01. Evans Syndrome Associated with Heart Abnormalities: A Case Report with Literature Review

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Background: Evans syndrome (ES) is an uncommon autoimmune disorder characterised by the simultaneous or transient occurrence of autoimmune hemolytic anemia (AIHA) and immune thrombocytopenia (ITP), sometimes additionally associated with autoimmune neutropenia (AIN). Cardiovascular manifestations have been occasionally reported in patients with Evans syndrome. However, valvular heart disease in adult patients with primary Evans syndrome, without underlying connective tissue disorders, has not been previously documented.

The Case

A 40-yr-old woman presented with dizziness, low-grade fever and episodic hematemesis. She had a history of unexplained pancytopenia for many years. Over time, she had required repeated blood transfusions for anemia, thrombocytopenia and leukopenia. Investigations revealed pancytopenia with a positive direct antiglobulin test (polyspecific anti-IgG and anti-C3d, 3+), supporting an autoimmune hemolytic process. No secondary cause was found for her hemolytic anemia, including nutritional deficiencies, current viral infections, and autoimmunity secondary to connective tissue disorders or underlying hematological malignancies. A diagnosis of primary Evans syndrome was made. Transthoracic echocardiography revealed thickened aortic valves with moderate aortic regurgitation and mild aortic stenosis and thickened mitral valves with mild mitral regurgitation. Valvular disease was noted to be out of proportion to the duration and absence of a prior diagnosis of any connective tissue disorder. The patient was started on systemic corticosteroids for her Evans syndrome and showed a good hematological response.

To the best of our knowledge, no previously published case has described this combination of valvular abnormalities in association with adult primary Evans syndrome. We cannot attribute the patient's findings to a particular mechanism of disease, whether it be chronic immune activation or endothelial injury. This case broadens the known clinical spectrum of Evans syndrome and suggests that cardiac evaluation should be considered in these patients.

Conclusion: This case draws attention to a previously unreported association between adult primary Evans syndrome and multivalvular cardiac disease and suggest that all patients with primary Evans syndrome undergo routine cardiac evaluation. Further studies will be necessary to clarify the spectrum of cardiovascular disease seen in Evans syndrome

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