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Medicine

TAKAYASU ARTERITIS VERSUS SLE VASCULITIS- TWO SIDES OF THE SAME COIN?'

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ABSTRACT **Introduction:** Takayasu Arteritis and Systemic Lupus Erythematosus affect the same age group and gender. Yet the former affects the large arteries, while the later affects the small blood vessels. Thus overlap of the two in the same patient is a rarity. **Case:** We present the case of a young woman who presented with hemiplegia and was first diagnosed as a case of Takayasu Arteritis but on further evaluation found to have Systemic Lupus Erythematosus also along with antiphospholipid antibody positivity. We also search the literature and explore and discuss various aspects of the case. **Conclusion:** We conclude that this rare overlap may be increasingly reported as awareness about it increases among clinicians. Also noted is the higher number of such cases reported from Japan, which also has higher geographical prevalence of Behcet's disease.

KEYWORDS : Intracranial Takayasu, Takayasu Arteritis, Large Vessel Vasculitis, Systemic Lupus Erythematosus

INTRODUCTION:

Takayasu Arteritis (TA) is a large vessel vasculitis mostly prevalent in young women affecting the aorta and its branches. Systemic Lupus Erythematosus (SLE) is a multisystem autoimmune disorder found mostly in child bearing age women and with mostly small vessels being affected. Both have the same age distribution. Yet overlap of these two divergent conditions has been rarely reported in literature. We present a case of a young woman who had both of the above as well as anti phospholipid antibody (APLA) positivity and presented with infarction of the brain resulting in hemiplegia. We also discuss the various aspects related to this case and their interpretation.

Case Report

A 41-year lady presented to medical emergency with complaints of sudden onset headache associated with vomiting, followed by left sided weakness for a day. She gave no history of loss of consciousness, seizures, or blurring of vision. General examination revealed a fully conscious, and oriented patient with left hemiplegia. However, her radial and brachial pulses were absent on the left side with diminished left carotid pulse. Non contrast CT of the brain revealed infarct.

Chest examination revealed bilateral pleural effusion. Examination of the heart and abdomen did not reveal any abnormality. Routine investigations revealed haemoglobin (Hb) of 9.3 g/dL, normal leucocyte and platelet counts, and renal and liver function tests. Her inflammatory markers Erythrocyte Sedimentation Rate (ESR) and C-reactive protein (CRP) were high at 52 mm in 1st hour and 10 mg/dL respectively.

The pleural fluid was exudative in nature with normal adenosine deaminase levels (ADA). Keeping in view the age of the patient, samples were sent for Anti-Nuclear Antibody (ANA) and Anti-Phospholipid Antibody (APLA) testing, and history reviewed again, which revealed polyarthralgia, malaise, fatigue, oral ulcers, and photosensitivity for the preceding 6 months. ANA was positive by both ELISA as well as indirect immunofluorescence (IIF), with ANA Profile revealing positivity for Anti Ro 52 and 60. APLA came out to be positive for anti cardiolipin (aCL-IgG), and anti β2 glycoprotein 1 (anti β2-GP1-IgG). Arterial Doppler of the left arm showed narrowing of the subclavian arteries, complete obstruction of the brachial, radial, and ulnar arteries.

Doppler of the Carotids showed diminished flow in the right common, external and internal carotids. CT Angiography revealed circumferential mural thickening of the proximal left subclavian artery as well as infra renal descending aorta, with luminal narrowing of proximal superior mesenteric artery (SMA), inferior mesenteric artery (IMA), and both the renal arteries (along with an accessory renal artery on the right side). CT Angiogram of the brain vessels revealed irregular beaded appearance of the M1 segment of the right middle cerebral artery (MCA). [Figure 1]

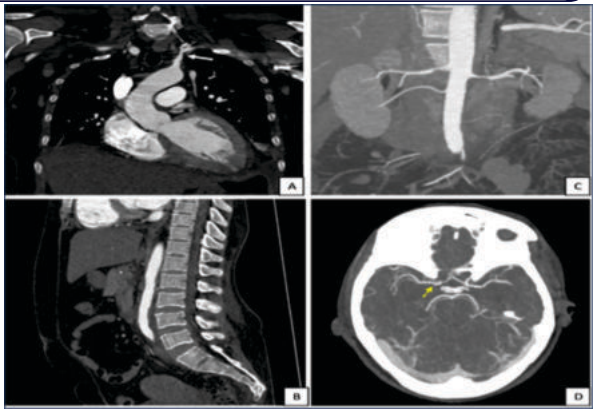

Figure 1- A) Coronal MIP CT image showing short segment circumferential wall thickening with luminal narrowing in proximal left subclavian artery. B) Sagittal MIP CT image showing circumferential symmetric hypodense wall thickening in infra renal abdominal aorta causing mild luminal narrowing. C) Coronal MIP CT image of abdominal aorta showing short segment areas of liminal narrowing in bilateral proximal renal arteries. Accessory right renal artery is noted supplying lower pole of right kidney. D) Axial MIP CT image showing irregular outline of M1 segment of right MCA (yellow arrow)-beaded appearance

Thus the final diagnosis was kept as Takayasu Arteritis with Systemic Lupus Erythematosus overlap with APLA positivity. She was started on Hydroxycholoroquine, steroids, and azathioprine along with aspirin. APLA remained positive on repeat testing after 12 weeks.

DISCUSSION

The association of TA with SLE is rare. Among the earliest reports was a series of 19 cases by Saxe PA and Altman RD[1], out of which five were from Japan, one each from India and Czechoslovakia, and three from Poland, thus showing a preponderance in Eastern Europe, and Japan. In recent times, the numbers of such cases reported in India has increased[2-4], and the numbers are likely to increase as awareness grows about this rare overlap.

Of interest is the question of whether the APLA positivity is secondary to SLE or to TA. APLA is commonly known to be positive in about 30%-50% patients of SLE. Reports of APLA in TA though are far and few[5-7], but almost a sixth of patients of TA had anti β2 GP1 or Lupus Anti Coagulant (LAC) without aCL positivity in a study by Senturk EF et al [8].

The impact of APLA positivity in a case of TA has contrasting views. The study by Jordan N et al [9] found vascular complications to be more prevalent in patients of TA with APLA positivity, especially those with

LAC. This is in contrast with the more recent study by Senturk EF et al [8] which found no differences in disease related complications between those positive and negative for these antibodies.

Another aspect which merits attention is the large vessel vasculitis (LVV) caused by SLE itself, reports of which are published intermittently[10-12]. Aortic dissection, aneurysm, and dissection have been described in SLE, with prolonged steroid use and inflammation being contributory factors. However, in our case, the symptomatology of SLE was of duration of 6 months only, and thus it is more likely that SLE developed in a case of pre-existing TA.

Our case has another rarity of affection of intracranial vessels in TA. There are few other case reports of such intracranial affection[13–19]. Intracranial vessel affection is missed if they are clinically silent as they are not evaluated in absence of manifestations.

Thus we conclude that our case is a rare manifestation of overlap of TA with SLE with APLA positivity, and with affection of intracranial arteries. With increasing awareness about this rare overlap among clinicians, more such cases are likely to be reported. We also note that many of the reported cases are from Japan, which also has a higher geographical prevalence of Behcet's disease.

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Conflicts of Interest: None

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