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Neuropsychiatric Disorders Revealing Autoimmune Polyendocrine Syndrome Type 3: Case Report

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Abstract **Case Report**

Multiple autoimmune syndrome is a rare pathological condition because it associates at least three different autoimmune diseases in the same patient. We discuss through this case and with a review of the literature MAS type 3 and its association with neuroanemic syndrome which remains exceptional. This is a 47-year-old patient, followed for vitiligo and a depressive syndrome, admitted for an ataxic gait lasting a month, in whom the neurological examination revealed a combined sclerosis syndrome of the spinal cord associated with a static cerebellar syndrome. Dermatological examination revealed diffuse acrofacial vitiligo. The biological assessment revealed Biermer's anemia and autoimmune thyroiditis. The clinical evolution was favorable under treatment. The discovery of Biermer's disease in a woman should prompt the search for arguments for one or other associated autoimmune diseases.

Keywords: autoimmune diseases, neuroanemic syndrome, neurological examination, Dermatology.

INTRODUCTION

Multiple autoimmune syndrome (MAS) is a rare pathological condition, recently described and particular because it combines at least three different autoimmune diseases in the same patient [1]. We discuss through this case and with a review of the literature MAS type 3 and its association with neuroanemic syndrome which remains exceptional in young subjects.

CLINICAL CASE

This is a 47-year-old patient, followed for vitiligo for 20 years and a depressive syndrome resistant to anti-depressants for 3 years. She was admitted for ataxic gait that had been going on for a month, with neurological examination showing a combined spinal cord sclerosis syndrome associated with a static cerebellar syndrome. Dermatological examination revealed diffuse acrofacial vitiligo (fig 1). The biological assessment showed macrocytic anemia (Hb: 89 g/l, VGM: 117 fl), vitamin B12 deficiency and elevated TSHus (7 mIU/l). The immunological assessment was positive with the presence of high levels of anti-intrinsic factor, anti-parietal cell, anti-thyroglobulin, anti-thyroperoxidase and antinuclear antibodies. The myelogram revealed medullary megaloblastosis. Gastro-duodeno-jejunal fibroscopy with biopsies revealed fundic atrophy without signs of malignant degeneration. The study of visual evoked potentials revealed bilateral retrobulbar optic neuritis with a prolongation of the P100 latency. Electroneuromyogram and electroencephalogram were free of abnormalities. In light of these data, our patient was put on vitamin B12 supplementation (Hydroxocobalamin) at a dosage of 5000 gamma/day intramuscularly for seven days, then 5000 gamma/week for one month and finally 5000 gamma/month at life. The evolution was very favorable under treatment with a clear improvement in walking, a correction of hematological disorders and a spectacular regression of neuropsychiatric symptoms after 6 months.

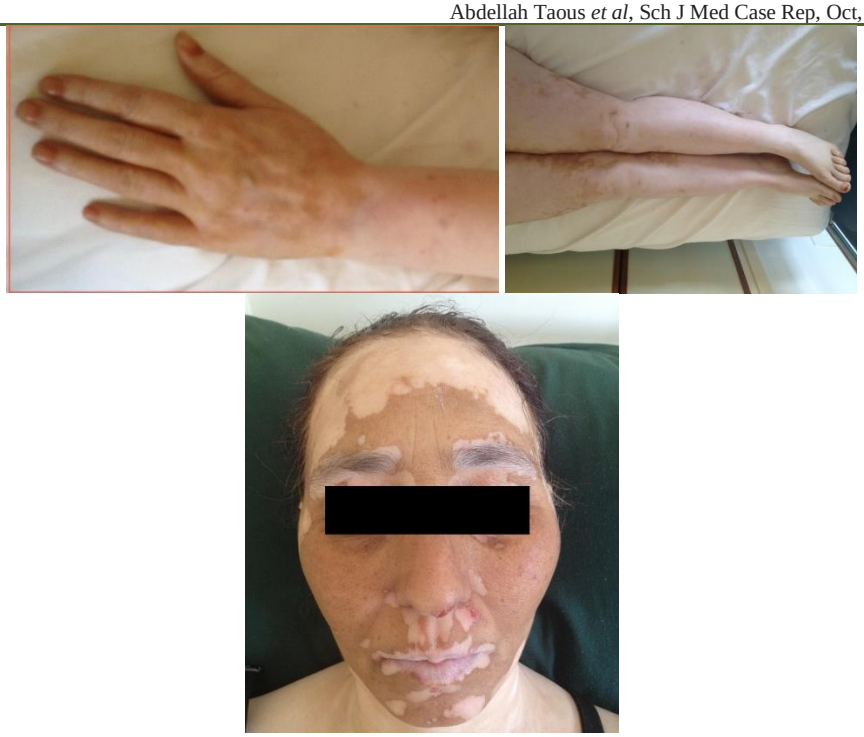

Figure 1: Diffuse acrofacial vitiligo

DISCUSSION

Multiple autoimmune syndromes constitute a rare nosological context, of which less than a hundred cases are reported in the literature [2]. In 1988 Humbert and Dupond [2] proposed a classification of multiple autoimmune syndromes, defined by the association in the same patient of at least three different autoimmune diseases. MAS type 3 combines autoimmune thyreopathy, myasthenia gravis or thymoma, Sjögren's disease, Biermer's disease, idiopathic thrombocytopenic purpura, Addison's disease, autoimmune diabetes, vitiligo, hemolytic anemia autoimmune disease and systemic lupus erythematosus [3]. In our patient, the diagnosis of MAS type 3 was made due to the association of vitiligo, neuroanemic syndrome and Hashimoto's thyroiditis. Our observation illustrates a real cascade of autoimmune diseases where the neuroanemic syndrome associated with vitiligo has prompted the systematic search for other associated autoimmune disorders thus revealing thyroiditis integrating into the context of a MAS type 3. In the literature, the neurological disorders of vitamin B12 deficiency are dominated by the combined sclerosis of the spinal cord [3, 4], as is the case in our patient. The other neurological signs are very polymorphic. In 41% of cases, they are related to mixed spinal cord and peripheral involvement [3]. These neurological disorders are generally discreet and are sometimes only detected by electromyogram. Peripheral neuropathies are described, most often purely sensory, predominating in the lower limbs [5], cases of cerebellar ataxia and involuntary movements [6]. Impairment of the cranial nerves such as diplopia and retrobulbar optic neuritis has also been described [7]. In the series by A.-A. Zulfiqar *et al* [2], Autoimmune thyreopathies represent a key element of multiple autoimmune syndromes. Within multiple autoimmune syndromes, vitiligo appears to be the most common autoimmune dermatological condition [8]. The acral and/or facial, bilateral and symmetrical character of vitiligo (Fig 1), as is the case in our patient, is a marker of multiple dysimmunity [9]. The treatment of neurological involvement in the context of vitamin B12 deficiency does not differ from the treatment of forms without neurological disorders. Neurological recovery seems essentially related to the precocity of the supplementation treatment. There is no consensus on the doses of vitamin B12 to be administered [10].

CONCLUSION

This succession of autoimmune diseases in this woman should lead us to monitor a patient suffering from an autoimmune disease in a prolonged and regular manner in order to detect in time the outbreak of new diseases which can occur at any time during of evolution and especially an autoimmune thyreopathy.

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