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Beyond the Usual Suspects: Emerging Insights into Takayasu's Arteritis and Its Role in Secondary Hypertension

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Authors' contributions

This work was carried out in collaboration among all authors. Author JP gathered the case from the emergency ward. Authors HMK and LSSN arranged the theoretical and the presentation review of the case report. Author HY aided in reviewing the literature part. Authors MN and PK chipped away at the case presentation alongside the remaining authors. Authors HMK and LSSN aided in drafting the presentation, discussion, and summarized the conclusion part of the case report. Author PKY alongside author JP dealt with the literature searches and other authors in the arrangement of the manuscript. All authors read, supported and approved the final manuscript.

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Case Report

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ABSTRACT

Background: Pulselessness is the main sign of Takayasu's Arteritis (TAK), a granulomatous large artery vasculitis that mostly affects the aorta, major aortic branches, and pulmonary arteries. Renal artery stenosis is the source of arterial hypertension, which is common in TA. It is estimated that there are 2.6 incidences of Takayasu's arteritis per million people worldwide each year.
Case Details: We report on an 18-year-old girl whose significant arterial hypertension on her left arm, identified during a routine check-up, was the primary manifestation of Takayasu's arteritis (TA). The left arm's systolic blood pressure was noticeably greater than the right's. Additionally, there was a difference in blood pressure between the legs and left arm. Auscultation revealed the presence of vascular bruits over the left subclavian regions. A CT angiography was used to establish the diagnosis of TA. The results indicated a substantial constriction at the right renal artery's origin and characteristics compatible with aortic arteritis of the thoracic and abdominal aorta with active disease given wall enhancement. Due to the disorder's rarity and the varied ways in which it manifests clinically, a delayed diagnosis and treatment are likely.
Conclusion: Our study emphasizes that this illness can and does affect young people, and as such, patients presenting with suggestive symptoms and signs—especially young patients with unexplained hypertension—should be taken into consideration. Appropriate imaging and clinical suspicion are essential for the accurate diagnosis and treatment of TA patients. Several findings were found during the clinical examination, including signs of claudication, fever symptoms, involvement of major vessels including the aorta and its branches. This illustrates the condition of Takayasu's arteritis and what medical professionals should anticipate when treating it.

Keywords: Fibrous thickening of the arterial walls; inflammatory vasculitis; pulseless disease; secondary hypertension; Takayasu's arteritis; thromboaortopathy.

1. INTRODUCTION

A kind of vasculitis known as Takayasu's arteritis (TA) mostly affects the aorta and its major branches, though it can also affect the coronary and pulmonary arteries. Compared to men, women experience it more frequently (80–90%), especially those between the ages of 10 and 40 [1]. Even yet, there have been several cases documented globally, with an annual frequency of 2.6 cases per million. The signs and symptoms of systemic and vascular inflammation, such as angina, arthralgia, hypertension, constitutional symptoms, and differences in blood pressure between the upper limbs, are among the clinical manifestations of TA [2]. Only TA along with hypertension seen in about half of patients because of renal artery stenosis or decreased aortic elasticity.

Usually, the American College of Rheumatology or Ishikawa criteria are used to diagnose TA. Giant cell arteritis (GCA) and TA are comparable disorders that might be confused with one another. The histological investigation is unable to distinguish between the two diseases [3]. Corticosteroids, either with or without a corticosteroid-sparing medication, are the mainstay of treatment for TA. In cases of irreversible artery stenosis and massive aneurysms, surgery may also be required. Usually, anemia and sedimentation rates improve. Rarely, immunosuppression might reverse vascular stenosis. However, the lesions may progressively worsen [4,5].

Although TA primarily affects young individuals in their second and third decades of life, cases have also been documented in children and those over 40. The oldest patient reported in a study was 75 years old, and the youngest was 6 months old [6,7]. Compared to men, women are more likely to be impacted. Approximately 80% of TA patients in adulthood are female, while reports from Japan, Mexico, and Israel have shown varying female-to-male ratios (of 9:1 to 6.9:1 to 1.2:1). The gender gap is less pronounced in the Pediatric population. The ratio of female to male participants in childhood TA studies from South Africa and India is 2:1. In Japan, there are about 200 new instances of TA reported each year [8-11]. In Minnesota Olmsted County, the incidence of TA is predicted to be 2.6 individuals per million population annually. It is unclear, therefore, how this figure would apply to the diverse population of the United States as a whole. One instance per million people is believed to occur annually throughout all of Europe. The yearly incidence is 0.15 cases per million in the United Kingdom and 1.2 cases per

million in Sweden [11-14]. It's unclear how often TA is in kids. The most prevalent clinical symptom of Takayasu's arteritis in both adults and children is arterial hypertension, even though patients may exhibit a wide range of other clinical symptoms [15].

2. CASE PRESENTATION

An 18-year-old adult female patient was admitted to the Emergency Ward with chief complaints of fever associated with chills, loss of appetite, and headache on and off (episodic) for 15 days. Her headache is usually aggravated by stress and while watching television but is relieved with rest. During the general examination, the pulse rate is 84 beats per minute, the respiratory rate is 18 cycles per minute, and the SPO_2 is 100% on Right Atrium. Vascular bruits were heard over the abdominal aorta. On obtaining the background information, she had a family history of consanguineous marriage (1st-degree consanguinity), and a history of irregular Menstruation (from the past four months). From the above complaints, Aorta Arteritis was suspected in the patient.

A physical examination revealed no signs of cardiac dysfunction. Heart sounds were typical. Over the heart apex, there was an audible faint systolic murmur. Auscultation revealed vascular bruits over the left supra- and subclavian areas, as well as the interscapular region. There was no enlargement of the spleen or liver. There was a mild atrophy and reduced tone in the muscles of right upper extremity. There was no pulse from the right radial artery or the right external carotid artery. There were equal and detectable femoral pulses. The left arm's systolic blood pressure was found to be significantly raised upon admission, measuring 187/85 mmHg, whereas the right arm's blood pressure was difficult to measure but was around 85/60 mmHg. Ultrasonography revealed no anomalies related to intracardiac architecture, except for a minor regurgitation of the mitral valve. The internal diastolic size of the left ventricle, which measured 54.7 mm, was within the normal range for the patient's age and weight and did not indicate hypertrophied walls.

The results of the laboratory indicated that the serum C-reactive protein level was 1.19 mg/dl (normal value <0.5 mg/dL) and the erythrocyte sedimentation rate was increased at 54 mm/h (normal value <20 mm/h). Moreover, elevated serum immunoglobulin G levels and anemia (Hemoglobin level of 7.4 mg/dL) were noted. The renal Doppler ultrasonography evaluation revealed no abnormalities.

CT Angiogram Impression shows features that are consistent with Aorta Arteritis of the thoracic and abdominal aorta with active disease given wall enhancement, diffuse smooth narrowing of thoracic aorta from D4-D5 level to the renal segment with service narrowing of 70% at the level of the vertebral body, and Mild Osteo-proximal narrowing was also noted at the origin of the celiac artery and moderate (<50%) narrowing of the origin of the Superior Mesenteric Artery. Severe narrowing at the origin of the right renal artery with post-strategic dilation. The above features are in favor of probable aortic arteritis involving the thoracic aorta, ductal to the origin of the left subclavian artery. It has also shown that dampened flow in renal arteries may be secondary to proximal aortic pathology.

The patient was started on Tab. Amlodipine 5mg OD, Tab Metoprolol 25mg Once a day, and Tab. Prednisolone 30mg/day. The patient was asked to continue taking the Tab. Prednisolone for 2 weeks and later tapered the dose to 25mg/day. Since the descending thoracic aorta's hypertension was not adequately controlled at first, stenting had also been explored. However, since the girl had never received treatment before and her nonspecific markers of inflammation were elevated, she was eligible for ongoing medical care. She is currently receiving treatment, and her left arm's blood pressure ranges from 134/84 to 154/92 mmHg. Her left arm and legs have the same reading. ESR is 23 mm/h in lab testing, while C-reactive protein is just slightly raised at 1.12 mg/dlThe patient is presently being monitored clinically for an extended period by a nephrologist and cardiologist. After the first follow-up, the patient was advised to undergo a Stent graft for the thoracic aorta and a POBA+/- Stent to the right renal artery.

3. DISCUSSION

The Etiology and pathogenesis of TA remain contentious despite a decade of significant advancements in our understanding of the disease. The underlying pathophysiology is thought to be inflammatory and has an unclear Etiology at this point. Numerous etiologic causes, such as circulating antibodies resulting from an autoimmune process, streptococci, spirochetes,

Mycobacterium tuberculosis, and hereditary variables, have been hypothesized [15,16]. According to one theory, when an antigen is deposited in vascular walls, CD4+ T cells become activated and release cytokines that attract monocytes. After transforming, these monocytes become macrophages, which cause endothelial damage and the creation of granulomas in the vessel wall. Increased expression of vascular cell adhesion molecule-1 and intercellular adhesion molecule-1 has been shown in human investigations, indicating endothelial cell activation in individuals with TA [17]. The pathogenesis may also include humoral immunity. Patients with TA have anti-monocyte and anti-endothelial cell antibodies, which are correlated with disease activity. The genetic predisposition to TA has been thoroughly investigated. Japanese patients showed a substantial correlation between HLA B-52 and DR-2; however, this finding was not replicated in Western nations [18].

Rarely, TA has also been linked to other autoimmune conditions that may point to the involvement of immune mechanisms in the pathophysiology, including Crohn's disease, Wegener's granulomatosis, anterior uveitis, sarcoidosis, juvenile idiopathic arthritis, TA, and systemic lupus erythematosus. It will need further research to fully understand the pathophysiology of Takayasu’s arteritis [19].

The abdominal aorta and renal arteries are more commonly impacted in patients from Korea, India, and Western countries with TA, whereas aortic arch involvement is more common in Japanese patients. All countries have observed all patterns of vascular alterations, nevertheless. The signs and symptoms of TA are not particular. There is an early, active, inflammatory phase to the disease's clinical course, followed by a late, chronic phase. There may be a remitting and relapsing course over the weeks to months-long active period. Systemic illness, including fever, general malaise, sweats at night, hunger loss, weight loss, headaches, dizziness, arthralgia, and skin rashes, are its defining characteristics.

It is important to emphasize that early diagnosis of TA is rarely made correctly. Aneurysms bruits, and other signs of vascular abnormalities that suggest inflammation may indicate TA. Organ ischemia, occlusion, and/or vascular stenosis cause the late chronic phase. The location of arterial lesions affects the variety of their clinical presentations [20]. The Etiology of TA-induced arterial hypertension is intricate, multifaceted, and poorly understood. Three mechanisms are currently thought to be responsible for it: (a) mechanical, where high resistance to cardiac output imposed by narrowing causes hypertension proximal to the narrowed aorta (atypical coarctation); (b) neural, where hypertension proximal to the narrowed aorta results from aortic arch baroreceptors readjustment, ensuring adequate blood supply to organs distal to the narrowed aorta; and (c) hormonal, where hypertension is brought on by renal hypoperfusion as a result of stenotic lesions of one or both renal arteries or the aorta alone. The TA-observed decrease in artery wall elasticity could potentially be a factor in the blood pressure rise [21].

Systolic arterial hypertension in our patient appeared to be caused by thoracic aortic constriction, renal ischemia, and most likely hyposensitivity of aortic arch baroreceptors. It should be noted that with TA, both the cephalic arteries of the aortic arch and both subclavian arteries may be damaged, causing an alternation in wave pulse propagation that could lead to an over- or under-estimation of blood pressure. In both groups, however, hypertension is the most prevalent symptom. Typically, hypertension, heart failure, or a neurological incident cause the symptoms to present. Although claudication, bruit, and a missing pulse in youngsters without symptoms are unusual presentations, our patient had these, and they were essential to making the diagnosis [22].

Immunosuppressive medications, such as Methotrexate and/or Prednisone, are used to reduce or eradicate inflammatory activity in the treatment of TA. A little over 60% of young TA patients react to glucocorticoids. But up to 40% of people who taper off steroids relapse. In TA, other treatments such as tacrolimus hydrate, cyclophosphamide, mycophenolate mofetil, and azathioprine are also utilized, particularly for diseases that are resistant to corticosteroids [23]. Pediatricians should be advised against using ACE inhibitors until renal artery stenosis has been ruled out while treating hypertension. Hypertension should be treated aggressively and often using a multimodal regimen. TA can significantly improve with anti-inflammatory medication. In adults, the 6-year survival rate can reach 89%. However, the death rate among youngsters might reach up to 28%. Novel targeted therapy, including antitumor necrosis factor agents, may eventually result from a

deeper understanding of the pathophysiology of Takayasu's arteritis [24]. While early diagnosis is crucial for better results, a novel biological approach to treatment may be less harmful and more successful than the current immunosuppressive regimens [25-27].

4. CONCLUSION

This study investigates Takayasu's arteritis, a common condition primarily seen in females aged 13-38. Symptoms include fever, claudication, and blood pressure changes. Imaging is the main diagnostic tool, revealing stenosis of major vessels and other conditions. The clot formation may cause heart function changes and related consequences. This provides a comprehensive understanding of Takayasu's arteritis and its potential treatment outcomes.

The case study presented here shows how a proper diagnosis can be reached with the help of a thorough physical examination, appropriate clinical history-taking, and well-chosen supplemental diagnostic testing. Indeed, the suspicion of TA was aroused by the co-existence of hard-to-control hypertension, anemia, stroke, pulsatile neck mass, renal artery stenosis, and carotid Doppler ultrasonography abnormalities. The diagnosis of TA, a rare and potentially dangerous illness, depends heavily on a doctor's suspicion and vigilance.

CONSENT

Every author proclaims that informed consent was acquired from the patient, and the signed consent form is submitted alongside the manuscript and submission form.

ETHICAL APPROVAL

As per international standards or university standards written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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