Case Report

Vasculitis: Many Faces and One Disease

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INTRODUCTION

Vasculitis encompass a group of heterogenous disorders with diverse clinical presentations, it involves the inflammation of the vessel wall of different size: large, medium size and small vessel vasculitis. There is no clear cut demarcations regarding the size of the inflamed vessels.

Case report:

A female patient 23 years old, from Tanta, Egypt, single, a manual worker and she has no special habits of medical importance.

Complaint:

Chest pain & Left upper limb pain of 1.5 year ago.

HPI:

The condition started 1.5 year ago by gradual onset and intermittent course of vague central chest pain referred to the left upper limb and sometimes to the back, it was accompanied by grade II dyspnea. There was no palpitation or syncopal attacks. The patient sought medical advice at a cardiologist diagnosed her as having rheumatic fever and she has been prescribed long acting penicillin but without any improvement.

She developed pain at left upper limb which extends from the shoulder to the finger tips, this pain increased on movement and on raising the arm this was associated with coldness and tingling sensation of the hands.

Five months later she developed recurrent headache and upper abdominal dull aching pain, sometimes colicky, referred to back, not related to meals for which she received daily NSAIDS with mild improvement. She sought medical advice again at a vascular surgeon who asked for upper limb duplex which revealed stenosed left subclavian artery. The patient was referred to the rheumatology unit at internal medicine department, Tanta University to be diagnosed and managed.

On examination:

She was fully conscious, alert, oriented of average body built, looks well with no special facial expression and no special decubitus.

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Vital signs:

- **Pulse:** On the Rt. side it was 80/minute, regular, of average force &volume no special character, arterial wall is not felt. On the Lt. side there is absent radial pulse, left brachial pulse but there is normal dorsalis pedis pulse bilaterally.
- **Blood pressure:** 140/90 on right upper limb absent on left upper limb, 150/90 in LL.
- **Respiratory rate:** 18 cycles /minute
- **Temperature:** 37 °C

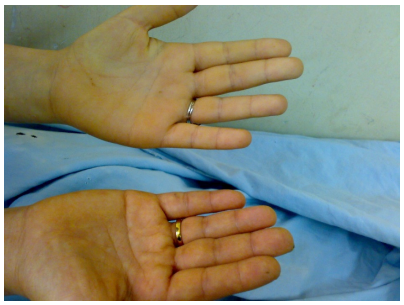
Head & Neck:

- Trachea is central, no enlarged thyroid gland; no congested neck veins.
- Bilateral bruit sound over Rt. & Lt. common carotid arteries with palpable thrill over right carotid artery.

Chest and heart examination are free

Abdomen: lax abdomen, no organomegaly nor ascites or bruit.

Extremities: left upper limb had pale fingers and hand, with slight coldness and ischemic atrophy of the thinner and hypothiner eminence of the palm delayed capillary filling, otherwise no skin lesions or lower limbs oedema.



Pale left hand with discrepancy in size between both hands; the Lt hand is smaller in size (ischemic atrophy).

Musculoskeletal examination:

No joint swelling, tenderness or limitation of movements

Neurological examination:

Motor system: power & deep reflexes were normal.
Sensory system: parathesia at left upper limb from fingers tips to the elbow.

Laboratory work up:

FBG: 78mg /dl BUN: 31 mg/dl
S. Creatinine: 0.9 mg /dl
CBC: HB: 10.10gm /dl WBCs: 7.200
Platelets: 440,000
L.F.T: Biliurbin: 0.7mg/l
SGPT: 26 u/l SGOT: 28 u/l
S. Albumin: 3.5 gm/l ESR: 95 1st hr, 104 2nd hr
CRP: + ve 48 mg% RF: -ve
ANA: -ve Anti ds DNA: -ve
C and P ANCA: -ve HBSAg –ve, HCVab -ve, HIV-ve

Chest X ray: Normal.

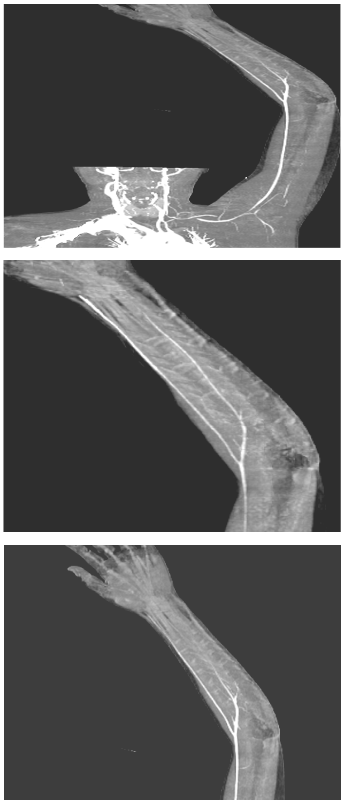
X ray of cervical spine (AP &Lateral):
Normal with no bony cervical rib.

Echo: Normal study.

Abdominal U/S : Normal study
Duplex on Lt. upper limb: Severe ischemia of the left UL below the level of the Lt. subclavian artery.

CT angiography on the left upper limb:

Significant stenoses of the first and second part of the Lt. subclavian artery ending with a totally occluded segment in the third part with reconstitution of the axillary artery through collateral networks, another significant stenoses is seen at the end of left axillary artery measuring 5 mm.



CT angiography of left upper limb showing marked stenoses of the left subclavian artery.

Duplex on both carotid arteries:

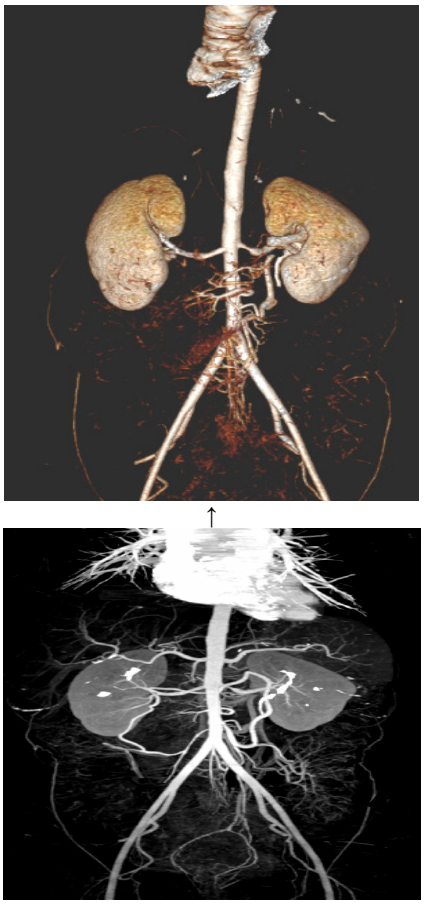
Marked thickened intema at both sides with 80% stenoses at the left side & 60% stenoses at the Rt. side with high peak velocity.

Multislice CT angiography on the chest:

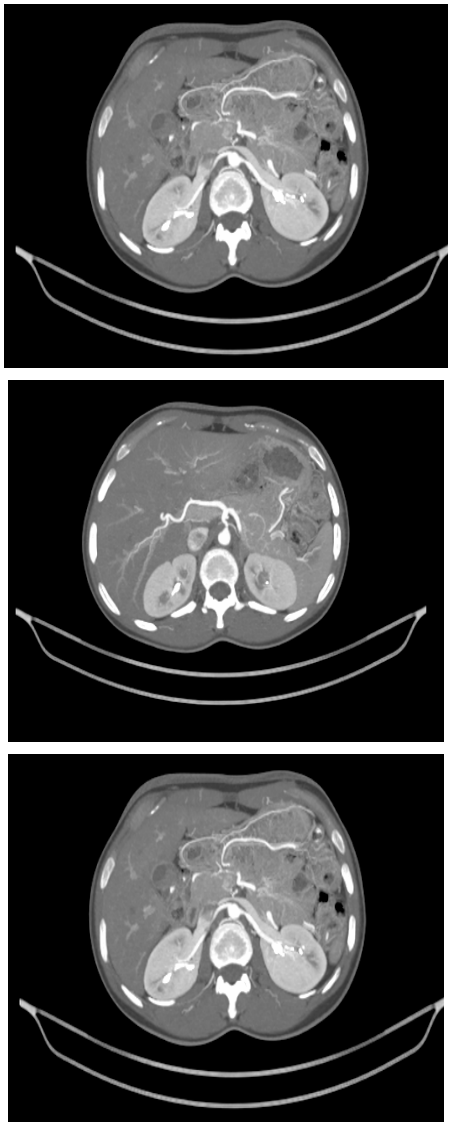
Circumferential uniform wall thickness of the major branches of the aortic arch, descending thoracic aorta with significant narrowing mainly affecting the left side arteries; Lt. common carotid and Lt. subclavian arteries.

CT angiography on abdomen:

Circumferential uniform wall thickness of the major branches of abdominal aorta with totally occluded origin of the superior mesenteric artery with filling of the distal segment through the collaterals from dominant inferior mesenteric artery, segmental left renal artery stenoses.



CT abdominal angiography showing stenosed Lt renal artery



CT angiography showing totally occluded superior mesenteric artery.

DISCUSSION

We diagnose this patient as a case of systemic large vessel vasculitis of Takayasu subtype depending on the following criteria:

1- Female <40 year.
2- Left UL claudication pain (ischemic pains due to stenosed vessels.
3- Unequal pulse & blood pressure between both arms.
4- Bruit over Rt. and Lt. carotid arteries.
5- Significant stenoses of aorta and its main branches proved by CT angiography.

Takayasu's is a large vessel vasculitis that involves the inflammation of the aorta and its main branches, this largely explains most of the clinical manifestations of the disease. The pattern of vessel wall inflammation is a granulomatous vasculitis, it ends with massive intimal fibrosis and vascular narrowing, resulting in significant reduction in the blood flow to certain limb/organ.[1] Takayasu's arthritis was 1st described in 1908 by Japanese ophthalmologist Mikito Takayasu during the Annual Meeting of the Japan Ophthalmology Society. Takayasu was described as a peculiar "wreathlike" appearance of the retinal vessels[2,3]

The American College of Rheumatology (ACR) has established a classification criteria in 1990 for Takayasu's vasculitis. The presence of three or more of these criteria, yields a diagnostic sensitivity of 90.5 % and a specificity of 97.8%.

Takayasu's classification criteria:

1- Age of onset < 40 years.
2- Claudication of the extremities: Development and worsening of fatigue and discomfort in muscles of one or more extremity while in use, especially the upper extremities.
3- Decreased pulsation of one or both brachial arteries.
4- Difference of at least 10 mm Hg in systolic blood pressure between both arms
5- Bruit over one or both of subclavian arteries or the abdominal aorta.
6- Arteriographic narrowing or occlusion of the entire aorta, its primary branches or the large arteries in the upper or lower extremities that is not due to arteriosclerosis, fibromuscular dysplasia or other causes.(14)

Treatment Plane

The goal of therapy in Takayasu's vasculitis is to induce disease remission; reducing systemic and vessel wall inflammation and to suppress autoimmunity.

1. Patient education:

We've explained the condition and the possible side effects to the patients. Eating a healthy diet, including fresh fruits and vegetables, whole grains, and lean meats and fish, drinking plenty of water while limiting salt, sugar and alcohol intake. Calcium and vitamin D rich diet. Exercise regularly; regular aerobic exercise, such as walking, can help prevent bone loss and improve the collateral circulation, high blood pressure and diabetes. Avoid smoking , to reduce the atherosclerotic risk of the already stenosed and injured blood vessels.

2. Medications:

The patient has been given pulse methylprednisolone 1 gram daily for 3 days, followed by 40 mg oral prednisolone to be tapered to 20 mg daily over a month. In addition to 6 cycles 1gm cyclophosphamide monthly followed by maintenance methotrexate 25 mg SC /week. Aspirin 75 mg/day and Trental 400 mg /day.

Clinically she was partially improved regarding her ischemic claudications pains. BP: RT UL 130/70 & in LL 140/70, ESR: 25 1st hr.

We refer the patient to vascular surgery, to assess for the possibility of surgical bypass operation or stenting for the stenosed arterial segments.

Now she has been prepared for angioplasty operation for her left subclavian artery. She is in need for stenting for her left stenosed renal artery. While the occluded superior mesenteric artery couldn't be operated upon.

The etiology of Takayasu's arteritis is unknown. The worldwide incidence is estimated to be 2.6 cases per million per year. Although the disease has a worldwide distribution, it is observed more frequently in Japan; about 100-200 new cases are registered each year. The disease is common in Asian countries: Korea, China, India, Thailand, Singapore. In United Kingdom, the annual incidence is 0.15 case per million. While the incidence in the United States is estimated to be 2.6 persons per million annually.[4]

Approximately 80% of patients with Takayasu's arteritis are women. Females are about 8–9 times more likely to be affected than males. In India, the female-to-male ratio is as low as 1.6:1. Most patients with Takayasu's arteritis are aged between15-30 years fewer than 15% of cases present in individuals older than 40 years.[5,6]

The symptoms that clinicians encounter depend on when the patient presents; most of the patients present late, mainly due to mis / delayed diagnosis.

The underlying pathologic process is inflammatory vasculitis, several etiologic factors having been proposed, including; spirochetes, mycobacterium tuberculosis, streptococcal organisms and circulating antibodies due to an autoimmune process. Genetic factors may play a role in the pathogenesis.

Takayasu's arteritis progresses through 3 stages:[7]

The first stage is an early systemic stage during which the patient may complain of constitutional symptoms (e.g., fatigue, malaise, giddiness, fever). This stage is considered to be prevasculitic.

The second stage is the vascular inflammatory stage, there is significant vessel wall inflammation and infiltration with inflammatory cells with vascular stenoses / aneurysms and vascular pains tend to occur.

Symptoms characterizing the vascular inflammatory stage include: fatigue, fevers, malaise, pain in extremities and joints, dyspnea, palpitations, headaches, rash (erythema nodosum or a lupus like butterfly rash, which can be photosensitive), hemoptysis, ulceration, and weight loss. A single case of thoracic and lumbar spine pain has been reported. Symptoms of vascular insufficiency include arm numbness, claudication pains in the legs (rare) or arms, blurred vision, double vision (which can be posture dependent), amaurosis fugax, stroke, transient ischemic attacks, hemiplegia, seizures and paraplegia.

The third stage is the burned-out stage, when fibrosis sets in and generally it is associated with remission. This stage does not occur in all patients, and even in patients who are in remission, relapses can occur. Presumably, the burned-out stage manifests with minimal symptoms, but little supportive evidence is found in the literature.[8-13]

Prognosis

Takayasu's arteritis is associated with substantial morbidity and may be a life-threatening disease. Its course usually extends for many years, with varying degrees of activity. Approximately 20% of patients have a monophasic and self-limited disease. In others; Takayasu is progressive or relapsing/remitting and requires prolonged immunosuppressive treatment.[(15)]

Angioplasty or vascular surgical procedure e.g. bypass grafting may be needed in certain situations, this may involve angioplasty (widening of a narrowed or blocked segment), with or without placement of stent to keep and ensure blood flow[(16)].

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