Clin Rheumatol (2007) 26: 821–822
DOI 10.1007/s10067-006-0277-3

CASE REPORT

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Rare association of antiphospholipid syndrome and Takayasu arteritis

Received: 25 July 2005 / Accepted: 29 August 2005 / Published online: 31 March 2006

Abstract The antiphospholipid syndrome (APS) is characterized by obstetric and thrombotic complications in the presence of antiphospholipid antibodies. It can happen in an isolated way or in association with diffuse connective tissue diseases, mainly systemic lupus erythematosus. The association of APS with Takayasu arteritis (TA) is rarely described in the literature. We described a case of primary APS in a female patient who developed obstruction in large-size arteries, in spite of the use of oral anticoagulant, and increase of erythrocyte sedimentation rate, suggesting TA. The favorable response to prednisone treatment and later to infliximab reinforced the diagnosis of TA. The present report illustrates the existence of APS associated to TA, whose recognition is very important once the therapeutic strategy is radically different.

Keywords Antiphospholipid syndrome · Systemic lupus erythematosus · Takayasu arteritis

Introduction

The antiphospholipid syndrome (APS) is characterized by obstetric and thrombotic complications in the presence of antiphospholipid antibodies. It can happen in an isolated way or in association with diffuse connective tissue diseases, mainly systemic lupus erythematosus (SLE) [1]. The association of APS with Takayasu arteritis (TA) has been described rarely in the literature [2, 3]. We report a case of primary APS in a female patient who developed obstruction in large-size arteries, in spite of the use of oral anticoagulant and increase of erythrocyte sedimentation rate (ESR), suggesting TA.

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Case report

A 33-year-old white female was previously diagnosed with primary APS based on history of spontaneous abortion, deep venous thrombosis, pulmonary embolism, and presence of high titles of anticardiolipin (aCL) antibodies and lupus anticoagulant, detected in several occasions, for which she was taking oral anticoagulant, maintaining an INR between 3 and 4. Six months before her admittance to our hospital, she started complaining of an excruciating pain in her left upper limb associated with cyanosis and decreased local temperature. On physical examination, her temperature was 36.5°C, and her blood pressure in right arm was 120/80 mmHg. Peripheral cyanosis of the left upper limb was observed as well as significant decrease in brachial and radial pulses on that extremity. There was also a systolic murmur in left infraclavicular area. The rest of the physical examination was unremarkable. Laboratory evaluation evidenced an ESR of 55 mm, ANA (IIF in HEp-2), anti-DNA, anti-Ro and anti-La negative, IgG anticardiolipin 63 GPL units (normal<10) and IgM 20 MPL units (normal<10), and lupus anticoagulant positive. Doppler ultrasonography demonstrated the presence of obstruction of the left subclavian artery, which was later confirmed by angio-NMR. Arteriogram revealed occlusion of the left subclavian artery; left vertebral artery had 30% of stenosis on its origin and left internal thoracic artery had 90% of stenosis on its origin. The patient was already taking oral anticoagulant, and then it was added with prednisone (1 mg/kg body weight), followed by methotrexate 15 mg/week, with significant improvement of the symptoms and fall of the sedimentation rate even if without improvement on the physical findings. Subsequently, she underwent angioplasty with implantation of stent in the left brachial artery.

Unfortunately, after tapering steroid, the symptoms worsened and the patient was started on anti-TNF∝ therapy, with infliximab 3 mg/kg body weight having a satisfactory response. This regimen has been maintained for 6 months.

Discussion

The present report illustrates a case of primary APS in a patient who, even on full dose of oral anticoagulant, developed manifestation of large-size artery obstruction. Such complication was initially interpreted as a thrombotic arterial obstruction in the context of APS. However, the arteriographic findings characterized by occlusion of subclavian, internal thoracic and vertebral arteries in the absence of thrombi and a significant elevation of ESR suggested that an inflammatory vascular disease such as TA could be responsible for those symptoms. The satisfactory response to prednisone and subsequently to infliximab gave support to such possibility.

At present, there is no evidence demonstrating any role for antiphospholipid antibodies in the pathogenesis of TA. Moreover, antiphospholipid antibodies are not usually found in TA [4]. On the other hand, the coexistence of thrombi and vascular inflammatory phenomena can be eventually demonstrated in histological analyses of samples from patients with APS. If, in selected cases, vascular inflammation may evolve to a well defined vasculitic disorder, such as TA, is not known. Therefore, the association of TA and APS, as occurred with our patient, may suggest either that a single primitive immunological disorder would be responsible for both manifestations or that the coexistence of these conditions may be only coincidental.

Although supported only by uncontrolled reports [5, 6], the use of infliximab seemed to be justified in our patient since she had already been on oral anticoagulant, methotrexate, high doses of prednisone, and experienced worsening in her symptoms after tapering steroid. However, additional studies are necessary to define the real value of the anti-TNF therapy in this clinical condition.

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