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Case Report
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Late Diagnosis of Takayasu's Arteritis Despite Multiple Unheeded Clues: A Lesson to Learn

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Abstract

Takayasu's arteritis is a large vessel vasculitis which affects the aorta and its major branches. The mean age of onset is approximately 30 years, with fewer than 15% of cases in the age group above 40 years. The diagnosis of Takayasu's arteritis can be challenging without a proper clinical suspicion and acumen. We report a case of Takayasu's arteritis diagnosed in a 63-year-old lady who presented with heart failure despite an obvious blood pressure discrepancy in her previous seven pregnancies.

Keywords: Takayasu's arteritis; Blood pressure discrepancy; Computed tomography aortogram

Introduction

Takayasu's arteritis is a large vessel vasculitis, involving the aorta and its major branches. It affects mainly the young females, with higher prevalence among Asian populations. Non-specific presentations and lack of specific laboratory markers make the diagnosis of Takayasu's arteritis challenging. The presence of characteristic features such as absent or diminished pulse and blood pressure discrepancy in the absence of proper clinical suspicion might result in missed diagnosis of Takayasu's arteritis. Early diagnosis and treatment are important to prevent any serious complications of the disease.

Case Presentation

A 63-year-old lady presented with chest pain and heart failure symptoms of orthopnea and reduced effort tolerance. The chest pain is heavy in nature, central and non-radiating. She has a background history of bronchial asthma diagnosed 40 years ago, in good control with turbuhaler symbicort. She has a history of admission to a district hospital 6 months ago with similar chest pain, treated as unstable angina with 3 days of subcutaneous fondaparinux and discharged home with aspirin and clopidogrel. She was referred to a tertiary centre for further management of ischemic heart disease, however, still awaiting an appointment date.

Upon presentation, she was hypotensive with a blood pressure of 85/50 mmHg and a heart rate of 103 beats per min. Her Glasgow Coma Scale was full and orientated and does not appear to be in respiratory distress with an oxygen saturation of 97% under room air. Her physical examination revealed a displaced heart apex beat to 6th intercostal space at left anterior axillary line with the presence of pansystolic murmur over the apex and early diastolic murmur over the left sternal edge. There were fine crackles heard at the lower thoracic region bilaterally with no pedal edema. Otherwise, her jugular venous pressure was not raised with good peripheral perfusion and other systemic examinations were unremarkable.

Her chest radiograph demonstrated a congested lungs field with bilateral blunted costophrenic angle and cardiomegaly (Figure 1). Her electrocardiogram showed sinus rhythm with left bundle branch block and a bedside echocardiogram revealed a reduced left ventricular ejection fraction of 40% with severely dilated left atrium and ventricle and the presence of severe mitral and aortic regurgitation. Otherwise, her serum creatinine kinase and troponin I levels were not raised at 41 U/L and 16.8 pg/mL respectively. Other baseline blood investigations including full blood count, renal profile, liver function test, and coagulation profile were normal.

She was admitted for cardiogenic shock secondary to acute coronary syndrome. She was clinically well in ward with no chest pain or shortness of breath. However, she remained hypotensive

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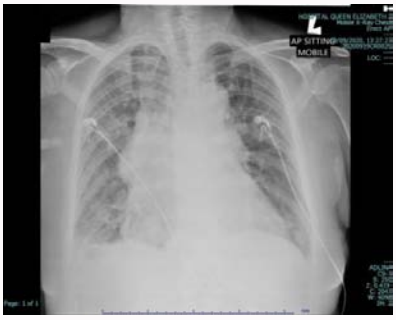

Figure 1: Chest radiograph: Congested lungs field with blunted bilateral costophrenic angle and cardiomegaly.

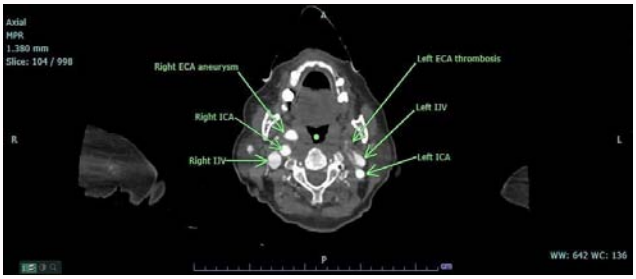

Figure 2: CTA: Right ECA aneurysm and left ECA thrombosis.
CTA: Computed Tomography Aortogram; ECA: External Carotid Artery; ICA: Internal Carotid Artery; IJC: Internal Jugular Vein

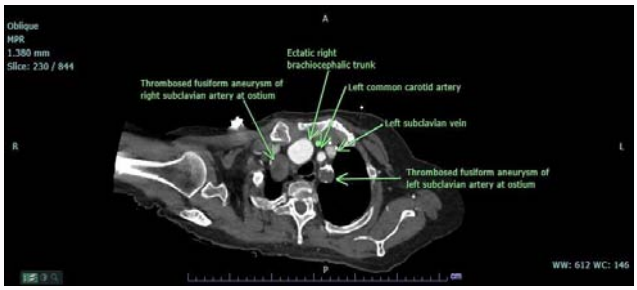

Figure 3: CTA: Thrombosed fusiform aneurysms of bilateral subclavian arteries at ostium and ectatic right brachiocephalic trunk.
CTA: Computed Tomography Aortogram

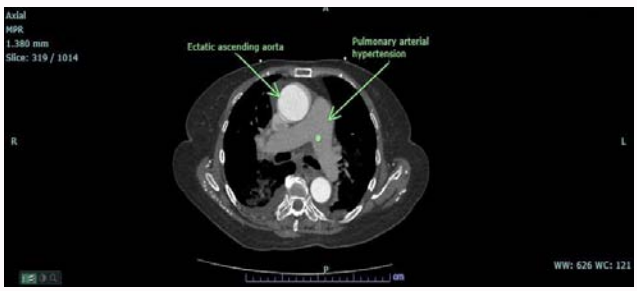

Figure 4: CTA: Ectatic ascending aorta and pulmonary arterial hypertension.
CTA: Computed Tomography Aortogram

with unexplainable increasing inotropic support. Further history taking revealed that she has the similar issue in her previous seven pregnancies in which she was noted to be hypotensive and only able to get a normotensive blood pressure by measurement on her lower limbs. Otherwise, her pregnancies were uncomplicated and uneventful. Detailed physical examination subsequently revealed weak bilateral brachial and radial pulses with good bilateral femoral,

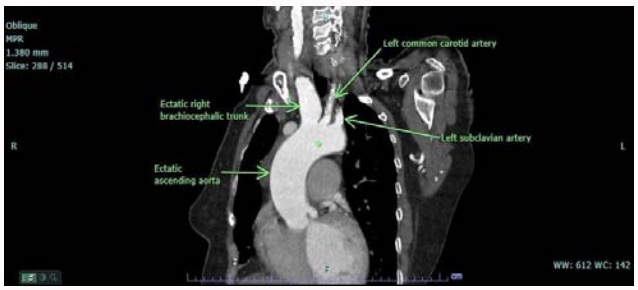

Figure 5: CTA: Ectatic right brachiocephalic trunk and ascending aorta.
CTA: Computed Tomography Aortogram

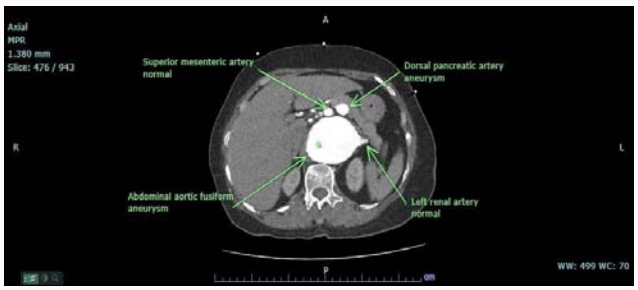

Figure 6: CTA: Abdominal aortic fusiform aneurysm and dorsal pancreatic artery aneurysm.
CTA: Computed Tomography Aortogram

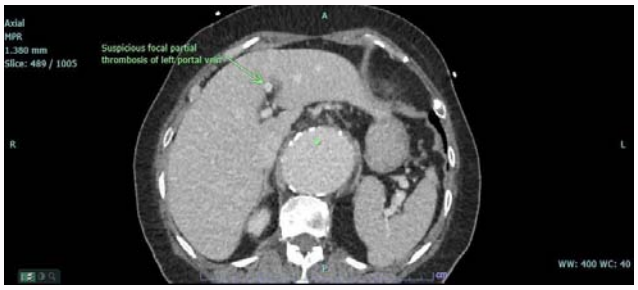

Figure 7: CTA: Suspicious focal partial thrombosis of left portal vein.
CTA: Computed Tomography Aortogram

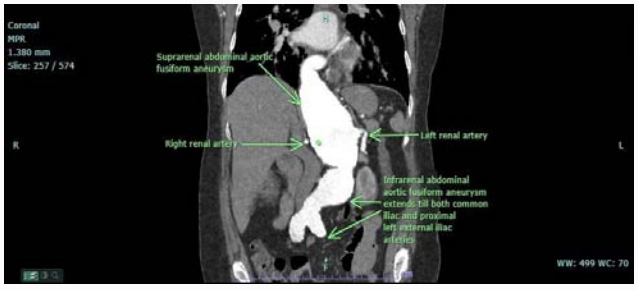

Figure 8: CTA: Suprarenal and infrarenal abdominal aortic fusiform aneurysms extending till both common iliac and proximal left external iliac arteries.
CTA: Computed Tomography Aortogram

popliteal, and dorsalis pedis pulses. Measurement of blood pressure on different limbs showed a significant discrepancy between upper and lower limbs with 91/57 mmHg over right arm, 90/50 mmHg over left arm, 179/69 mmHg over right leg, and 185/62 mmHg over left leg.

A computed tomography aortogram was done and revealed an ectatic ascending aorta and right brachiocephalic trunk with multiple

Figure 9: CTA: Infrarenal abdominal aortic fusiform aneurysm extends till bilateral common iliac and left proximal external iliac arteries, and bilateral internal iliac arteries thrombosis.
CTA: Computed Tomography Aortogram

aneurysms over proximal right external carotid artery, dorsal pancreatic artery, and suprarenal and infrarenal abdominal aorta, with thrombosed aneurysm of bilateral subclavian arteries, complete stenosis of left external carotid artery, and thrombosis of bilateral internal iliac arteries (Figures 2-9). A diagnosis of Takayasu's arteritis was made based on 1990 American College of Rheumatology criteria for Takayasu's arteritis. She was discharged with Cardiprin 1 tablet OD, Bisoprolol 5 mg OD, Perindopril 8 mg OD, and Spironolactone 6.25 mg OD, and given outpatient follow-up under the Cardiology, Rheumatology, and Vascular teams.

Discussion

Takayasu's arteritis has been reported in all parts of the world. However, data on epidemiology of Takayasu's arteritis is limited, probably due to the rarity of the disorder. It appears to be more prevalence among the Asian populations, affecting mostly young females in 80% to 90% of the cases, with an age of onset between 10 to 40 years old [1].

The onset of symptoms in Takayasu's arteritis tends to be sub acute with a wide range of presentations. In the early phase of Takayasu's arteritis, non-specific constitutional symptoms such as weight loss, low grade fever, night sweat, arthralgia, myalgia, malaise, and fatigue are common [1,2]. As the inflammation progresses, leading to permanent vascular injuries, characteristic features become more apparent. One of the common features is hypertension, which can be a result of renal artery stenosis or coarctation of aorta, with the former being the more common cause [1-3]. More than 80% of cases have diminished or absent pulses associated with limb claudication and blood pressure discrepancies. Vascular bruits present in up to 94% of cases, particularly affecting the carotid, subclavian, and abdominal vessels [2]. Chest pain may occur due to coronary artery ostial narrowing from aortitis or coronary arteritis. Mesenteric artery involvement will result in post-prandial abdominal pain, diarrhea, and gastrointestinal hemorrhage. Neurological symptoms such as light-headedness, vertigo, headache, convulsions, strokes, and visual impairment may result from carotid and vertebral arteries stenosis. About 10% to 30% of patients experienced carotidynia, which is tenderness of a carotid artery [1].

The diagnosis of Takayasu's arteritis is often delayed due to the non-specific presentations and lack of specific laboratory markers. Time from onset to diagnosis may take several months up to 11 years. Without a proper clinical suspicion, the diagnosis can be missed even with apparent signs such as loss of peripheral pulses. Significant delayed in diagnosis of >12 months may subject patients to angioplasty and surgery, which may be prevented by an increased clinical awareness of Takayasu's arteritis for earlier diagnosis [4]. In the present case, she was asymptomatic with only blood pressure discrepancies noted in multiple pregnancies, however, were unheeded leading to delay diagnosis with complication of heart failure.

There is no unified guideline for the management of Takayasu's arteritis due to the lack of complete placebo-controlled and randomized clinical trials. High dose prednisolone (1 mg/kg/day) is the standard initial treatment for Takayasu's arteritis, with conventional immunosuppressive agents such as methotrexate and azathioprine to be added as steroid sparing agent while tapering down steroids. Some physicians prefer an initial treatment with the combination of steroids and another immunosuppressive agent. To date, there is no evidence on clinical outcomes of different immunosuppressive agents. Biologic agents such as anti-TNF agents, rituximab and tocilizumab are choices of treatment for patients who are resistant or intolerant to initial therapy. Antiplatelet may help in lower down ischemic events in patient with Takayasu's arteritis. Endovascular interventions including balloon angioplasty and stent graft replacement play an important role in revascularization of the affected organs with arterial stenosis and occlusion, especially in late diagnosis of Takayasu's arteritis [5].

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