

Takayasu Arteritis with Systemic Lupus Erythematosus: A Rare Association

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Abstract

Case Report

Takayasu arteritis (TA) is a systemic vasculitis typically occurring in young female patients, and still of unknown etiology. This vasculitis rarely coexists with systemic lupus erythematosus (SLE). We report the observation of a 45-year-old patient diagnosed with TA who presented with upper extremity ischemia and wall thickening with a good response to the association of methotrexate and corticosteroids. Five years later, she presented with bilateral inflammatory arthralgia of the middle joints with malar rash. Biological assessment showed lymphopenia, positive anti-nuclear antibodies at 1/1280 with a homogeneous fluorescence, positive anti-dsDNA and proteinuria at 4.3g/day. The diagnosis of SLE associated with TA was established.

Keywords: Lupus- Takayasu arteritis –Corticosteroids.

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INTRODUCTION

The frequency of the association of systemic lupus erythematosus (SLE) and Takayasu arteritis (TA) is unknown and probably underestimated. TA is an idiopathic, systemic inflammatory disease, typically involving the aorta and its main branches. While SLE affects mainly the small vessels. The coexistence of these two diseases has been rarely reported. Here we report a case where SLE was preceded by TA.

CASE REPORT

A 45 year-old woman was diagnosed for 16 years with TA revealed by an intermittent claudication of upper limbs, without ocular or cutaneous features. Clinical examination showed an asymmetric blood pressure in upper limbs (Systolic blood pressure was at 150mmHg on the right and 110mmHg on the left), a reduction of radial and humeral pulses, without vascular bruits.

Doppler ultrasound of the upper limb showed an inflammatory arterial wall thickening of subclavian arteries with a low velocity biphasic flow of the axillary and humeral right arteries. Computed tomography angiogram showed partial stenosis of the right humeral artery and infrarenal abdominal aorta. The patient was treated with corticosteroids (1 mg/kg/day), methotrexate

(20 mg/week) and platelet antiaggregant, with a good clinical evolution.

Five years later, the patient presented with inflammatory polyarthralgia involving the middle joints with pitting edema of the lower limbs.

Biological findings showed lymphopenia and a biological inflammatory syndrome (accelerated erythrocyte sedimentation rate of 99 mm; a CRP of 20 mg/l and anemia of inflammation with haemoglobin of 10.7 g/dl). Anti-nuclear and anti-dsDNA antibodies were positive.

Proteinuria was positive at 4.3g/day and the renal function was normal. Renal biopsy revealed extramembranous glomerulonephritis. The diagnosis of SLE associated with TA was established. The patient was treated with corticosteroids and cyclophosphamide for lupus nephritis with a good evolution.

DISCUSSION

Takayasu arteritis is a granulomatous vasculitis of the aorta and its branches. It is predominantly a disease of young adults in the second and third decades of life, most commonly seen in Japan and South East Asia [1].

As with SLE, there is a marked female predominance of TA. It can be associated with systemic or granulomatous diseases such as ulcerative colitis, Crohn's disease, ankylosing spondylitis, and more rarely SLE and sarcoidosis. These associations are exceptional but seem to be non-fortuitous. The simultaneous development of SLE and TA is rare with only twenty observations reported in the literature. According to some authors, the association between SLE and TA remains unclear since aortitis are sometimes observed during SLE [2-4].

Vascular disease is frequent in patients with SLE (15% of patients), and is associated with high morbidity and mortality. This may be explained by the presence of anti-phospholipid antibodies, diffuse vasculitis or secondary atherosclerosis. It generally involves small vessels, thus aortic damage, aneurysms, occlusions and dissections are very exceptional [5].

On the other hand, vascular damage in TA is due to mononuclear cells and granulomatous infiltration associated with parietal fibrosis leading to aortic obstruction [6].

The diagnosis of SLE is certain in our patient. TA diagnosis was made according to the American College of Rheumatology criteria including (1) age < 40 years; (2) intermittent claudication of the upper limbs; (3) decreased left brachial artery pulse; (4) blood pressure difference >10 mm Hg; and (5) arteriogram abnormality [7].

In our case, the coexistence of both vascular and renal damage was a therapeutic dilemma. However, after the remission of the arteritis, cyclophosphamide was started for the proliferative nephritis. The follow-up showed an improvement of all systemic manifestations.

CONCLUSION

The association of SLE and TA is very rare. Since T cell-mediated immunity plays a major role in both the diseases it may be an overlap of the immune mechanism which leads to the coexistence of both diseases. The absence of specific SLE markers in patients with TA who subsequently develop SLE suggests that the coexistence of these conditions may be coincidental.

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