

Primary Biliary Chirrhosis Associated with Scleroderma: Case Report and Review of The Literature

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Abstract

Case Report

A 57-year-old housewife with primary biliary cirrhosis associated with scleroderma but without features of the calcinosis-Raynaud's phenomenon-sclerodactyly-telangiectasia syndrome is reported. The mechanisms responsible for the co-occurrence of these diseases are largely unknown. Genetic, epigenetic, environmental, and infectious factors appear to be important for the pathogenesis of the disease, but the hierarchy of events are not well defined. Some case reports suggested that the presence of SSc in PBC patents is associated with a more favorable prognosis of the liver disease, whereas others report an increased mortality in patients with PBC and SSc compared to patients with PBC alone.

Keywords: scleroderma, calcinosis-Raynaud's phenomenon-sclerodactyly-telangiectasia syndrome, PBC and SSc.

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INTRODUCTION

Scleroderma is a disease of connective tissue often affecting middle-aged females which is characterized by leathery, symmetrical skin induration and involvement of muscle, bone, and various internal organs [1]. Primary biliary cirrhosis (PBC) is a chronic cholestatic liver disease characterized by immune-mediated chronic nonsuppurative cholangitis that mainly affects interlobular and septal bile ducts, affecting middle-aged women and often accompanied by generalized pruritus, skin hyperpigmentation, and thickening [2]. Many cases of primary biliary cirrhosis occurring in association with scleroderma have been reported and in every instance features of eRST have been evident [3]. We report here the coexistence of primary biliary cirrhosis and scleroderma (Reynolds syndrome) without features of the CRST syndrome in a 57-year-old woman.

CASE REPORT

A 57-year-old housewife without any medical history (no alcohol consumption, smoking, or taking any medication) and there was no family history of note. She was referred to our gastroenterology department. She was admitted to our department for an Iron deficiency anemia with jaundice. She denied having choluria, acholia and haematemesis, as well as

fever, night sweats, myalgia and weight loss. On physical examination, the patient was haemodynamically stable and afebrile. She was a well nourished woman with generalized hyperpigmentation. The skin was tight and thickened over the fingers, forearms, chest, back, and face. Superficial telangiectasias were present over the malar areas of the face. Elbow extension was limited to 160° and shoulder abduction to 90°. No sclerodactyly, calcinosis or xanthomas were present. Her abdomen was soft, but tender, without guarding or rebound. A large splenomegaly of 17cm, the liver was not enlarged and no other masses were palpated. On percussion, a shifting dullness was observed, which suggested ascites. Examination revealed no peripheral stigmata of chronic liver disease.

Initial laboratory investigations showed an abnormal liver profile, with hemoglobin 10 g/L, leukocytes 4370 /L and platelets $175 \times 10^3/L$, aspartate aminotransferase 66 U/L, alanine aminotransferase 73 U/L, alkaline phosphatase 254 U/L, gamma glutamyl-transpeptidase 113 U/L, bilirubin 42 $\mu\text{mol/L}$, albumin 34 g/L, prothrombin time 96%, total serum protein 65g/l. No abnormalities of the esophagus, stomach were demonstrated on an upper gastrointestinal. Colonoscopy revealed *rectal varices*. An aetiological study was performed, showing a positive antimitochondrial

antibody (AMA) level and an elevated immunoglobulin IgM. Anti-LKM1, antismooth muscle antibody, antinuclear antibody ANA and anti-SLA were normal. Ultrasonographic examination of abdomen showed a normal-sized liver with homogenous structure signs of portal hypertension. Skin and muscle biopsy from the forearm revealed scleroderma in the sclerotic stage and minimal muscle atrophy.

DISCUSSION

Reynolds syndrome is an autoimmune disease characterized by the co-occurrence of primary biliary cholangitis (PBC) and limited cutaneous systemic sclerosis (LCSS). This association was first described to co-occur by Milbradt in 1934, and it has been noted historically in several case reports. The patient reported here has primary biliary cirrhosis associated with scleroderma without the features of CRST syndrome. She has no calcinosis, Raynaud's phenomenon, or sclerodactyly, and the telangiectasias present are superficial and not those seen in the CRST syndrome [4]. The random association of primary biliary cirrhosis and sclerosis is highly unlikely since both disorders are relatively uncommon. The nature of this association remains obscure as the etiology of both diseases is largely unknown. The pathological tissue reaction in both diseases reveals some general similarities. An early inflammatory reaction followed by fibrosis [5, 6]. This case indicates that the coexistence of primary biliary cirrhosis and scleroderma can occur without features of the CRST syndrome. Fox *et al.*, have described impaired delayed hypersensitivity in patients with primary biliary cirrhosis as measured by their response to dinitrochlorobenzene and tuberculin skin tests [7, 8]. From the practical point of view, it is important to evaluate liver function in all patients with

scleroderma and to investigate all patients with primary biliary cirrhosis for symptoms of connective tissue disorders. These signs are commonly attributed to the multiorgan involvement of the hepatic disease, and in fact are caused by a separate nosological unit.

REFERENCES

1. Kaplan MM, Gershwin ME. Primary biliary cirrhosis, *The New England Journal of Medicine*, 2005; 353(12):1261-1273.
2. Hohenester S, Oude-Elferink RPJ, Beuers U. Primary biliary cirrhosis, *Seminars in Immunopathology*, 2009; 31(3):283-307.
3. Neuberger J. Primary biliary cirrhosis, *The Lancet*, 1997; 875-879.
4. Winterbauer RH. Multiple telangiectasia, Raynaud's phenomenon: sclerodactyly and subcutaneous calcinosis: A syndrome mimicking hereditary hemorrhagic telangiectasia. *Bull Johns Hopkins Hosp*, 1964; 114:361-383.
5. Reynolds TB, Denison EK, Frankl HD, Lieberman FL, Peters RL. Primary biliary cirrhosis with scleroderma, Raynaud's phenomenon and telangiectasia: New syndrome. *The American journal of medicine*. 1971 Mar 1;50(3):302-12.
6. Murray-Lyon IM, Thompson RP, Ansell ID, Williams R. Scleroderma and primary biliary cirrhosis. *Br Med J*. 1970 Aug 1;3(5717):258-9.
7. Fox RA, Scheuer PJ, James DG, Sharma O, Sherlock S. Impaired delayed hypersensitivity in primary biliary cirrhosis. *The Lancet*. 1969 May 10;293(7602):959-62.
8. Fox RA, James DG, Scheuer PJ, Sharma O, Sherlock S. Impaired lymphocyte response in primary biliary cirrhosis. *Proc R Soc Med*, 1970; 63:357-358.