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Takayasu arteritis masquerading as recurrent abdominal pain

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Abstract

Takayasu arteritis (TA) is a rare granulomatous inflammation of large arteries, specifically the aorta and its branches. It is more prevalent in young Asian women with the highest incidence in Japan. There is a delay in diagnosis due to nonspecific early symptoms and unfamiliarity with this disease. We present a case of a young female who presented with recurrent abdominal pain suggestive of mesenteric angina and constitutional symptoms over one-year. Her radial pulses were absent, and she had multiple bruits on examination over the right subclavian and carotid arteries and the epigastrium, along with high inflammatory markers. Her CT angiography of large vessels confirmed the diagnosis of TA and she was treated with high-dose steroids with good clinical remission confirmed by a reduction in inflammatory markers and improvement in symptoms.

Key words: Takayasu arteritis, large and medium vessel vasculitis, angiography, recurrent abdominal pain

Introduction

Takayasu arteritis (TA) is a large and medium vessel vasculitis, mainly involving the aorta and its major branches due to granulomatous inflammation. The inflammation results in the infiltration of lymphocytes, plasma cells, and giant cells leading to stenosis, occlusion, and aneurysms of the vessels. (1,3) The estimated worldwide incidence is 2.6 cases per million. Although TA has a worldwide distribution, it is most frequently seen in Asian countries.(2) Females are more commonly affected than males with the mean age of onset being 30. Clinical presentation of TA is nonspecific during the early phase of the disease and clinical symptoms depend on the site of involvement. Due to this early nonspecific presentation diagnosis is delayed. The aetiology of TA is unknown. TA should be one of the differential diagnoses in a patient who presents with constitutional symptoms specifically in our region. Patients with suspected TA should undergo CT/MRI angiography. The mainstay of treatment is systemic glucocorticoids. TA patients respond to steroids well. However, the majority of cases will have a relapsing and remitting course of illness. This is a case report of a 20-year-old woman who presented with constitutional symptoms, ischaemic abdominal pain, multiple bruits with absent radial pulse and high inflammatory markers. We proceeded with CT angiography on suspicion of TA. Arteriography revealed thoracic and abdominal aorta narrowing and involvement of the superior mesenteric artery and bilateral renal arteries. The patient was started on high-dose prednisolone and she showed marked improvement in her symptoms as well as a reduction in inflammatory markers.

Case presentation

A 20-year-old previously healthy woman presented with recurrent abdominal pain. She described it as a burning type of severe pain associated with meals for one-year. There was no history of radiation of pain, associated regurgitation, or altered bowel habits. She tried antacids and proton pump inhibitors

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with no response. She consulted a gastroenterologist and underwent upper gastrointestinal endoscopy which was reported to be normal. Along with abdominal pain, she developed a progressive weight loss of 10 kg with poor appetite and low-grade fever. She was also complaining of on and off non-productive cough without hemoptysis or contact history of Tuberculosis. Hence, she was evaluated and found to have a high ESR of 110 mm/1st hour. Her full blood count revealed mild normochromic normocytic anaemia. Other cell lines were normal. Her liver profile, renal functions and chest x-ray were normal, and her Mantoux test had an induration of 7mm. She was started on anti-tuberculosis (anti-TB) treatment as a trial by a chest physician. However, she did not respond to anti-TB drugs even after one month. She continued to have constitutional symptoms with high ESR and was referred to the internal medicine team. A detailed history revealed right arm pain following exertion and when exposed to cold suggesting an ischaemic aetiology . On examination, her BMI was 18.5 kg/m^{-2}. On physical examination, blood pressure readings in both arms were normal. However a systolic blood pressure difference of 15 mmHg was noted between the right and left arms. Both the right radial and right brachial pulses were impalpable. Bruits were heard over the right subclavian, right carotid, and epigastrium. The cardiac apex was heaving in nature at the left 5th intercostal region and there was no cardiac murmur. She had a heart rate of 100 beats per minute and a body temperature of 37.9°C. Abdomen, respiratory, and neurological examinations were normal.

Clinical features suggestive of mesenteric ischemia, presence of arm claudication with weight loss prompted the working diagnosis of Takayasu Arteritis. Hence, a CT angiogram of the thoracic and abdominal aorta was performed and it showed diffuse thickening of ascending and descending aorta, arch of aorta, carotid arteries, coeliac axis, superior mesenteric artery, and renal arteries, suggestive of acute active phase of Type iii TA (figure 1). Echocardiography revealed left ventricular hypertrophy and good ventricular function.

The diagnosis was confirmed and she was started on high-dose prednisolone. She showed a dramatic response and constitutional symptoms subsided. Her inflammatory markers came down. Currently, her prednisolone is being tailed off and she is clinically improved.

Discussion

The exact pathophysiology of TA is not known(4), however, it is thought to be due to pan arteritis resulting in intimal hyperplasia, infiltration of mononuclear cells, and occasionally giant cells, medial and adventitial thickening is thought to be the cause.(5)

Clinical presentation of TA is nonspecific and patients present with a wide variety of symptoms during the early phase of the disease. Due to this nonspecific presentation diagnosis is delayed. Our patient

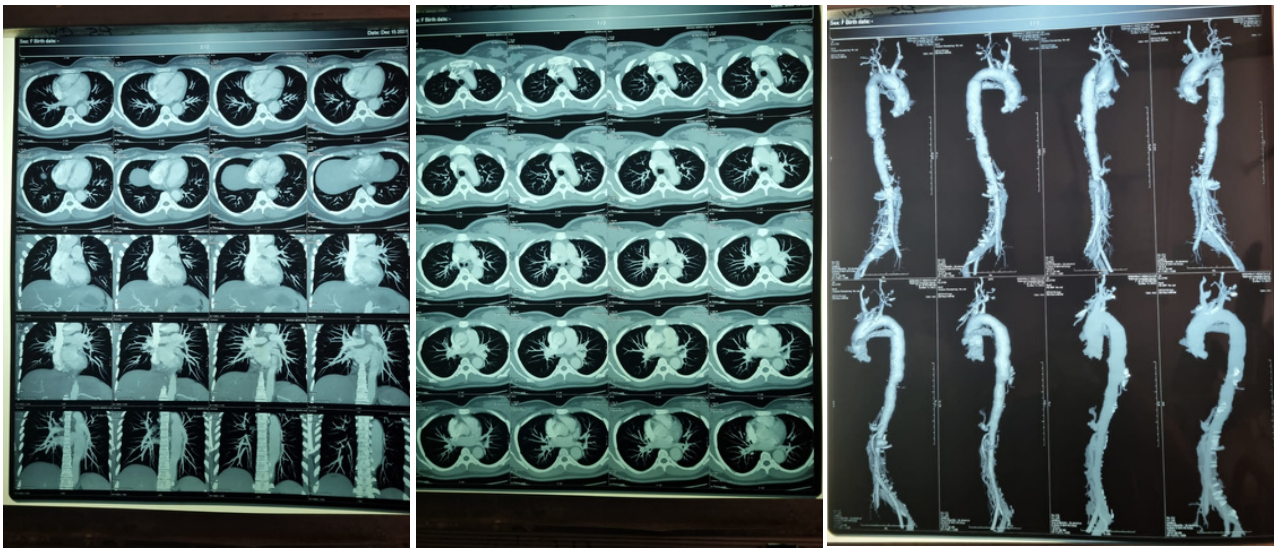

Figure 1 - CT angiogram of the thoracic and abdominal aorta showing diffuse thickening of ascending and descending aorta, arch of aorta, carotid arteries, coeliac axis, superior mesenteric artery, and renal arteries, suggestive of acute active phase of Type iii TA.

presented with nonspecific symptoms which delayed the diagnosis for about one year.

The diagnosis of TA should be suspected in a patient with constitutional symptoms, absent or diminished pulse, and /or arterial bruit, claudication and symptoms suggestive of organ ischaemia. In our case, the patient had long-standing abdominal pain related to meals due to the narrowing of the mesenteric artery reflecting organ ischemia. Narrowing of subclavian and carotid arteries were reflected as a carotid bruit and absent pulse.

The main differential diagnoses in our patients were chronic infection, especially Tuberculosis and autoimmune disorders.

The diagnosis of TA is made by clinical presentation along with CT/MRI angiography. The 1990 American College of Rheumatology (ACR) classification criteria aid the diagnosis of TA. According to ACR classification, 3 or more criteria have a sensitivity of 90.5% and a specificity of 97.8% in diagnosing TA.(6)

The 2022 ACR/European Alliance of Associations for Rheumatology (EULAR) classification for TA uses a weighted algorithm that includes clinical and imaging criteria. These criteria were constructed in part to reflect the growing role of non-invasive imaging in the evaluation of patients with TA.(9)

High levels of Acute phase reactants favour the diagnosis. The autoimmune profile will help to rule - out the differential diagnoses.

Patients with suspected TA should undergo CT/MRI angiography to demonstrate arterial luminal narrowing and occlusion of large and medium-sized vessels.

In our case, the patient was under 40 years old, and had a systolic blood pressure discrepancy of 15 mmHg between the right and left arms with impalpable right radial and right brachial pulses. Bruits over the right subclavian and carotid arteries, epigastrium and abnormal arteriography confirmed the diagnosis of TA.

The mainstay of treatment is high-dose systemic glucocorticoids and glucocorticoid-sparing agents, such as Methotrexate, Azathioprine, leflunomide, and Mycophenolate mofetil, especially in patients whose disease relapses after treatment with an extended course of glucocorticoids alone.(7)

High-dose steroids should be continued for 2-4 weeks and then tapered gradually when patients demonstrate clinical improvement.

In cases of critical organ ischemia due to irreversible arterial narrowing, there is a place for surgical intervention for endovascular procedures and stenting.(8)

TA patients respond to steroids well as in our case. However, the majority of cases will have a relapsing and remitting course of illness. Hence, TA patients need a long-term follow-up.

Conclusion

In young patients, especially females with constitutional symptoms and systemic manifestations, TA should be considered as one of the differential diagnoses. Varied clinical presentations lead to delayed diagnosis associated with increased morbidity due to irreversible stenosis of arteries and organ dysfunction. This case emphasises the importance of symptom analysis, thorough examination, and medical knowledge on differential diagnosis which lead to better clinical outcomes.

Declarations

Conflicts of interest
No conflicts of interest to be addressed regarding this case report.

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