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Autoimmune Hepatitis as Part of Multiple Autoimmune Syndrome: About A Rare Case

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Abstract **Case Report**

Multiple autoimmune syndromes (MAS) is a rare entity defined by the presence in the same person of at least three different auto-immunes diseases. The association of autoimmune hepatitis with other autoimmune manifestations apart from primary biliary cirrhosis (PBC) or primary sclerosing cholangitis (PSC) is exceptional. It can be with any type of known localized or systemic autoimmune disease. We report through this observation the case of autoimmune hepatitis discovered as part of a multiple autoimmune syndrome made of vitiligo, autoimmune thyroiditis and type 1 diabetes.

Keywords: Multiple autoimmune syndrome, autoimmune hepatitis, MAS type 3.

INTRODUCTION

Multiple autoimmune syndromes (MAS) is a rare combination in the same patient associating at least three different organ-specific and/or non-specific autoimmune diseases (AI).

Autoimmune hepatitis (AIH) is a chronic inflammatory liver disease characterized by auto-antibodies and hypergammaglobulinemia, which can progress to hepatic cirrhosis in the absence of treatment. It is frequently associated with other autoimmune diseases such as primary biliary cirrhosis (PBC) or primary sclerosing cholangitis (PSC), which explains the current tendency to make systematic screening as soon as the disease is revealed.

We report a particular observation of multiple autoimmune syndrome combining type 1diabetes, vitiligo, autoimmune thyroiditis and autoimmune hepatitis.

CASE REPORT

A 50-year-old woman, followed since the age of 7 for type 1 diabetes (T1D) on Basal – Bolus insulin therapy (Rapid Insulin 10 units 3 times a day and 14 units of slow insulin per day). She has had a vitiligo since the age of 18 (Figure 1). The patient is also followed for autoimmune thyroiditis retained in front of peripheral hypothyroidism and positive antithyroid antibodies.

Admitted for a cholestatic jaundice (dark urine, pale stools, generalized pruritus), evolving for 1 month in a context of apyrexia and conservation of the general state.

The biological assessment showed cholestasis with high alkaline phosphatases (APL) and gamma glutamyl transferase (GGT) (ALP= 210 U/L, GGT= 120 U/L), hyperbilirubinemia at 368 mg/L predominantly conjugated, associated with hepatic cytolysis with high liver enzymes (ALT=558 U/L and AST= 1591 U/L). Total IgG levels were high at 22.9 g/L, with positives anti-smooth muscle antibodies and anti-Liver and Kidney Microsome (LKM1) antibodies. Viral serologies were negative, eliminating viral hepatitis.

The liver histology was in favor of autoimmune hepatitis with moderate lymphocytic interface hepatitis associated with a plasma cell-rich infiltrates. These arguments led to the diagnosis of autoimmune hepatitis.

The patient was treated with 1 mg/kg/day oral corticosteroid therapy in progressive degession, combined with treatment with Azathioprine as long-term maintenance therapy.

The evolution was favorable, marked by the regression of symptomatology and normalization of liver function tests.

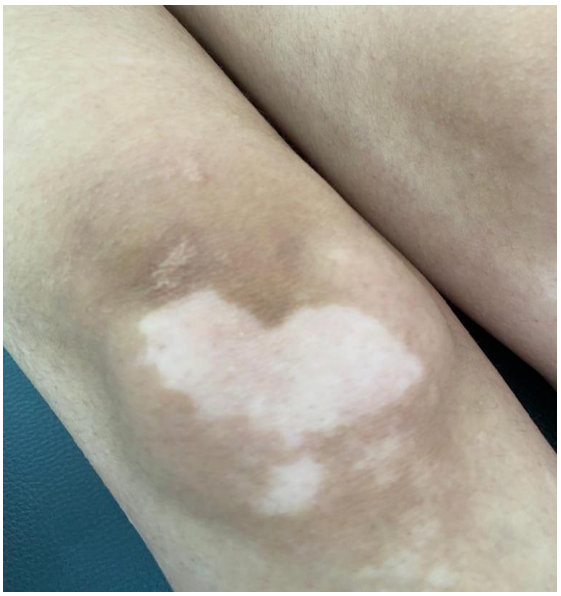

Fig-1: Vitiligo visible in the knee

DISCUSSION

Multiple autoimmune syndrome (MAS) is a rare entity defined by the association of at least three different autoimmune diseases in the same person(Sow *et al*., 2019).

The pathogenesis of MAS remains unknown to date. It probably involves environmental factors in genetically predisposed individuals inducing immune dysregulation. Studies have shown the role of the HLA-DR3 or HLA-DR4 haplotype in the association of multiple autoimmune diseases(Sow *et al*., 2019). On the other hand, some mono- or polyclonal autoantibodies specific to one organ may be reactive to both other organs(COJOCARU *et al*., 2010).

Three types of MAS are distinguished according to the associated autoimmune diseases. Type 1 MAS includes autoimmune myasthenia gravis, thymoma, polymyositis and autoimmune myocarditis. Type 2 MAS includes Sjögren-Gougerot syndrome, rheumatoid arthritis, primary biliary cirrhosis, scleroderma and autoimmune thyroiditis. Type 3 MAS contains autoimmune thyroiditis, myasthenia gravis, thymoms, Sjögren-Gougerot syndrome, Biermer's disease, idiopathic thombopenic purpura, Addison's disease, type 1 diabetes, vitiligo, auto-hemolytic anemiaimmune, lupus and herpetiform dermatitis(Cherif *et al*., 2014).

Association of autoimmune hepatitis with other autoimmune manifestations outside of primary biliary cirrhosis (PBC) or primary sclerosing cholangitis (PSC) is rare. It can be with any type of known autoimmune disease, whether localized (uveitis for example) or systemic (lupus for example)(Teufel et al., 2010).

The frequency of patients with autoimmune disease associated with AIH is around 40% in the Teufel(Teufel *et al*., 2010) and Choudhori (Choudhuri *et al*., 2005) series on all patients with AIH included in their studies.

Autoimmune thyroiditis is by far the most associated autoimmune manifestation of AIH outside of CBP and CSP. (10.1% of patients in the Teufel series(Teufel *et al.*, 2010) and 8% in the Choudhori study(Choudhuri *et al*., 2005)), hence the interest of the early research of autoimmune thyroiditis in patients with autoimmune hepatitis.

The association of AIH with type I diabetes (T1D) is described in 11% of patients in the Choudhori study(Choudhuri *et al*., 2005), unlike 0.7% in the Teufel series (4) of patients with HAI. Homberg *et al*. reported that T1D was found in 6.2% of AIH patients with LKM-1 antibodies and in 1.7% of patients without LKM-1 antibodies(Homberg *et al*., 1987).

Our observation corresponds to a particular association made of autoimmune hepatitis associated with three other autoimmune diseases: autoimmune thyroiditis and type 1 diabetes and vitiligo as part of a multiple autoimmunity syndrome.

The association of other autoimmune manifestations with autoimmune hepatitis does not alter the clinical evolution or severity of this pathology. However, the relapse rate is higher in patients with additional autoimmune diseases (Teufel *et al*., 2010).

CONCLUSION

We have reported through this observation a special and rare case of an association of autoimmune hepatitis and a type 3 multiple autoimmune syndromes.

Multiple autoimmune syndromes are a rare situation that demonstrates a common genetic substratum among some groups of autoimmune diseases. Their screening during follow-up allows better management of patients.

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