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Case Reports

Systemic Lupus Erythematosus with Takayasu's Arteritis - A Rare Co-existance

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Abstract:

We report the case of a 30-year-old normotensive, nondiabetic lady diagnosed case of Systemic Lupus Erythematosus(SLE); who came with the complaints of limb claudication. After clinical and lab evaluation she was diagnosed as a case of Takayasus Arteritis along with SLE. Though the co- existence of SLE and Takayasu's Disease is very rare, we should search for the development of arterial occlusive diseases in SLE cases if the patient has got suspicious symptoms.

Key words*: Systemic lupus erythematosus, Takayasu's arteritis*

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Introduction:

SLE is a complex heterogeneous autoimmune disease most frequently affecting the small and medium sized vessels.[1,2] Takayasu's arteritis, also known as 'Aortic arch syndrome' or 'Pulseless disease' is a form of large vessel granulomatous vasculitis[3] with massive intimal fibrosis and vascular narrowing[4]. Association of SLE with Takayasu's arteritis is very rare. Only 22 published case reports are available and of them, four had associated antiphospholipid antibody syndrome. Here we report a case of SLE, which later on diagnosed as Takayasu's arteritis along with SLE.

Case Report:

A 30-year-old lady diagnosed case of SLE, presented with progressive disability to perform the household activities and pain with numbness in the upper extremities for 5 months. The problem was more prominent in the left side, usually worsened on exertion and relieved by taking rest. For SLE, she was taking Hydroxychloroquine and Azathioprine orally in an irregular manner. She has two healthy children and does not have the history of any fetal loss.

On examination, the patient had butterfly rashes on her cheeks. Her radial, brachial, popliteal, arteria dorsalis pedis of both sides were absent. Femoral pulses were present but very feeble. Blood pressure was undetectable in both the upper and lower limbs. Carotid bruit was present. An early diastolic murmur was audible in left lower parasternal area, best heard with patient sitting and bending forward and breath hold after expiration. Bilateral renal bruit was present.

Her ESR was high (84 mm in first hour). ANA, Anti-ds DNA and Anti-Sm antibody were positive with negative IgM and IgG Antiphospholipid antibodies. Serum C_3 was decreased(0.56gm/dl) with normal C_4 level. Echocardiogram revealed dilated ascending aorta (40 mm), mild Aortic Regurgitation and ectatic and narrowed descending aorta. Duplex study of the arch of the aorta revealed 100% occlusion in Common Carotid and Left Subclavian artery along with coarctation of the descending thoracic aorta. So the finding was suggestive of Takayasus Arteritis with Left Subclavian Steal Syndrome and Acquired Coarctation of Descending

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Aorta.

Coronary angiogram revealed 3-vessel- disease with 30-40% stenosis in left anterior descending artery with an impression in favor of minor LAD [Fig-1]. Peripheral angiogram showed aneurysmal and narrowed Left Subclavian artery and 100% occlusion of Right Subclavian artery. It also revealed 100% occlusion of Right Renal artery and 80% occlusion in the middle of descending aorta[Fig-2]. The impression was

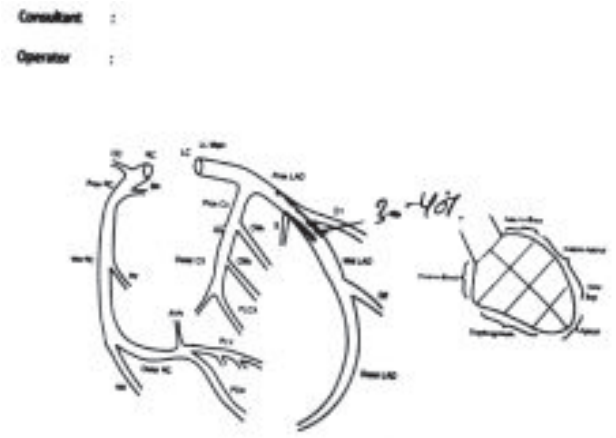

Figure 1: *Coronary Angiogram*

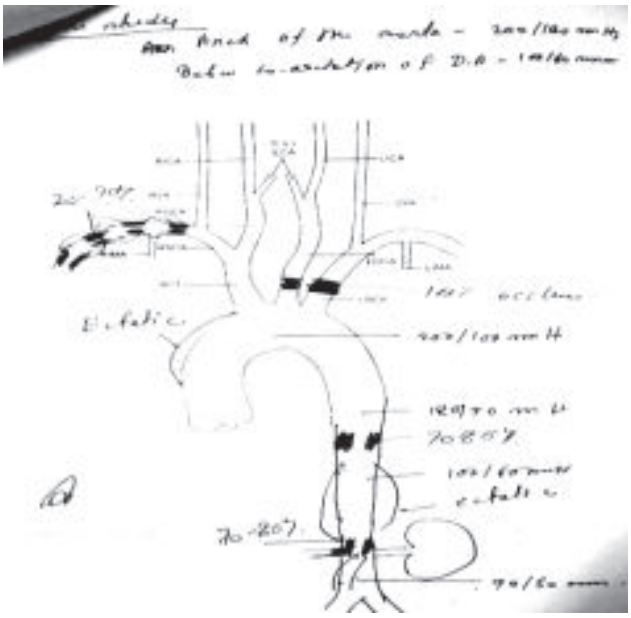

Figure 2: *Peripheral Angiogram*

Takayasus Arteritis Type V.

The patient was treated with steroid and beta blocker along with Hydroxychloroquine and was discharged. Follow-up after four weeks revealed that her weakness and fatigue was improved and arteria dorsalis pedis pulsation of both sides were present but feeble.

Table I

ACR diagnostic criteria for Takayasu's Arteritis.[6]

	Criteria	Definition
1	Age at disease onset <40 years	Development of symptoms or findings related to Takayasu's arteritis at or before 40 years
2	Claudication of extremities	Development and worsening of fatigue and discomfort in muscles of 1 or more extremity while in use, especially upper extremities
3	Decreased brachial artery pulse	Decreased pulsation of one or both brachial arteries
4	Blood pressure difference	Difference of >10 mm Hg in systolic blood pressure
5	Bruit over subclavian arteries or aorta	Bruit audible in auscultation over 1 or both subclavian arteries or abdominal aorta
6	Arteriogram abnormality	Arteriographic narrowing or occlusion of the entire aorta, its primary branches or large arteries in the proximal upper or lower extremities, not caused by arteriosclerosis, fibromuscular dysplasia or similar causes; changes are usually focal or segmental

Table II

Angiographic classification of Takayasu's arteritis.[7]

Type	Vessel Involvement
I	Branches from the aortic arch
IIa	Ascending aorta, aortic arch and its branches
IIb	Ascending aorta, aortic arch and its branches, descending thoracic aorta
III	Descending thoracic aorta, abdominal aorta, and/or renal arteries
IV	Abdominal aorta and/or renal arteries
V	Combination of IIb and IV

Discussion:

Takayasu's arteritis or pulseless disease is an inflammatory and stenotic disease of medium and large sized arteries affecting mainly the young females and characterized by a strong predilection for the aortic arch and its branches. It is a very uncommon disease with an estimated annual incidence of 1.2-2.6 cases per million.[5] Our patient met most of the diagnostic criteria of Takayasu's arteritis set by the American College of Rheumatology[6], according to which at least three out of six criteria should be met for confirmation of the diagnosis [Table-I]. According to angiographic involvement, there are five types of Takayasu's arteritis[7][Table-II]. Our patient had type V Takayasu's disease, though according to Hata A et al, Asians usually have type III of this disease.[8] It usually progresses through three stages. In the first stage, patient complaints of formless symptoms; like fatigue, malaise, fever etc. This stage is considered to be prevasculitic. The second stage is due to vascular inflammation, like claudication pain, palpitation, headache TIA, stroke amaurosis fugax, double vision etc. The third stage is the burnt-out stage when fibrosis sets in and usually associated with remission of symptoms.[9]Our patient was diagnosed as a case of SLE few years back and this time she came with rash, hypocomplementemia with positive ANA, Anti-ds DNA and Anti-Sm antibodies fulfilling the SLE diagnostic criteria set by Systemic Lupus International Collaborating Clinics(SLICC).[10]

Vascular involvement is quite frequent in SLE and represents the most frequent cause of death[1]. It may manifest in nearly 56% of SLE cases throughout their life. Vasculopathy may be directly related to the pathogenesis of the disease, presenting as acute or subacute manifestation of SLE; as for example- antiphospholipid syndrome and lupus vasculitis. It may also develop as an important co morbidity, like steroid-related atherosclerosis.[11]In most of the cases the small and peripheral vessels are involved. In some cases, medium-sized arteries are involved, which are associated with more frequent thrombotic events and higher morbidity.[11,12]In SLE, involvement of aorta is extremely rare.[13] In our case who was a diagnosed case of SLE, we found the typical angiographic evidence of Takayasu's arteritis effecting the great vessels.

In Takayasu's disease, there is granulomatous infiltration of aortic tissues by killer cells especially cytotoxic T-lymphocytes.[14]Absent or diminished pulsation is due to thickened arterial wall or thrombosis of aortic branches, which if left untreated, may lead to gangrene and tissue loss.[15] In SLE, autoantibodies and immune complexes trigger both B and T cell activation.[16] Since T-cell mediated immunity plays an important role in both the disease , there may be an overlap of immune mechanism that leads to the co-existence of the disease.[17]

Conclusion:

SLE and Takayasu's arteritis both are disease of immune dysfunction. Combination of these two are very rare, but not impossible. So if a young female with SLE comes with any abnormal features like limb claudication or unusual fatigue, treating physician should crucially search for the presence of any co-existing illness. Any undue delay may lead to life-threatening events.

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