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Letter to the Editor

Pericarditis and possible antiphospholipid syndrome on primary biliary cirrhosis

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Dear Sir,

Primary biliary cirrhosis (PBC) is a chronic liver disease, which may be associated with other autoimmune diseases.[1] Pericardial effusion and detectable antiphospholipid antibodies without clinical evidence of antiphospholipid syndrome have been described in only one patient with PBC.[2] We report a case of PBC who developed pericarditis and possibly antiphospholipid syndrome.

A 68 year-old woman was admitted to our Department with fever (38.5° C), cough, and back pain. She had PBC being treated with ursodeoxycholic acid for the last 8 years. She had also been taking digitoxin for atrial fibrillation since 20 years and acenocumarol for an episode of arterial occlusion of the right leg since three years. Physical examination showed normal blood pressure and pulse rate without paradoxical pulse. Abnormal laboratory tests were: Ht: 34%, Hb: 10.5 gr/dL, ESR: 122 mm, prothrombin time: 27 sec (INR: 2.4), CRP: 6.12 mg/dL. Liver tests were: AST: 34 IU/L (ULN<40), ALT: 17 IU/L (ULN<40), bilirubin: 1.4 mg/dL, alkaline phosphatase: 199 U/L (ULN<130), GGT: 99 U/L (ULN<75). Antimitochondrial antibodies were positive (AMA: 1/160) and antinuclear antibodies positive in low titer (ANA: 1/80). Electrocardiogram was normal, while chest radiograph showed an enlarged cardiac shadow. Transthoracic echocardiography revealed a large pericardial effusion.

Pericardiocentesis was performed, after correction of patient's bleeding tendency and discontinuation of acenocoumarol. The pericardial fluid was bloody with Ht of 12% and protein of 6.3 g/dL. All stains, cultures and cytologic examination were negative. Extensive investigation revealed no specific cause for pericarditis, and all virologic and bacteriological tests were negative. The patient had a good recovery without residual pericardial fluid, but five days after admission she had an episode of transient ischemic attack and anticoagulation therapy was restarted. Thrombophilic study revealed only positive anticardiolipin antibodies (ACA) in high titers (IgM ACA: 630 MPL, normal level<10 MPL) on two 6-monthly occasions. Nine months later, the patient remains in a stable good condition without further complications.

PBC is considered to be an autoimmune liver disease, which may be associated with increased incidence of other autoimmune disorders.[1] Pericarditis may develop due to unknown (idiopathic) or infectious causes or less frequently in association with autoimmune diseases, when it may be associated with circulating immune complexes.[3] Development of pericarditis on PBC is rather rare and there are only two previous case reports suggesting such a possible relation.[2,4] Our patient further supports an association between PBC and pericarditis, since all known causes of pericarditis were excluded.

Hemopericardium has been associated with anticoagulation therapy, but only in case of severe prolongation of prothrombin time.[5] The bloody nature of pericardial fluid in our patient was probably due to her previous anticoagulation therapy.

Antiphospholipid antibodies, with or without clinical syndrome, have been detected in patients with autoimmune hepatitis[6] and in one case with PBC.[2] Patients with antiphospholipid syndrome may have various complications from the cardiovascular system, such as valvular disease, cardiomyopathy and coronary artery disease.[7] Recently, a case of primary antiphopholipid syndrome and large pericardial effusion with cardiac tamponade was described.[8] In addition, pericardial effusion was also observed in the only case of PBC with detectable antiphospholipid antibodies.[2] Although the atrial fibrillation in our patient may have predisposed to her ischemic attack via arterial embolism, particularly after discontinuation of anticoagulation therapy, the positive antiphospholipid antibodies and the previous arterial leg occlusion might favor the diagnosis of antiphospholipid syndrome.

Regardless of possible pathogenetic associations, the coexistence of such diseases poses several therapeutic di-

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Abbreviations: PBC, Primary biliary cirrhosis.

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lemmas, since pericardial effusion is an indication to discontinue anticoagulation therapy that results in increased risk for clinical complications of antiphospholipid syndrome. The better understanding of the associations between PBC and such clinical manifestations will result in better therapeutic decisions for these rare patients.

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