



PAEDIATRIC DERMATOLOGY

THE WISKOTT-ALDRICH SYNDROME: A DIAGNOSIS NOT TO IGNORE

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Introduction: The Wiskott-Aldrich syndrome is a rare immunodeficiency 1/200, 000 births recessive transmission linked to chromosome X. This deficit manifests itself in young boys by skin type of eczema signs, signs of bleeding associated with thrombocytopenia, recurrent infections related to immune deficiency and signs of self immunity.

Methods: A male infant of 18 months was admitted to the pediatric ward for ear infections with a syndrome colitis. Cutaneous examination revealed lesions of eczema in the lower limbs, purpuric lesions with diffuse cutaneous xerosis. NFS objectifying severe thrombocytopenia 20,000 / mm³ smear confirmed that the realization of a myelogram which eliminated a central origin the presence of megakaryocytes without morphologic abnormalities view.

Immunological tests showed a clear elevation of IgE undisturbed IgG, IgM and IgA. The patient received treatment with corticosteroids and immunoglobulins, the evolution was very satisfactory both in terms clinical and biological (increased to 60,000 / mm³ platelets).

Discussion: The Wiskott-Aldrich syndrome is due to a molecular defect with mutation of the WAS gene (located in xp 11.22 containing 12 exons) this mutation leads to a loss of forming the protein. It is characterized by the combination of often severe thrombocytopenia, eczema and immune deficiency. The genetic study is useful to confirm the diagnosis, identify the type of mutation and detect conductive.

The differential diagnosis is mainly with X-linked thrombocytopenia. The treatment goal for the correction of hemorrhagic syndrome and immune deficiency and is mainly based on corticosteroids and immunoglobulins intra venous. The outcome was favorable in our patient due to its rapid treatment

Conclusion: The Wiskott-Aldrich syndrome is rare, however, before any lesion of eczema associated with a syndrome recurrent bleeding and infection in a report type NFS is needed to make an early diagnosis, support fast and prevent the occurrence of complications, monitoring is important to detect malignancies that may arise.

Keyword: eczema, thrombocytopenia, Wiskott Aldrich

