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"DELAYED DIAGNOSIS OF PEDIATRIC TAKAYASU ARTERITIS: A CASE OF MULTISYSTEM PRESENTATION OVER EIGHT YEARS"

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Abstract
Takayasu's arteritis (TA) is a rare type of vasculitis, a group of disorders that causes blood vessel inflammation. In Takayasu's arteritis, the inflammation damages the large artery that carries blood from your heart to the rest of your body (aorta) and its main branches. In this case, a 6-yearold girl who repetitively presented to OPD with complaints of abdominal pain and fever, each time was misdiagnosed for eight years. She underwent extensive investigations but all of them went uneventful. The unusualness here lies in repetitive presentation with abdominal symptoms (pain/distension that were not explained). Afterwards she underwent aortogram that revealed some significant findings including stenosis of both subclavian arteries, tapering of thoracic aorta and subtotal occlusion of abdominal aorta. She met EULAR/PRES criterion for pediatric Takayasu Arteritis and was diagnosed henceforth. This case highlights that Takayasu Arteritis can present with abdominal distension and generalized symptoms, particularly due to unusual findings like subtotal occlusion of the abdominal aorta. Given that Takayasu arteritis is rare in children, reporting such cases aids pediatricians and rheumatologists in recognizing this condition earlier and improving treatment outcome.

INTRODUCTION

The systemic inflammatory disease known as Takayasu arteritis(TA), often called pulseless disease, causes damage to the medium and large arteries as well as their branches[1].The condition may result in weaker arterial walls that could burst (aneurysm) or lead to the narrowing or clogging of arteries. High blood pressure, arm or chest pain, and ultimately heart failure or stroke can also result from it[2].Takayasu's arteritis commonly affects adults between the ages of 20 and 40. Patients are often diagnosed between the ages of 15 and 35, with approximately 80–90% of those affected being female.Takayasu's arteritis is more frequently diagnosed by medical professionals in Japan and in individuals of Mexican, Indian, and East Asian heritage[3].

European League Against Rheumatism/Paediatric Rheumatology International Trials Organisation/ Paediatrics Rheumatology European Society (EULAR/PRINTO/PRES) proposed validated classification criteria for the diagnosis of TA that includes angiographic abnormalities, alterations in peripheral arterial pulses, systolic blood pressure (BP) discrepancy in any limb, arterial hypertension, large artery bruits, and elevated acute phase reactants[4].While TA primarily affects the ascending aorta and its major branches, there are rare instances where a patient may exhibit coarctation or

hypoplasia of the mid-aorta, including the descending thoracic or abdominal aorta.[5]

Here we present a case of a young girl who remained undiagnosed for eight years. The peculiarity here lies in the diverse range of symptoms exhibited at such a young age and the atypical involvement of the vascular system. These unusual findings contributed to a significant diagnostic challenge.

CASE PRESESENTATION:

A 6-year-old female presented to the Outpatient Department with fever and abdominal distension. An abdominal computed tomography scan revealed lymphadenopathy and fluid-filled dilated bowel loops. Exploratory laparotomy for lymph node biopsy was advised, but the patient's guardians opted for leave against medical advice, receiving supportive antipyretic medications.

After approximately two and a half years, the patient returned with fever, cough, and dyspnea. Echocardiography demonstrated pancarditis with mild left ventricular systolic dysfunction. Laboratory tests showed elevated C-reactive protein and erythrocyte sedimentation rate, with negative anti-nuclear antibody titers. Physical examination revealed near-absent upper limb pulses, initially attributed to pancarditis. Anti-failure medications were initiated, and she was discharged after one month following clinical improvement.

Less than a year later, the patient presented with fever and tachycardia, diagnosed with complicated pneumonia. Hepatomegaly was noted but remained unexplained. She was discharged after one week of treatment.

After about four years, the patient returned with a three-month history of progressive shortness of breath, low-grade intermittent fever, and generalized abdominal pain associated with anorexia and vomiting but no diarrhea. Shortness of breath was accompanied by facial swelling and neck vein distension, without cough or wheezing. Physical examination revealed a temperature of 100°F, SpO2 of 97%, and heart rate of 130/min. Blood pressure measurements were 110/66 mmHg in the right upper limb, 107/68 mmHg in the left upper limb, 77/54 mmHg in the right lower limb, and 80/65 mmHg in the left lower limb. Bilateral upper limb pulses were weak, and bilateral lower limb pulses were absent. Cardiovascular examination showed a normal chest shape, no precordial bulge, and an S1 + S2 + S3 gallop rhythm. The patient had coarse facies with facial and periorbital edema. Abdominal examination revealed hepatomegaly 4 cm below the right costal margin, with no other abnormalities noted.

A computed tomography aortogram revealed narrowing of bilateral subclavian arteries (Figure 1), tapering of the thoracic aorta, and subtotal occlusion of the abdominal aorta (Figure 2). These findings were highly suggestive of systemic vasculitis, and a diagnosis of Takayasu arteritis was established.

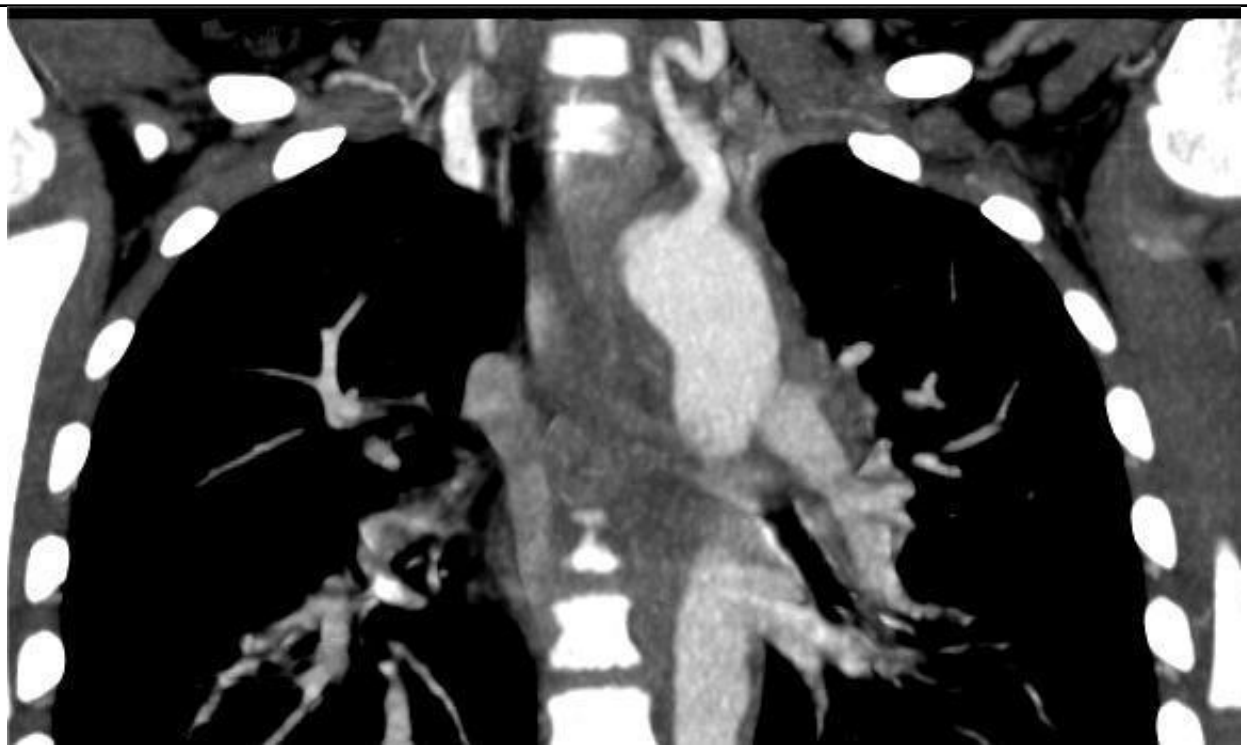

Figure 1: Computed tomography(CT) showing narrowing of Bilateral Subclavian arteries.

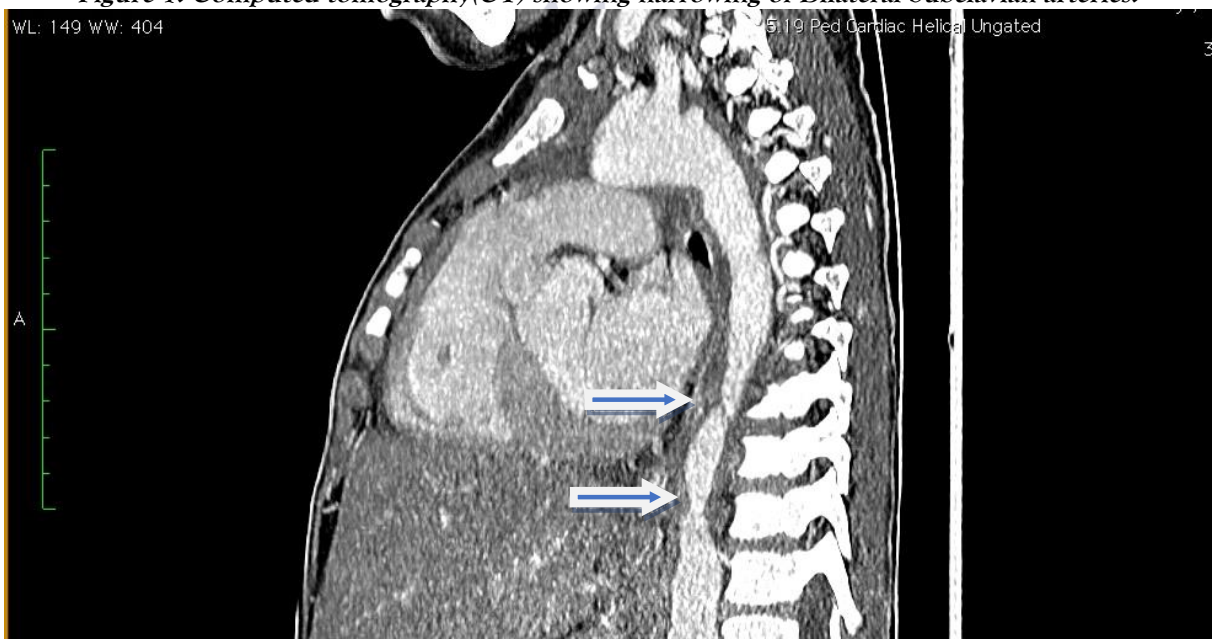

Figure 2: Computed tomography(CT) showing tapering of the Thoracic aorta, and subtotal occlusion of the Abdominal aorta.

DISCUSSION:

The aorta and its branches are involved in Takayasu's arteritis (TA), a chronic granulomatous vasculitis of unclear etiology that causes vascular sequelae with thrombus development and/or stenotic lesions. Known as Takayasu, Japanese eye specialist who initially explained the illness in a young female with retinal abnormalities in 1908. Afterwards Shimzu and Sano (1928) called it pulseless disease[6]. Its annual incidence is expected to be 2.6/1,000,000.

Although the illness is present all over the world, it is more typical in China, India, and Japan[7]. In Pakistan, Takayasu arteritis (TA) has been observed in both adults and children aged 12 to 19. While the illness has been documented in this age group, it is extremely rare in children under the age of ten[8]. This illustrates the uncommon nature of presenting Takayasu arteritis in very young childern and emphasizes the significance of evaluating a broad differential diagnosis in this age range to ensure quick and correct diagnosis.

In our case, a 6-year-old girl presented with generalized symptoms including abdominal pain and fever. She was continuously misdiagnosed due to overlapping nature of her symtoms. And after years of extensive workup during one of her visits CT aortography revealed stenosis of subclavian artery, tapering of thoracic aorta and subtotal occlusion of abdominal aorta. She met the EULAR/PRES criterion for paediatric Takayasu arteritis patients. This criterion requires angiographic anomalies (conventional, CT, or MR) in the aorta or its main branches with at least one of the following four characteristics: Reduced peripheral arterial pulse(s) and/or claudication of limbs, blood pressure difference > 10 mmHg, bruits over aorta and/or its major branches, and hypertension (linked to childhood normative data)[9]. Such findings of Takayasu Arteritis presenting in childhood with abdominal symptoms have also been reported in the past[10]. "To date, there have been no reported cases of Takayasu arteritis in children exhibiting subtotal occlusion of the abdominal aorta, despite this symptom being documented in some adult cases.

The primary takeaway lesson of this case report is that physicians should suspect rare diseases whenever facing diagnostic dilemma, which in our case was due to very overlapping nature of symptoms. The early presentation of Takayasu arteritis in children underscores the critical need for heightened awareness among healthcare providers. Recognizing the symptoms and understanding the potential impacts of these conditions can lead to timely medical intervention, which is essential for managing health effectively. Early diagnosis not only helps mitigate complications but also significantly improves the overall quality of life for affected children. Yet another fact that adds to the peculiarity of this case is subtotal occlusion of abdominal aorta due to Takayasu arteritis. Such rare findings of Takayasu arteritis have been reported in this region in the past.

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