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Case Report

Takayasu arteritis: In a middle-aged Sri Lankan male - A case report

[1]Gayathri Gnanaruban, [1]Sivansuthan S

[1] Teaching Hospital Jaffna

Abstract

Takayasu arteritis is a vasculitic condition which affects large and medium sized vessels primarily aorta and its main branches. It is a rare condition affecting females more than males between 10 and 40 years of age with spectrum of clinical presentation. Here we present a case of 58 year old man who presented to us with non-specific gastrointestinal symptoms who was eventually diagnosed with Takayasu arteritis.

Keywords

Takayasu arteritis, vasculitis

Introduction

Takayasu arteritis (TAK) is classified into large and medium vessels vasculitis with the predilection for aorta and its major branches (1). Its aetiology is still unknown but proposed as autoimmune process associated with antibody following infections (2). It is a granulomatous inflammation of the vessel wall leading to stenotic, occlusive or aneurysmal changes in the vessels (3,4). Worldwide incidence of TAK estimated 2.6 cases per million per year with more case detection from Asia, Africa, central and south America (4). About 80 percentage of the patients with TAK are females and the mean age of onset is approximately 30 years (3,4). Because of the rarity of this condition diagnosis of TAK is often delayed.

Case report

A 58-year-old male who did not have any significant co-morbidities in the past, presented with one week history of watery stools mixed with blood and mucus. He did not have fever or vomiting. There was no contact history of diarrhoeal illness. He had consumed food from outside home before the onset of illness. He also had reduced appetite and significant unintentional weight loss of 2 stones over the past two years. He also had epigastric abdominal pain for the last two years which was colicky in nature, radiating to back, aggravated by food intake and relieved by leaning forward. He had belching and heartburn. He needed to open his bowel following each meal and he used to pass loose stools without blood or mucus. He did not have headache, joint pain, skin rashes, skin ulcers, oral or genital ulcers, chronic cough with sputum or history of tuberculosis in the past. He did not have chest pain, dyspnoea or limb claudication. He had history of alcoholism and smoking which he gave up four and two years before respectively. There was no history of high-risk sexual behavior. His past medical history is not significant otherwise. He underwent right side inguinal hernial repair 30 years back. He was a fisherman, married and has two children.

On examination, he looked well. His body mass index was 19.27 Kg/ m^2. He was neither pale nor icteric. There was no clubbing, lymphadenopathy, skin ulceration, mucosal ulceration or ankle edema. Capillary refilling time was less than two seconds. On cardiovascular system examination, pulse rate was 56 beats per minute which was regular with good volume. Distal pulses in right side limb namely brachial, radial and dorsalis pedis were not palpable. Blood pressure discrepancy was noted in bilateral upper limb with the blood pressure of 77/ 59 mmHg in right arm and 100/60 mmHg in left arm with the arterial oxygen saturation of 90 % on right arm and 99% on left arm. On auscultation bruit was heard over both subclavian arteries. Precordial auscultation findings were normal. His abdomen was soft without organomegaly, he had mild tenderness over umbilical region. Respiratory system and neurological examination were unremarkable.

On investigation his complete blood count revealed WBC count of 14.86 x 10^9/L, haemoglobin of 13.3 g/dL and normal platelet count. ESR was 72 mm/first hour and CRP was 11.5 mg/L. Renal function, serum electrolytes and serum calcium were normal. Liver function was otherwise normal except mild elevation of ALT which was compatible with grade 1 fatty liver on ultrasound abdomen. Urine full report, stool full report, urine and stool culture were normal. VDRL and Mantoux test were negative. Ultrasound scan of the abdomen revealed normal size kidney in both side with normal architecture. 2D echo was normal. Upper GI endoscopy revealed severe gastritis with mild duodenitis. Chest X-ray was normal. Plain X-ray abdomen did not reveal pancreatic calcification. CT angiogram of thoracic and abdominal aorta

Corresponding Author: Gayathri Gnanaruban. Email: gayathri.kugangananthan@gmail.com . https://orcid.org/0000-0003-1006-8781,
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and its branches revealed subtle irregular thickening of ascending aorta, narrowing of right brachiocephalic trunk and left common carotid artery without complete obstruction, and stenosis at the origin of celiac axis, superior and inferior mesenteric arteries. (Figure 1)

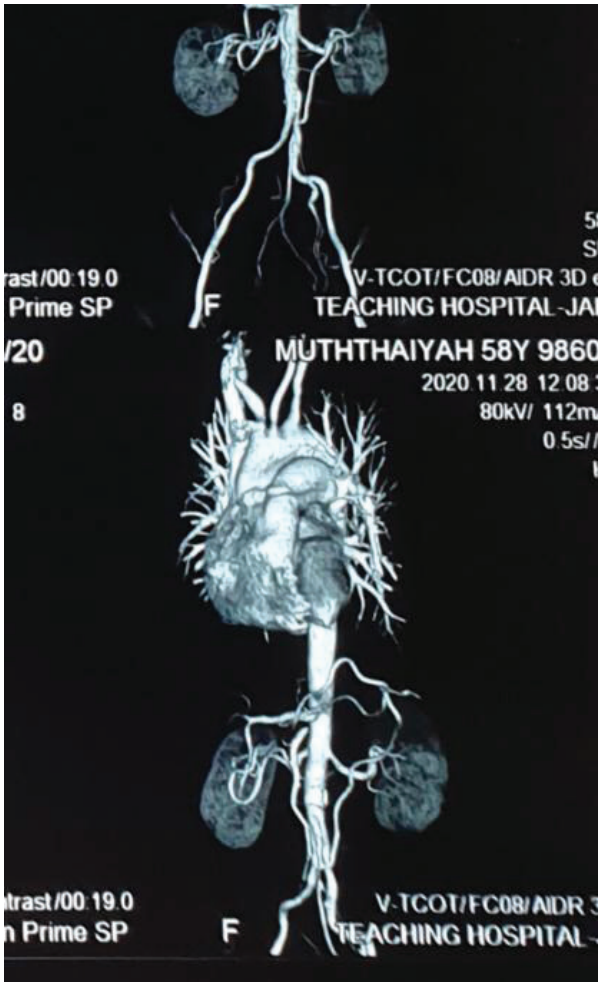

Figure 1: CT angiogram

Based on the clinical findings from the history, examination and investigation diagnosis of Takayasu arteritis type V is made. He was started on oral prednisone 1mg/Kg/day with bone mineral prophylaxis. He was also started on treatment for gastritis and duodenitis with proton pump inhibitors coupled with *Helicobacter pylori* eradication regime. A month later, on review he was asymptomatic.

Discussion

TAK is a rare condition often present with absence of peripheral pulses so called 'pulse less disease' (4). It is diagnosed by using American College of Rheumatology 1990 criteria (2). Following are the six components of the criteria 1) age of 40 years or younger at disease onset 2) claudication of the extremities 3) decreased pulsation of one or both brachial arteries 4) difference of at least 10 mmHg in systolic Blood pressure between arms 5) bruit over one or both subclavian arteries or the abdominal aorta 6) arteriographic narrowing or occlusion of the entire aorta, its primary branches, or large arteries in the upper or lower extremities that is not due to arteriosclerosis, fibromuscular dysplasia, or other courses. Presence of three or more of six criteria supports the diagnosis of TAK with the sensitivity of 90.5% and the specificity of 97.8% (5). According to Takayasu conference held in Tokyo in 1994 (5,6), TAK is sub-grouped into six types according to angiographic findings of arterial involvement as follows, type I- branches from the aortic arch; type IIa- ascending aorta, aortic arch and it's branches; type IIb- ascending aorta, aortic arch and its branches, thoracic descending aorta; type III-thoracic descending aorta, abdominal aorta, and/or renal arteries; type IV- abdominal aorta and/or renal arteries; type V- combined features of types IIb and IV. Our patient fulfilled four criteria to make the diagnosis of Takayasu arteritis, type V.

Presentation of TAK can vary from asymptomatic to non-specific symptoms such as fever, weight loss, malaise and myalgia with chronic cause to life threatening condition like CVA, myocardial infarction or mesenteric ischemia according to the area of vessel involvement. There are studies done on distribution of TAK related to sex and ethnicity respectively, which revealed females have higher tendency for the involvement of supra diaphragmatic portion of the aorta and its branches such as aortic arch and its branches whereas males have higher tendency for the involvement of infra diaphragmatic portion of the aorta and its branches such as abdominal aorta, renal arteries, mesenteric arteries and iliac arteries which is present in our patient. Patients with TAK from Japanese descent, among whom most are females (90%) have involvement of the aortic arch and its major branches but in patients from Indian descent where significant number of patients are males (37%) have involvement of the abdominal aorta and its branches (1).

Conclusion

Because of the rarity of the condition, TAK is often overlooked especially when patient present with nonspecific symptoms. Therefore, clinical suspicion of TAK should be entertained irrespective of the age, sex and ethnicity of the patient whenever clinical findings denote the possibility of Takayasu arteritis. It is pivotal for preventing major and life- threatening complications of TAK and for providing symptomatic improvement with the commencement of appropriate medical treatment.

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