



New Case of Anti-Synthetase Syndrome Associated with Cirrhosis Post HVB

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Abstract

The anti-synthetase syndrome is represented by the association of inflammatory myopathy (polymyositis/dermatomyositis), interstitial pneumonitis, Raynaud's syndrome, inflammatory polyarthritis, and characteristic skin abnormalities. We found also a clinical aspects of Mechanic's hand with high level of anti-synthetases. We report the case of a 66-year-old patient with polyarthralgias of large and small inflammatory joints, myositis, Raynaud's phenomenon, and cutaneous involvement of the hands and feet, with fissure hyperkeratosis feet and an appearance of mechanic's hands. The patient had an inflammatory syndrome, elevated muscle enzymes, positivity of anti-Jo1 antibodies, PID at chest CT and the presence of inflammatory mononuclear cell infiltrates at muscle biopsy. The evolution under immunosuppressive treatment combining corticosteroids and methotrexate was marked by an oedemato-ascitic decompensation of a post-HVB cirrhosis unknown.

Subject Areas

Epidemiology, Gastroenterology & Hepatology

Keywords

Cirrhosis, Hepatitis, HVB, Anti-Synthetase

1. Introduction

The anti-synthetase syndrome is an autoimmune disease clinically associating with interstitial lung disease (ILD), inflammatory myopathy (polymyositis/dermatomyositis), Raynaud's phenomenon, inflammatory polyarthritis and mechanic-type skin abnormality [1]. It is characterized by the presence of anti-aminoacyl transferase RNA synthetases, and an association with post-viral pathologies is possible. Our patient presents an anti-synthetase syndrome associated with post

HVB cirrhosis in oedemato-ascitic decompensation.

2. Observation

This is a 66-year-old patient of Caucasian origin, having a history of pulmonary tuberculosis treated thirty years ago, being a chronic smoking drug weaned a year and a half ago. He presented a symptomatology evolving since one year made of polyarthralgias of large and small joints of inflammatory pace, a myositis with a myalgia associated with symmetrical proximal muscle weakness, a phenomenon of Raynaud as well as a cutaneous involvement of the hands and feet, with fissured hyperkeratosis of the feet (**Figure 1**) and an aspect of mechanic's hands.

The paraclinical assessment highlights:

We found an inflammatory syndrome with CRP = 28 mg/l and CPK = 4521 U/l.

The immunological report shows a positivity of antinuclear antibodies at 1/640 and in particular anti-nuclear antigen-resistant antibodies of anti-Jo-1 type at 90 U/ml. Anti-native DNA, rheumatoid factors and anti-CCP are normal.

The radiological assessment showed:

The absence of structural damage to X-rays of the various joints, a myogenic syndrome with EMG, an aspect of inflammatory myositis predominant on muscles proximal to muscle MRI and inflammatory infiltrates of perivascular mononuclear cells strongly suggestive of a polymyositis at the muscle biopsy. Diffuse interstitial syndrome with a micronodule appearance and thickening of the peri-bronchovascular and sub-pleural interstitium in CT and a mild-moderate restrictive syndrome with decreased CO₂ transfer capacity indicating significant membrane involvement alveolar.



Figure 1. Shows the aspect of fissured hyperkeratosis of the hand and the feet.

ECG and transthoracic echocardiography are normal:

The diagnosis of anti-synthetase syndrome is based on clinical presentation, the presence of CPK at 6321 U/l, the positivity of anti-Jo1 antibodies, the presence of inflammatory mononuclear cell infiltrates on muscle biopsy and PID on the thoracic plane. The patient is treated with oral corticosteroids and methotrexate. The evolution was marked 3 months later by the appearance of an oedemato-ascitic syndrome made of soft edema of the lower limbs taking the bucket and a syndrome of peritoneal effusion of average abundance.

The biological assessment showed a disturbed hepatic assessment with ASAT at 83 U/l, ALAT at 50 U/l, GGT at 121 U/l, a TP at 58%, a Factor V at 53% and a hypo albuminemia at 19 g/l. A normal renal balance with a urea at 0.27 g/l, a creatinine at 10 g/l and a proteinuria of 24 h at 0.06 g/24 h.

The radiological assessment showed an atrophic liver with irregular finely micronodular contours with ascites of great abundance Actitest: A2 Fibrotest: F4. The exploratory ascites puncture showed transudative ascites to 7 g/l sterile with a gradient (albumin-blood albumin ascites) > 11. Viral serologies have shown that HVB positive with a positive HBsAg at 2501.53 S/Co, and anti-HBc positive total at 11.49 S/Co and a viral load at 8.74 Log. The serology of HVC was negative. FOGD showed grade I oesophageal varices.

The diagnosis of post-HVB cirrhosis in oedemato-ascitic decompensation associated with an antisynthetase syndrome was selected and the patient was put on diuretic and direct antiviral therapy. The evolution was marked by death following respiratory decompensation.

3. Discussion

The anti-synthetase syndrome is a systemic autoimmune disease characterized by autoantibodies directed against one of the many aminoacyl transfer RNA (tRNA) synthetases and which associates an inflammatory myopathy (polymyositis/dermatomyositis), an interstitial lung disease (ILD), Raynaud's phenomenon, inflammatory polyarthritis, and mechanic-type skin abnormality [1]. Among these criteria, our patient had clinical features consistent with PM/DM, Raynaud's phenomenon, and pigmented hyperkeratosis of the glabrous skin of the hands associated with fissures.

Anti-aminoacyl-transfer RNA synthetases, which characterize AS, include anti-histidyl (OJ) detected in our case. The other antibodies that are part of the syndrome are: anti-threonyl (anti-PL-7), anti-alanyl (anti-PL-12), anti-glycyl (anti-EJ), anti-asparginyl (anti-KS), phenylalanyl (anti-ZO) and anti-tyrosyl-tRNA (anti-YRS) [2] [3]. Of these, anti-OJ-1 was the first antibody to be recognized and was detected in 20% to 30% of patients with PM or MS [4]. The pulmonary involvement is the severity of the syndrome, generally evolving to fibrosis, in the absence of treatment. It is responsible for an excess mortality of 40%. It does not differ from that of other connectivity's [5] [6] and is initially manifested by a dry cough, then a dyspnea of effort.

The different treatments used in case of SAA are corticoids and immunosuppressors.

The efficacy of corticosteroids has been proven in articular, muscular and general manifestations, as well as in some pulmonary forms [7] [8]. Some observations of favorable evolution under cyclophosphamide [9] or tacrolimus [10] have been reported as well as the interest of cyclosporine A [11] [12]. Joffe *et al.* note a good response in methotrexate and especially azathioprine in the 2nd line of treatment in 143 patients [13] with idiopathic inflammatory myopathies including 34 SAS. Intravenous immunoglobulins, validated in the second line in inflammatory myopathies [14], have not been specifically studied in pulmonary involvement of SAS. On the other hand, they have an efficacy on myositis with improvement of muscle testing and normalization of muscle enzymes [15] [16]. Our patient has been treated with oral corticosteroids and methotrexate. Hepatic involvement during SSA is rare and often caused by an autoimmune mechanism whose detailed pathogenesis is unknown [17].

Cases of patients with HCV infection who have developed inflammatory myopathy and interstitial lung disease have been reported [18], some of whom have developed cirrhosis and HCC [19]. The combination Polymyositis and HCV infection at or without HCC has been described with worsening clinical signs of myositis after hepatitis exacerbation, suggesting a close association between hepatitis B infection and myositis [20] [21].

Thus, immunosuppressants have been shown to pose a significant risk of stimulating the replication of many viruses, and increase the progression of fibrosis during viral hepatopathies B and C. Therefore, in post-HVB cirrhosis, administration of these drugs should be associated with antiviral treatment [22]. For post-HVC cirrhosis, increased monitoring is recommended [23].

In our case, it was an SSA treated with immunosuppressants and triggering a œdemato-ascitic decompensation of an unknown post-HVB cirrhosis. In general, the poor prognosis of SAS is related to the frequency and severity of interstitial lung disease and its corticosteroids resistance, requiring the frequent use of immunosuppressants. Thus the association with other pathologies such as viral infections and cirrhosis makes the prognosis worse.

4. Conclusion

The syndrome of antisynthetases is an entity to know because of high level of morbidity and mortality. The diagnostic reaches the value for idiopathic inflammatory myopathies and the severity of the pulmonary involvement; treatment with corticosteroids and immunosuppressant should be initiated promptly, taken into consideration an association with other infectious or hepatic diseases.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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