DQ2 and DQ8 were negative but HLA DQ6 and DQ7 were positive. Because of the persistent diarrhea, sigmoidoscopy was performed.
done which revealed edematous mucosa of rectum, sigmoid and descending colon. Biopsy from these sites showed chronic non-specific inflammation. Patient was kept on parenteral nutrition and GFD but diarrhea was not settled. Later on, Computerized Tomography (CT) abdomen was done to rule out Vasoactive Intestinal peptide (VIP)-oma. It showed minimal ascites which was not tapable with few subcentimetric lymph nodes in the para-aortic region. After ruling out all causes of diarrhea, Immunohistochemistry (IHC) of the duodenal biopsies was requested which revealed positivity of IELs for both CD3 and CD8. The positivity of CD8 was noted in > 50% of IELs, suggestive of RCD, type 1 (Figure 1). Patient was started on high dose steroids, 1 mg/kg and showed marked improvement in diarrhea. He then presented in summer with profuse watery diarrhea with metabolic derangements requiring hospitalization and institution of parenteral nutrition. The precipitating factor was most probably MRSA infection.

This case is a rare presentation of celiac crisis with neurological manifestations as the first symptoms, presenting with a history of seizures and after evaluation found to have right occipital lobe encephalomalacia and ex-vacuo dilation of occipital horn of right lateral ventricle on Magnetic Resonance Imaging (MRI) brain. His Electroencephalogram (EEG) was also consistent with seizure disorder along with encephalopathy.

**REFERENCES**

7. Jamma S, Tapia AR, Kelly CP, Muray J, Sheth S, Schuppan D.

