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Research Article

POLYRADICULONEUROPATHY INDICATIVE OF WISKOTT SYNDROME - ALDRICH ON A CASE

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Abstract

Wiskott-Aldrich syndrome (WAS) is a rare inherited immune deficiency of recessive transmission linked to the X chromosome, manifested by hemorrhagic signs, eczema and whose hematological study shows severe thrombocytopenia. Cellular and humoral responses, which are triggered during infection can lead to the development of neuropathies

Keywords: Poly radiculoneuropathy, Wiskott syndrome – Aldrich.

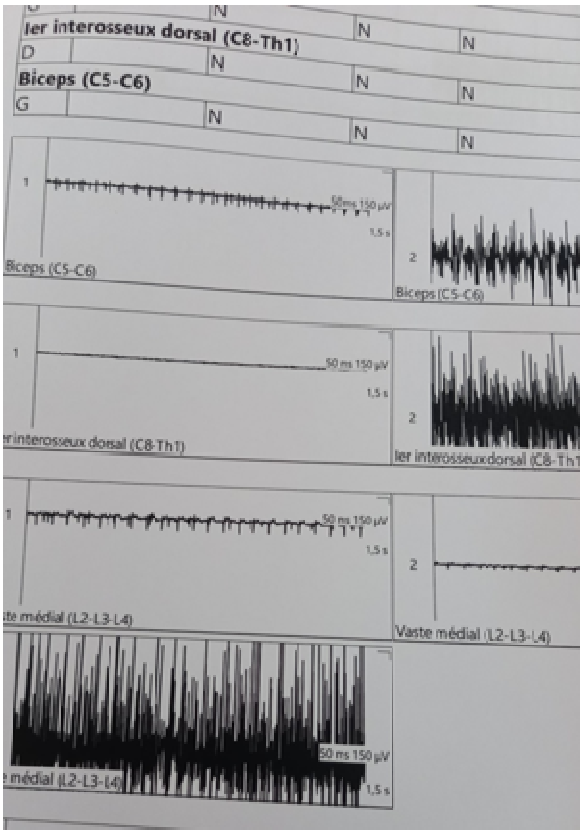
INTRODUCTION

Wiskott-Aldrich syndrome (WAS) is a rare hereditary immune deficiency (1/200 000 births) of recessive transmission linked to the X chromosome. This deficiency manifests itself in young boys, by hemorrhagic signs (purpura, petechiae, ecchymosis, epistaxis, bloody diarrhea or other), recurrent infections (bronchial, pulmonary, ENT), eczema and sometimes signs of autoimmunity. The haematological study reveals severe thrombocytopenia, often less than 50,000 platelets, as well as small platelets (4 to 5 microns). Auto-reactive immune reactions the cellular and humoral response, which are triggered during a infections can lead to the development of neuropathies

DISCUSSION

Clinical presentation is based on a clinical score (Mahlaoui et al). This disease consists of a primary immunodeficiency, characterized by a triad: • recurrent infections; • eczema; • thrombocytopenia, thrombopathy-small platelets. However, only 27% of patients present this triad at diagnosis. This disease can also be complicated by autoimmune pathologies, such as vasculitis or autoimmune hemolytic anemias but also lymphoma-type cancers (Watson RD et al) the clinical triad as well as autoimmune damage was present in our patient. Small platelet thrombocytopenia is a constant feature in all patients whether the form is severe or not. X-linked thrombocytopenia (XLT) is described as a variant of this disease, the immune deficiency is less severe or absent (Villa et al), and however with some variability and possible progression to severe complications (autoimmunity in particular) at all stages of life (Albert et al). Vasculitis is also a classic complication (13%) which occurs in both severe and hitherto minor forms. The localization is variable, ranging from cutaneous vasculitis to cerebral vasculitis. Vasculitis can cause the walls of blood vessels to thicken or narrow, thereby cutting off the vital supply to tissues and organs thus triggering peripheral neuropathy. The prognosis is bleak, leading to debate on the indication for hematopoietic stem cell transplantation (HSC).

Other autoimmune complications are listed such as arthritis (10%), IgA glomerulonephritis (12%) and neutropenia (sometimes pan cytopenia) and inflammatory colitis. Lymphomas and leukemias constitute the majority of the malignant pathologies reported, the most frequent of which is non-Hodgkin's malignant lymphoma (l Dupuis-Girod et al). Lymphomas can be induced by means of proximal lymphomatosis infiltration of the roots; of peripheral nerve damage is given a picture of polyradiculo neuropathy. Wiskott-Aldrich disease is therefore currently called WAS / XLT (Villa A et al) including these patients with a less severe initial presentation corresponding to a score often less than 3. Another phenotype is X-linked neutropenia with a gain-of-function mutation in the WAS gene (Massaad MJ, et al).



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Conclusion

Hereditary pathology with varied clinical manifestation, the diagnosis of which is based on a score described by Clinical Score according to Imai et al. And peripheral nerve damage which can be directly linked to the disease via the immune system or indirectly via damage by compression or by disturbance of the vascularization. The lack of financial means did not allow us to do genetic research that would provide more for the diagnosis.

All the authors participated in the development of the document and clarified in any conflict of interest

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