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Evans Syndrome Secondary to Celiac Disease - A Case Report

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Abstract: ***This article presents a rare case of Evans syndrome, a challenging autoimmune disorder marked by concurrent autoimmune hemolytic anemia AIHA and immune thrombocytopenic purpura ITP, occurring secondary to celiac disease. A 36 - year - old female, adherent to a gluten - free diet due to her celiac condition, presented with symptoms of fatigue, weakness, and easy bruising. Clinical findings, including severe anemia and thrombocytopenia, along with positive serological tests, confirmed the diagnosis of Evans syndrome in the context of active celiac disease. Treatment involved corticosteroids, intravenous immunoglobulin IVIG, and Rituximab, leading to significant improvement. This case underscores the need for vigilant monitoring for autoimmune complications in patients with celiac disease, highlighting the potential interconnection between these conditions. The article calls for further investigation into the mechanisms linking Evans syndrome and celiac disease to improve patient outcomes***

Keywords: Evans syndrome, celiac disease, autoimmune hemolytic anemia, immune thrombocytopenic purpura, autoimmune disorders

1. Introduction

Evans syndrome is a rare autoimmune disorder characterized by the presence of both autoimmune hemolytic anemia (AIHA) and immune thrombocytopenic purpura (ITP). It is a challenging condition to manage and can be associated with other autoimmune diseases. Here, we present a case report of Evans syndrome secondary to celiac disease, along with relevant images.

2. Case Report

A 36 - year - old female presented to our hospital with a history of persistent fatigue, weakness, and easy bruising. She had a previous diagnosis of celiac disease, for which she was adherent to a gluten - free diet. Physical examination revealed pallor and petechiae on her extremities. Laboratory investigations showed severe anemia with a hemoglobin level of 6 g/dL and severe thrombocytopenia with a platelet count of 14, 000/µL.

Further workup revealed a positive direct Coombs test, confirming AIHA, and a low platelet count, consistent with ITP. The patient was diagnosed with Evans syndrome secondary to celiac disease. She was started on corticosteroid therapy and intravenous immunoglobulin (IVIG) with gradual improvement in her hemoglobin and platelet levels.

A diagnostic workup for celiac disease, including serologic testing, positive anti tissue transglutaminase antibody and endoscopic biopsy, confirmed the presence of active celiac disease despite adherence to a gluten - free diet. The patient was referred to a gastroenterologist for further management of her celiac disease. The patient was managed with IV glucocorticoids and Rituximab and showed significant improvement.

3. Discussion

Evans syndrome is a complex disorder that often requires a multidisciplinary approach for management. In this case, the association of Evans syndrome with celiac disease is an important finding, as it suggests a possible link between the two conditions. Patients with autoimmune disorders, such as celiac disease, should be monitored closely for the development of Evans syndrome and other autoimmune complications. The responses were most likely with glucocorticoids, rituximab, and splenectomy. [1] Case reports have described responses to hematopoietic stem cell transplantation and IVIG [2, 3, 4]

4. Conclusion

This case highlights the rare association of Evans syndrome with celiac disease and emphasizes the importance of a comprehensive evaluation in patients with autoimmune disorders. Early recognition and management of Evans syndrome in the setting of celiac disease can lead to better outcomes for affected individuals. Further research is warranted to better understand the underlying mechanisms linking these two conditions.

5. Images

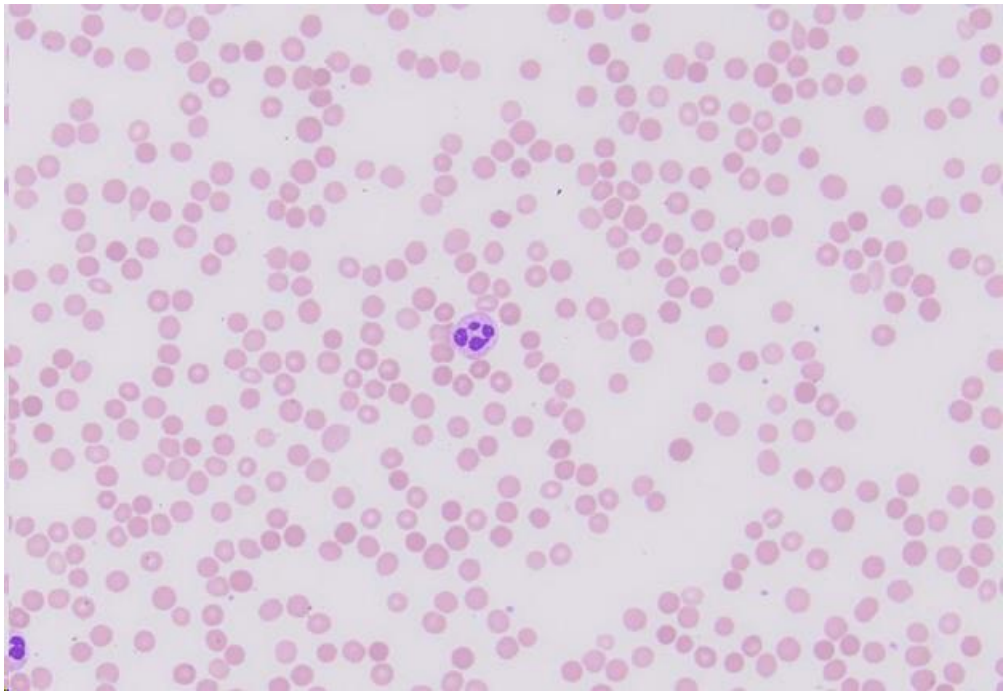

Figure 1: Peripheral blood smear showing spherocytes and thrombocytopenia

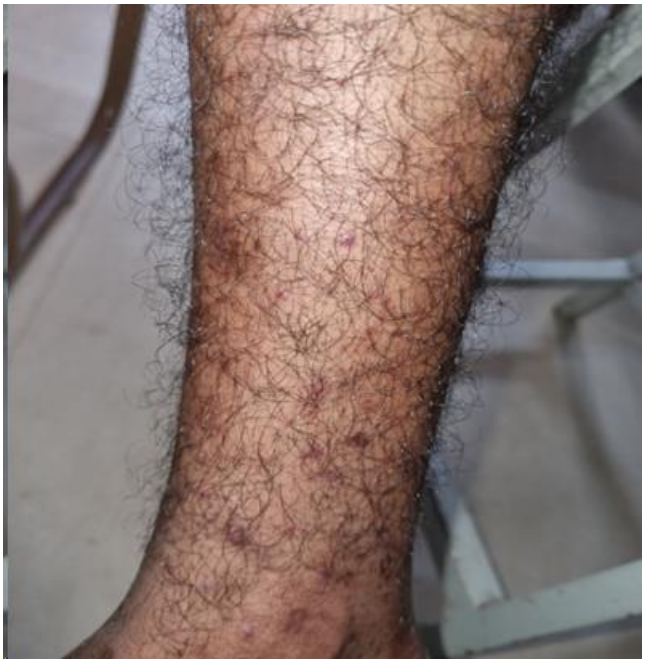

Figure 2: Petechiae on the lower extremities

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