Celiac disease presenting as acute unilateral blindness

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Abstract

Background: Celiac disease is known to present with extra gastrointestinal manifestations.

Case Characteristics: An eight years old male child presented with painless acute loss of vision in left eye.

Observation: MRI orbit was suggestive of demyelinating optic neuritis. Serology and duodenal biopsy confirmed celiac disease. Vision regained completely with steroids and gluten free diet.

Message: Acute loss of vision or blindness is an uncommon presentation of celiac disease.

Key words: Celiac disease, blindness, optic neuritis

Introduction: Celiac disease (CD) is an autoimmune condition affecting the small intestine, triggered by the ingestion of gluten, the protein fraction of wheat, barley, and rye. When patient with celiac disease ingest gluten, an immunologically mediated inflammatory response occurs that damages the mucosa of their intestines, resulting in malabsorption of food nutrients. Apart from gastrointestinal symptoms, celiac disease has extraintestinal symptoms like anemia, osteopenia, motor weakness, ataxia, seizures, peripheral neuropathy, dermatitis herpetiformis, bleeding diathesis and ophthalmic manifestations like uveitis, cataract, orbital myositis, retinopathy, occipital calcification and rarely optic neuropathy. We are reporting a case of celiac disease who presented as acute loss of vision of left eye.

Case report: An eight year old boy presented with painless acute loss of vision of left eye in July 2018 which was preceded by blurring of vision for few hours. There was no history of fever, rashes, vomiting, diarrhea, constipation, seizures or headache. On examination vitals were stable, weight: 18.1 Kg (5-10th centile), Height: 114 cm (5-10th centile). There was pallor on general examination. Systemic Examination was normal. He was referred to ophthalmologist and pediatric neurologist. Ophthalmologist's evaluation showed no abnormal finding. Paediatric neurologist advised MRI brain and Orbit. MRI showed Inflammatory changes in left preseptal orbital region extending into the post septal and retro bulbar region showing moderate contrast enhancement, left orbital optic nerve showed focal loss of perineural CSF signals, focal hyperintensity with focal acute neuritis changes (Fig. 1 A). CSF was advised but was refused by the parents. CRP was 1.2mg/L (0-5).

In view of anemia and poor growth following investigations were done: Hb 7.3 gm/dl, MCV: 55.5 fL, Total Leucocyte count 9100/µL; Platelets: 458000/µL, RDW: 23.3 %. Serum tissue transglutaminase IgA >300 unit/ml (Upper limit is 18unit/ml).UGI endoscopy was suggestive of scalloping of duodenal mucosal folds. Duodenal biopsy showed Marsh grade III b changes (Fig. 1 B)

He was put on strict gluten free diet. In view of acuity of symptoms and changes of demyelination he was given 3 doses of injection methyl prednisolone followed by oral prednisolone which was gradually tapered and stopped over next 4 weeks. The child regained

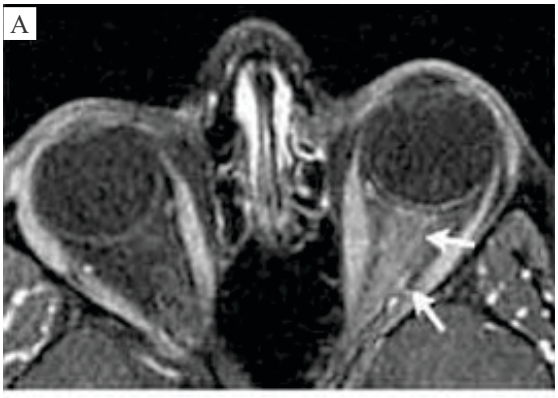
full vision in left eye after six days of treatment. On follow up till 6 months, his weight and height centiles have increased to 50%ile and there was no further episode of vision loss.

Discussion: Atypical forms of celiac disease without prominent gastrointestinal symptoms and with frequent extra-intestinal manifestations, are being increasingly recognized, especially over the past decade, both in adult and pediatric patients. The ophthalmic manifestations can be divided into autoimmune disorders and absorptive disabilities. The manifestations related to malnutrition are correlated to the low levels of vitamin A, Vitamin D, and calcium which could cause retinopathy, cataract, dry eye and pseudotumor cerebri. The manifestations related to autoimmune disorders are orbital myositis, uveitis, thyroiditis associated with orbitopathy and brain occipital calcification. [1]

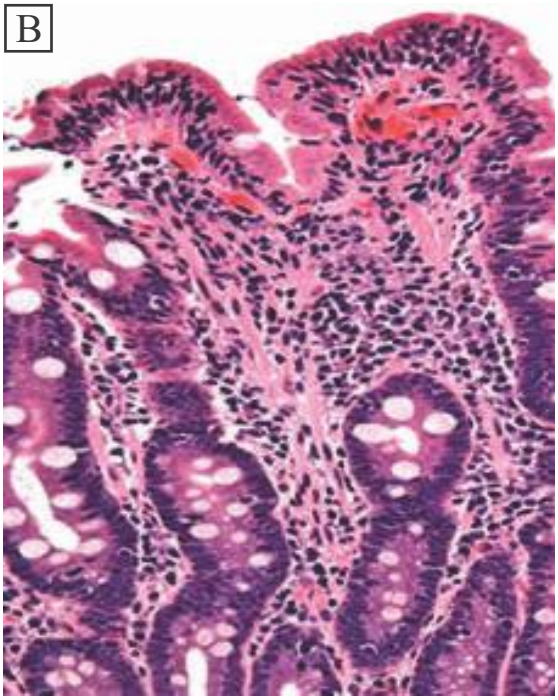
Similar presentations of celiac disease with acute loss of vision related optic neuropathy has been reported in literature. Boushehri et al. reported an adult who responded to immunosuppressive therapy and gluten free diet. [2] Another case of celiac disease with right sided optic neuritis was reported in which the neurological examination was unremarkable and brain MRI showed non specific white matter lesions. Vasculitis tests were normal and anti-aquaporin 4 antibody was negative. CSF was normal and oligoclonal band was negative. The optic neuritis improved with intravenous methylprednisolone pulse therapy for 5 days. [3] A pediatric case has also been reported with recurrent optic neuritis, celiac disease, partial IgA and IgG3 deficiency. Treatment with Tacrolimus was successful in preventing disease relapses. [4]

To conclude, we report the unusual association of optic neuritis and celiac disease. The fact that celiac is an autoimmune disease and one autoimmune disease predisposes to another, the optic neuritis in this case with response to immunosuppression appears to be linked to celiac disease.

Fig1 – (A) MRI orbit showed loss of perineural CSF signals, focal hyper intensity suggestive of demyelinating optic neuritis.



(B) Duodenal mucosal biopsy demonstrated crypt hyperplasia, lymphocytic proliferation and villous atrophy consistent with Marsh grade IIIb.



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