ISSN- O: 2458 - 868X, ISSN–P: 2458 – 8687
Index Copernicus Value: 68 . 16
PubMed - National Library of Medicine - ID: 101731606

International Journal of Medical Science and Innovative Research (IJMSIR)
IJMSIR : A Medical Publication Hub
Available Online at: www.ijmsir.com
Volume – 5, Issue –3, June - 2020, Page No. : 47 - 51

Takayasu Arteritis with Systemic Lupus Erythematosus and persistent triple-positive Antiphospholipid profile – A case report with diagnostic dilemma.

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Citation this Article: Rajdeep Basu, Bhaswar Bhattacharya, Sumantro Mondal, Omkarr De Hazra, Jayaditya Ghosh, Soumitra Ghosh, "Takayasu Arteritis with Systemic Lupus Erythematosus and persistent triple-positive Antiphospholipid profile – A case report with diagnostic dilemma.", IJMSIR- June - 2020, Vol – 5, Issue -3, P. No. 47 – 51.

Type of Publication: Case Report

Conflicts of Interest: Nil

Abstract

Association of Takayasu arteritis (TA) and systemic lupus erythematosus (SLE) is rare in literature and the presence of TA, SLE, and antiphospholipid syndrome (APS) altogether is rarer. A 38-years-old female presented with the asymmetric pulses and blood pressure in upper and lower limbs was diagnosed as having TA by various imaging methods, there were no symptoms on the presentation that could suggest SLE. The clues for SLE were normal leukocyte count with lymphopenia and high erythrocyte sedimentation rate with normal C-reactive protein, which were not usually found TA. Later on, a persistently positive high titer of anti-cardiolipin, anti-β2 glycoprotein and lupus anticoagulant created diagnostic confusion, whether the APS should be considered to start anticoagulation.

Keywords: Takayasu arteritis, Systemic lupus erythematosus, Antiphospholipid syndrome.

Introduction

Takayasu arteritis (TA) is a form of idiopathic large vessel arteritis which involves mainly aorta and its major branches [1]. The systemic lupus erythematosus

(SLE), an autoimmune condition involves multisystem, commonly seen in females. Vascular involvement in SLE is mainly small to medium vessel vasculitis and thrombosis in the association of antiphospholipid syndrome (APS) [2].

Associations of TA with SLE were shown in many literatures. Here, we described a case of TA with SLE with no symptoms of the later and high titer persistently triple-positive antiphospholipid profile without any vascular thrombosis.

Case Report

A 38-year-old non-hypertensive, non-diabetic, euthyroid female from Kolkata, India presented with tingling and claudication over left arm and both legs for the last 2 months, it was progressively increasing, reached its severity within 20 days. Also, the history of Raynaud's phenomena involving all fingers and toes was there for the last 5 months. These symptoms were associated with weight loss (not significant) for the same duration. There was no history of fever, joint pain, skin tightening, oral ulcer, skin rash, hand and foot gangrene, body ache, shortness of breath, cough, chest pain or tightness, unconsciousness, weakness, dizziness. Obstetrics history was not significant and no history of abortion was given.

On physical examination, there was mild pallor, clubbing was present on left-sided fingers and all toes [Figure 1A, 1C, 1D]. Pulses were 90 per minute, regular, normal volume but absent in left brachial, radial, ulnar arteries, feeble in left subclavian artery, absent in both arteria dosalis pedis, feeble in both posterior tibial artery, rests are well palpable and no bruit was auscultated. Blood pressure was 90/60 mmHg at right arm at the supine position, but in other limbs, it was not recordable. There were punctate

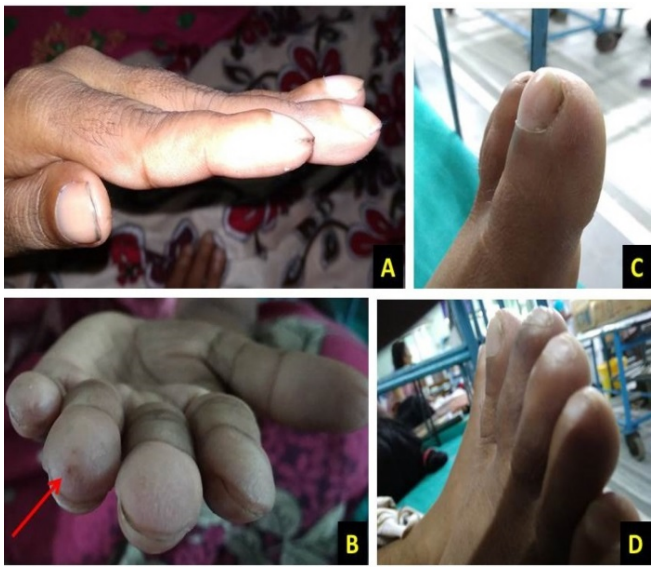

Figure 1: A: Clubbing of the left hand; B: Punctate blackish spots over tips of left-sided fingers (red arrow); C, D: Clubbing of all toes.

Blackish spots over tips of left-sided fingers [Figure 1B] also in the left great toe and no skin tightening.

On routine blood investigations, hemoglobin was 10.0 g%, total leucocyte count, 3800 /mm^3 (N70/L22/E04/M04/B00), lymphocytes were 836 /mm^3 platelets, 1.0 lacs. Liver and renal function tests, serum electrolytes, coagulation profile, urine microscopic examination, all were within normal limits.

On ultrasound Doppler study of both lower limbs, there was monophasic flow with the continuous diastolic flow in right popliteal artery and left anterior tibial artery and very poor flow with absent end-diastolic flow in right anterior tibial, both sided posterior tibial artery and arteria dorsalis pedis.

In CT angiography of lower limb vessels, a long segment (47.6 mm in craniocaudal) narrowing was seen in the mid-part of the right superficial femoral artery at the adductor canal. The right deep femoral artery is dilated communicating with the lateral- superior genicular artery. Also, focal narrowing was there in the

distal part of both right and left ATA and lateral peroneal artery. Both ADP was not well defined. Remaining arteries were normal in course, caliber and branching. Collaterals were present [Figure 2].

MR angiography of the arch of the aorta and its branches showed a severe narrowing of left

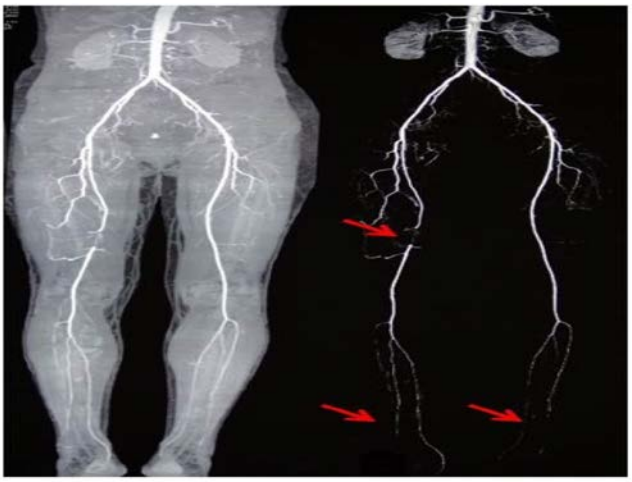

Figure 2: CT angiogram of the lower limbs showing narrowing of the left superficial femoral and distal arteries (red arrows).

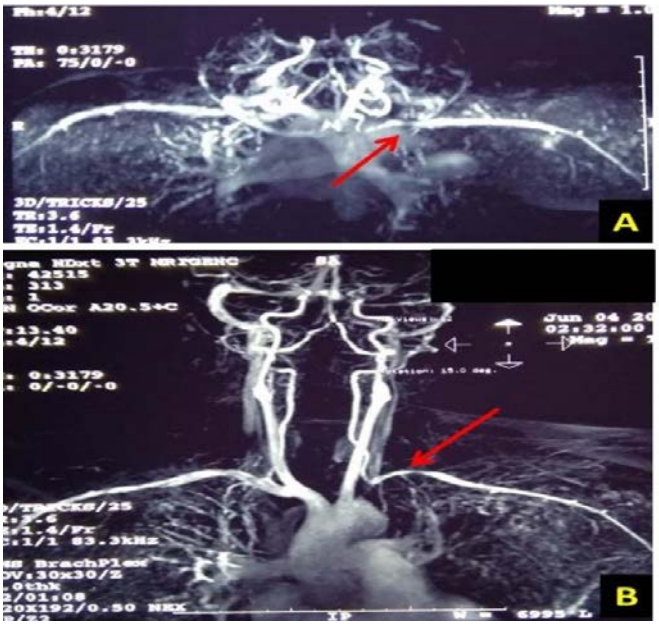

Figure 3: MR angiography of the arch of the aorta and its branches showed a severe narrowing of left subclavian artery, A: Transverse view; B: Coronal view.

Subclavian artery just distal to ostium up to the origin of left vertebral artery and short segment moderate narrowing measuring 21.3mm in the left subclavian artery after the origin of the left vertebral artery with collaterals [Figure 3].

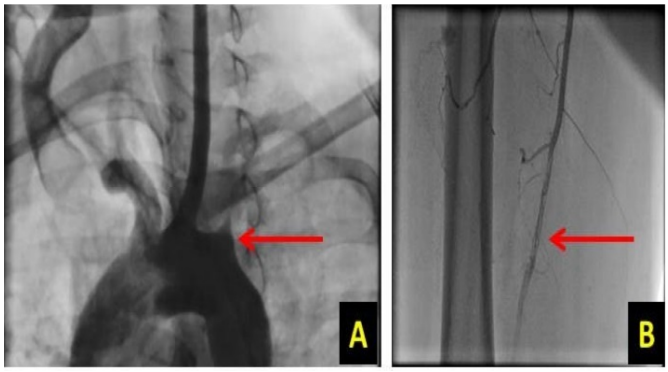

Figure 4: Invasive angiogram showing A: No flow in the left subclavian artery (red arrow); B: Narrowing of the right superficial femoral artery (red arrow).

Invasive angiogram was also suggestive of no flow in the left subclavian [Figure 4A] and narrowing of the right superficial femoral and bilateral distal lower limb arteries [Figure 4B].

Erythrocyte Sedimentation Rate (ESR) was 130mm in 1st hr., but C-reactive protein (CRP) was 0.03 mg/dl; Antinuclear antibody (ANA) – 4+ Homogenous; SS-A native (60 kDa), Ro-52 recombinant, AMA-M2 was positive in ANA profile; anti-ds DNA - 774 IU/ml; Anti-Cardiolipin IgM – 32.8; IgG – 52.9 [normal < 12 PL-U/ml]; Anti-β2 glycoprotein IgM – 200; IgG – 95.17 [normal < 20 RU/ml]; Lupus Anticogulant: screening 59.7 s [normal 28-47 s]; Ratio 1.71confirm 48.7 s [normal 32-46 s]; Ratio 1.33; ANCA – Negative; C3, C4 – Normal; Direct Coomb's test (DCT) – Negative.

Chest X-ray, pulmonary function test and ultrasound whole abdomen were normal.

Stenting of Left subclavian artery was done with the return of normal flow intraoperative. The patient was

started with Aspirin 75mg daily; Methotrexate 15 mg weekly (with Folate supplement); Prednisolone 0.5mg/kg body weight and Nifedipine.

Anti-Cardiolipin, Anti-β2 glycoprotein antibodies were persistently positive even after 12 weeks.

Discussion

The patient had Takayasu arteritis according to The American College of Rheumatology classification criteria 1990 [1], with high disease activity as per Indian Takayasu Clinical Activity Score (ITAS-A) [3], that is 21. So, the patient was given both Prednisolone and Methotrexate before stenting of the subclavian artery to reduce the inflammation. Her disease scenario was also matching with SLE as stated in 2019 European League Against Rheumatism/American College of Rheumatology classification criteria (Total score of 11 with ANA positivity) [4]. But there was no specific clinical symptom for SLE. Suspicion began on the ground of leukopenia and low CRP level with high ESR, where we usually get leukocytosis and increased inflammatory markers in Takayasu [5].

There are many associations TA with SLE cited in previous literature [6-8]. All cases had clinical symptoms of SLE. In our case, the unusual thing was there was no suggestion of SLE until we got the blood reports. SLE has the propensity to cause vasculitis but it is of small to medium vessels in majority of cases in form of fibrinoid degeneration, thickening of the intima, thrombosis, and sclerosis, large vessel vasculitis in SLE is rare [2].

In the question of antiphospholipid profile positivity (Anti-Cardiolipin, Anti-β2 glycoprotein and Lupus anticoagulant, persistent triple positive) and the starting of anticoagulant, there was neither vascular thrombosis demonstrated nor any pregnancy morbidity, which are the main clinical aspects of APS. In some case reports it was shown that TA may be associated with APS [9-11], also two reports shown TA SLE APS, all three were present together [12, 13]. We deferred the diagnosis of APS in our patient, as the APS profile may be positive in SLE and even with TA [14]. We followed the case more than 1 year without any anticoagulation and luminal narrowing was still present in lower limb arteries (as described earlier) on repeat vascular imaging, it did not show any thrombosis even then.

Unusual arterial involvement can be found in TA, like superficial femoral and other lower limb arteries [15, 16]. Thus, we can explain our case. The cause of such associations is not clearly understood yet, T cell mediated autoimmunity may be an explanation for these [8].After subclavian artery stenting improved left upper arm claudication significantly but pulses are still absent in distal lower limb arteries. At present, the patient can continue her daily activities with ease.

Conclusions

So, though there is a list full of mimickers of vasculitis and many rheumatological diseases share the features of one another, careful categorization and classification are required for diagnosis and management.

Acknowledgment: We are very much thankful to Dr. Saroj Mondal, Department of Cardiology, Seth Sukhlal Karnani Memorial Hospital and Professor Dr. Parasar Ghosh, Department of Rheumatology, Seth Sukhlal Karnani Memorial Hospital, for their constant guidance and support in managing this patient.

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