



Case Report

Association of Immune thrombocytopenia with Celiac Disease in a young female

Authors

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Abstract

A 18 year young female who was known case of celiac disease presented with epistaxis, petechial rash and menorrhagia. Routine workup revealed anemia with severe thrombocytopenia. Bone marrow investigation revealed immune thrombocytopenia. Celiac disease (CD) and Immune thrombocytopenic purpura (ITP) may occur together as a result of similar autoimmune mechanisms.

Keyword: celiac disease, immune thrombocytopenic purpura, autoimmune.

Introduction

The pediatrician K.W. dicke of Utrecht and the Hague, in the early 1930s, was the first to link celiac disease to wheat consumption. He witnessed the confirmation of his hypothesis when his patients' symptoms improved during the grain shortages prevailing toward the end of the second world war.

Celiac disease (CD) is an immunological disease induced by intolerance of the small bowel to gluten. In celiac subjects the ingestion of gluten leads to enteropathy with an impairment of the mucosal surface and, consequently abnormal absorption of nutrients. Many auto immunological disorders may accompany the disease, or there may be extra intestinal findings, such as growth retardation, developmental delay, impaired hepatic functions, skin manifestations, osteoporosis, or hematological disorders. As a

result of having similar autoimmune mechanism, in some publications, CD has been reported as a risk factor for Immune thrombocytopenia (ITP).

Case Report

A 18yr old young female known case of celiac disease (Biopsy proven) presented with an episode of epistaxis, petechial rash over extremities and involving oral cavity to the emergency department, was managed conservatively. On initial presentation she was conscious, oriented. She was hemodynamically stable. Physical examination was unremarkable except anterior nasal pack was present. There was no history of any trauma, use of NSAIDs, fever or sore throat in past few months. There was history of menorrhagia in last 3-4 months. She was diagnosed with CD 9 years prior to the date of presentation and was put on celiac free diet. For

past few months she was not following the proper celiac diet. Routine laboratory investigations were done. Her CBC revealed normocytic normochromic anemia with severe thrombocytopenia (20,000 /uL) for which further workup including bone marrow was done. Bone marrow investigation revealed increased number of megakaryocytes hence she was managed in line of ITP. Platelet transfusion was done in view of severe thrombocytopenia but her platelets counts were consistently on the lower side and did not increase. After bone marrow report she was started on IV steroids but still the platelet counts did not increase hence she was put on IVIG therapy. Later, her platelet counts improved subsequently. At the time of discharge patient's platelet counts was 1.3lac/ cu mm.

Investigations

RBC Total	2.78 millions/ul
MCV	78.1 fl
MCH	22.3 pg
MCHC	28.6 g/dl
Hemoglobin	6.2 gm/dl
WBC Total	6.60 ths/ul
Platelet count	20 ths/ul
ESR	02 mm /1 st hr
Anti-H pylori IgA	Negative
Anti mycoplasma pneumonia IgM	Negative
Antibodies to tissue Transglutaminase- IgA	Positive

Bone Marrow Biopsy interpretation–

Dimorphic erythroid hyperplasia with megakaryocytic thrombocytopenia

Duodenal biopsy which was done 9 yrs before -

Microscopic findings showed fragments of duodenal mucosa having partial villous atrophy with crypt hyperplasia. There was marked chronic duodenitis with increase in intraepithelial lymphocyte giving the impression which was consistent with celiac disease (Modified marsh classification –grade III A)

Discussion

In cases of CD, generally, the clinical findings become manifest due to intestinal mucosa injury and resultant malabsorption^[1]. Various

hematological symptoms can be seen in CD due to deficient intestinal absorption, and autoimmune disease. In CD, apart from anemia other hematological abnormalities like leukopenia and thrombocytopenia may develop due to micronutrient deficiencies, and immune cytopenias, such as ITP, can develop via different autoimmune mechanisms^[2]. In this case no micronutrient deficiency was seen.

Concomitancy between CD and ITP was first described in 1988, and studies performed have demonstrated the presence of similar autoimmune mechanisms in the pathogenesis of both diseases^[10]. It has been determined that the native immune system is important in the pathogenesis of CD, and that toll-like receptors (TLRs) also play a key role^[3]. Zanoni et al.^[4] demonstrated that in some cases of CD, tissue transglutaminase antibody (tTG) antibodies induce TLR4 activation. Presumably, TLR4 expression in platelets leads to thrombocytopenia^[5]. CD is a frequently seen disease in the community; however, diagnosis can be overlooked due to its multivariant symptoms^[6].

Conclusion

ITP is one of the atypical findings of CD that may accompany CD due to a similar autoimmune mechanism. Although investigation for CD is not required in every case diagnosed as ITP, in ITP patients with developmental retardation or malabsorption, considering CD in the differential diagnosis is important so as to prevent unnecessary treatment. We therefore suggest there should be an increased awareness of CD in patients with ITP.

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