Aortoarteritis with systemic lupus erythematosus and secondary antiphopholipid syndrome

Sir,

A 39-year- old female patient presented to us with arthritis of small and large joints along with generalized eruptions all over her body [Figure 1] for the past 2 months. She had history of Raynaud's phenomenon and claudication of both upper and lower limbs for the past 15 years. In 1999, she had undergone an amputation of left forefoot due to gangrene. At that time she was diagnosed as a case of non-specific aortoarteritis. In 1990, she had been treated for pulmonary tuberculosis (TB). She suffered from three successive spontaneous mid-trimester abortions within last 3 years. She has no live offspring at present. There was no history of diabetes, hypertension, seizure, swelling of body, chest pain, or abdominal pain. She had received steroids irregularly in the past outside. Examination revealed an average built with mild pallor but no jaundice, edema, or lymphadenopathy. Pulses of left upper limb and popliteal, posterior tibialis, and dorsalis pedis of left lower limb were impalpable. There was no arterial tenderness or bruit. Blood pressure was 130/80 mm Hg in right upper limb; in left-sided limbs it could not be recorded. Skin involvement revealed lesions of a mixed character-some papulosquamous, others annular. There were also oral ulcers and malar rash. Ophthalmic and ENT evaluations were non-contributory. Cardio-respiratory, gastrointestinal, and neurological examinations were normal except slowed mentation. Investigations revealed Hb 11 g, %TC 9000, P 65%, L 30%, E 3%, B 2%, platelet count normal, and erythrocyte sedimentation rate 80 mm/h. Urine RE/ME was normal, fasting sugar 98 mg/dl, and creatinine 0.9 mg/dl. Chest X-ray revealed right upper lobe calcification. Sputum was negative. Mantoux test was positive (12 mm induration). HIV serology was negative. Antinuclear antibody (ANA) was positive at a titer of 1:320 by Hep 2 method, while Double stranded DNA (DSDNA) was negative. Considering the presence of malar rash, oral ulcer, arthritis of more than two joints, and this ANA titer, the diagnosis of systemic lupus erythematosus (SLE) was established (four American Rheumatological association (ARA) criteria fulfilled). History of recurrent fetal losses and gangrene of toes prompted us to go for anticardiolipin antibody. The results were IgM 9.5 μ/ml IgG 62 μ/ml (normal IgM < 5 unit/ml and IgG < 16 unit/ml). It was repeated after 6 weeks when values came out to be 10 and 70 μ/ml, respectively. Prothrombin time (PT) and apTT (Activated Partial thromboplastin time) were also prolonged. Presence of two clinical events (fetal loss and thrombosis) and positive anticardiolipin antibodies on two occasions point to a concomitant secondary antiphospholipid syndrome (APLA).

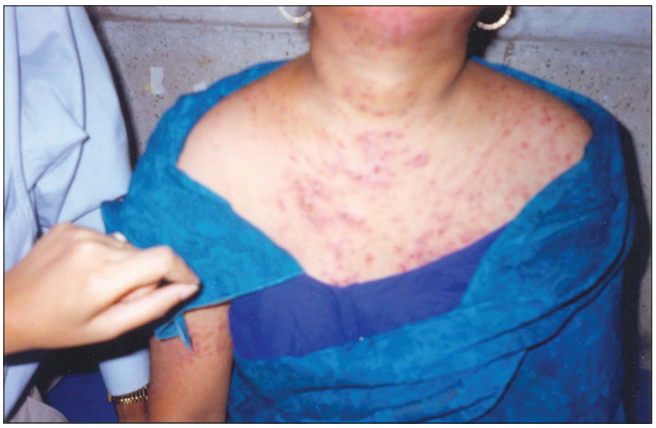

Figure 1: Photograph showing papulosquamous skin lesion

Doppler ultrasonography of peripheral vessels showed diastolic dampening of left brachial artery. Magnetic resonance (MR) angiogram of brain showed bilateral lacunar infarcts. MR angiography of lower limbs revealed narrowing of left femoral artery from mid-thigh extending along popliteal and anterior tibial arteries [Figure 2] and Computed tomography angiogram of aortic arch and its branches failed to visualize left subclavian artery beyond the first part [Figure 3]. Biopsy was taken from left temporal artery that showed accumulation of lymphocytes and few plasma cells in the tunica media with increased vascularity. There was some intimal proliferation as well. A diagnosis of nonspecific aortoarteritis and SLE, and secondary APLA was made. She was given local and oral steroids, hydroxychloroquine, and anticoagulants. Her skin lesions and arthritis gradually subsided.

Aortoarteritis is more prevalent in Japan and Afro-Asian countries. Autopsy incidence in Japan is quoted as 33%.[1] It rarely coexists with SLE. Lupi herrara found ANA in 6% cases of Takayasu arteritis and LE cells in 2% cases.[2] Saxe and Altman reviewed 18 cases of non-specific aortoarteritis with SLE. Patients of both diseases have similar age of onset and female preponderance but presence of anticardiolipin antibody along with this is unique.[3] Most of the cases reported in literature had aortic aneurysms or dissections detected at autopsy or during surgery for dissections.[4]

In our case, five of six ACR criteria for classification of nonspecific aortoarteritis or Takayasu arteritis have been satisfied including age, h/o claudication, asynchronus pulses, and blood pressure and MR angiographic findings. SLE with

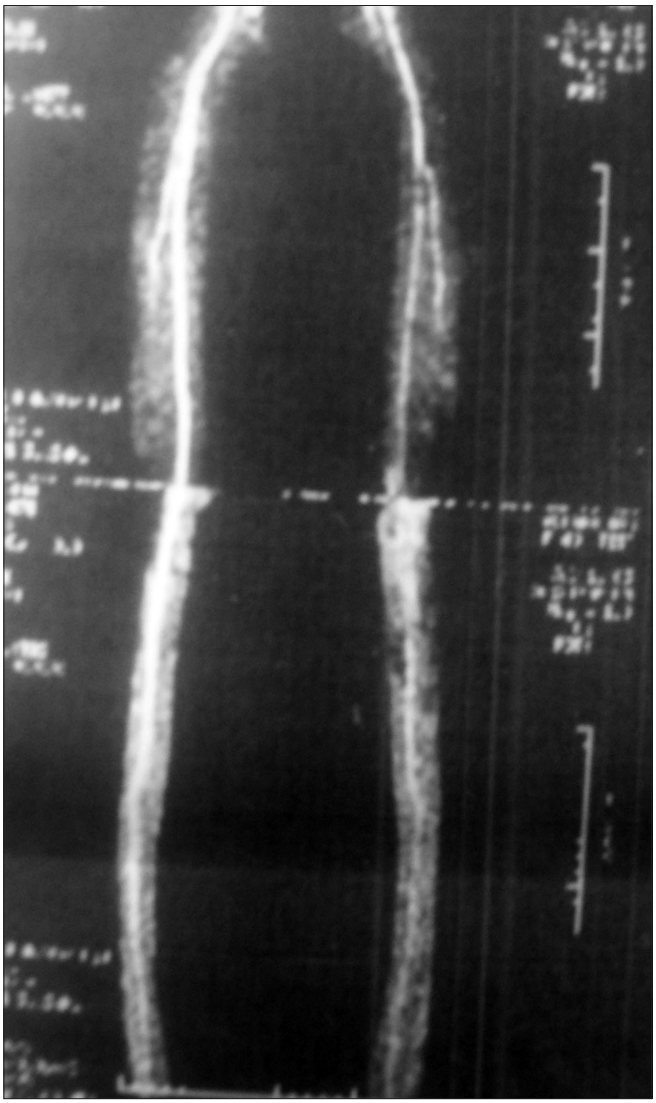

Figure 2: MR angiogram of lower limbs-narrowing of left femoral artery extending down left popliteal and anterior tib

secondary APLA was established on the basis of skin lesions, arthritis, and antibodies. The appearance of rash, arthritis, and abortion was preceded by claudication and gangrene by more than a decade. Possibly antibody work-up for APLA was not performed at that time. However, histopathology done later revealed features suggestive of large vessel vasculitis instead of the classical noninflammatory bland thrombosis so characteristic of APLA vasculopathy. Usually, SLE involves small and peripheral vessels but here involvement spans the whole gamut from small to medium to large vessels. This makes it a rare case report.

Another fascinating point to be noted is the occurrence of pulmonary TB in this patient. Pantell and Goodman had reviewed literature from 1961 to 1981 and found tuberculin test to be positive in 73.3–100% cases and active TB in 0.26–4.2%.[3] However, a large case series from central India

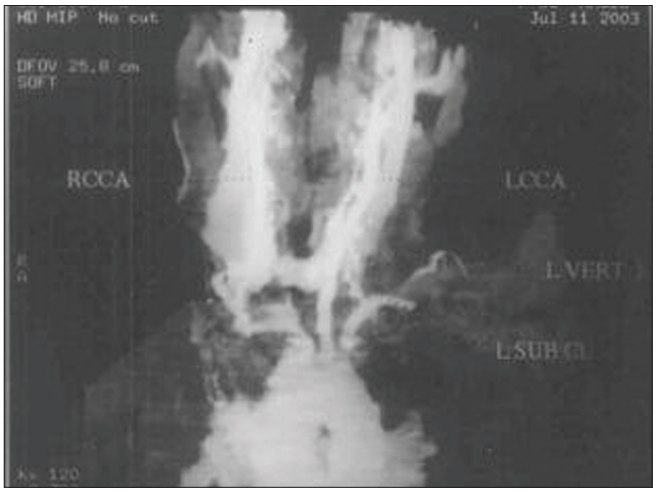

Figure 3: Computed tomography angiogram of aortic arch and main branches—left subclavian artery not visualized beyond first part

has documented mantoux positivity in only 3.03% cases of aortoarteritis.[4] Several researchers from Western and Asian countries have attributed the association as coincidental.[5] A state of immunosuppression due to steroid therapy in our patient could have been the predisposing factor. The natural history of non-specific aortoarteritis is variable. But it usually progresses slowly with the development of hypertension, retinopathy, and aneurysm formation. The coexistence of the above with secondary APLA creates a hypercoagulable milieu that adversely affects the prognosis.[6] Furthermore, as the therapeutic strategies are radically different, recognition of the coexistence of antiphospholipid syndrome and aortoarteritis is of prime importance.[7]

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Access this article online

Quick Response Code:

Website: www.idoj.in

DOI: 10.4103/2229-5178.93483

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