Unusual presentation of more common disease/injury

Abdominal angina: an unusual presentation of Takayasu's arteritis

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Takayasu's arteritis (TA) is an idiopathic large vessel vasculitis of young adults that affects the aorta and its major branches. The authors hereby present a case of TA that presented with abdominal aorta thrombosis. She was put on low-molecular weight heparin, antiplatelets, corticosteroids, cyclophosphamide along with haematinics and was referred to Department of Cardiothoracic and Vascular Surgery for further management.

BACKGROUND

Takayasu's arteritis (TA) is a large vessel vasculitis in which arterial stenosis is three to four times more common than aneurysm. The most common site of stenosis is subclavian and innominate arteries. Renal artery involvement is especially common in Indian patients, explaining the elevated incidence of hypertension. Cardiac, renal and central nervous system involvement are the principal causes of severe morbidity and mortality. In this report, we describe a patient who presented with rare manifestation of abdominal aorta thrombosis.

CASE PRESENTATION

A 35-year-old woman presented with pain in both lower limbs on walking for past 3 years and pain in abdomen after meals for past 2 years. Initially pain developed on walking for around 500 m but gradually the distance reduced to 200 m on level ground. The pain used to get relieved by rest and was associated with numbness in both lower limbs and for past 1 year she felt that her lower limbs were cooler than rest of the body. She also had abdominal pain of moderate to severe intensity for past 2 years which started after taking meals and usually persisted for 1–2 h. Patient gave a history of significant weight loss over past 3 years. No history suggestive of claudication of upper limbs was present. On examination, her pulse rate was 84/min, radial pulses were bilaterally palpable; however, femoral, popliteal, posterior tibial and dorsalis pedis pulses were absent bilaterally. Blood pressure in upper limbs was 140/40 mm of Hg bilaterally and was not recordable in lower limbs. Respiratory rate was 16/min and pallor was present. There was no fever, clubbing or lymphadenopathy. On examination, lower extremities were cold, skin was smooth and shiny with sparsely distributed hair. Rest of the systemic examination, including abdomen, were normal.

INVESTIGATIONS

Investigation revealed haemoglobin of 5.5 gm/dl, total leucocyte count 12050 cells/mm³ (P 70, L 25, E 5, B 0) and

platelet count 135 000/mm³. Liver function tests including prothrombin time and kidney function tests including serum electrolytes were normal. Urine examination revealed 10–15 pus cells/HPF, fair number of red blood cells and growth of *Escherichia coli* (2000 colonies/mm³) on culture/ sensitivity. Rheumatoid factor and C reactive protein (CRP) were negative. Total serum cholesterol was 91.8 mg/dl, high-density lipoprotein 29.5 mg/dl, triglyceride 83.4 mg/dl, Very-low-density lipoprotein 16.6 mg/dl and Low-density lipoprotein cholesterol 45.6 mg/dl. CT aortography of abdomen showed abdominal aortic thrombosis extending from D 11 and L 2 vertebrae, just above the bifurcation of renal artery however the thrombus was not extending into renal arteries. Origin of celiac axis was not well visualised suggesting probable involvement. Multiple collateral channels were seen in intraperitoneal and retroperitoneal region with enlargement of bilateral superior and inferior epigastric arteries suggesting probable feeding arteries of collaterals (figure 1). The screening of thoracic aorta was normal. The echocardiography and ultrasound abdomen of the patient were within normal limits. Possibility of TA as a cause of abdominal aortic thrombosis was kept.

TREATMENT

She was treated with low-molecular weight heparin followed by acitrom, antiplatelets, pentoxifylline, prednisolone and cyclophosphamide along with hematinics.

OUTCOME AND FOLLOW-UP

Finally, she was referred to Cardiothoracic and Vascular Surgery Department for further management where she expired.

DISCUSSION

TA is an idiopathic large vessel vasculitis of young adults that affects the aorta and its major branches. Women are affected 10 times more commonly than men and median age at onset is 25 years. TA is best known to occur in Asia

Figure 1 CT aortography of abdomen showing abdominal aortic thrombosis extending from D 11 and L 2 vertebrae, just above the bifurcation of renal artery without renal arteries involvement along with multiple collateral channels.

although the distribution of the disease is worldwide and has been reported in all races and ethnicities.

In TA, arterial stenosis is three to four times more common than aneurysm. Claudication (upper limb 60% vs lower limb 30% approximately) is the most common complaint while the presence of bruits, increased blood pressure and pulse asymmetries are the most common physical findings. Cardiac, renal and central nervous system vascular disease are the principal causes of severe morbidity and mortality.

In 1990, the American College of Rheumatology proposed a simple classification with six inclusion criteria: age of disease onset < 40 years, claudication of extremities, decreased brachial artery pulse, blood pressure difference

> 10 mm Hg, bruit over subclavian arteries or aorta and arteriogram abnormality. Three criteria are required to diagnose TA, differentiating it from other vasculitis.¹ Our patient had three criteria of TA (age < 40, claudication of lower extremities and aortographic abnormality – abdominal aorta thrombosis) and fulfilled the definition of it.

Systemic symptoms including fatigue, weight loss and low grade fever are common in an early phase of disease. Vascular symptoms are rare at presentation, but with progression of disease evidence of vascular involvement and insufficiency become clinically apparent due to dilation, narrowing or occlusion of the proximal or distal branches of the aorta.² Increasing extremity or visceral ischaemia, malaise, myalgias, arthralgias, night sweats and fever may

indicate active disease. In the absence of constitutional or new vascular symptoms, active TA can be diagnosed by presence of new vascular abnormalities on sequential angiographic studies or by the presence of inflammatory changes in bypass biopsy specimens from patients in whom surgery was performed because of critical flow abnormalities in the setting of clinically quiescent disease.³

⁴ In active phase of the disease there is mild leukocytosis along with markedly elevated erythrocyte sedimentation rate (ESR). ESR > 40 is considered by many as an indicator of active disease with or without constitutional symptoms. Antistreptolysin O titre and CRP demonstrated in more than 50% cases but does not correlate with the disease. Studies using refinements in MRI⁵ and positron emission tomography technique may enable the clinician to detect qualitative abnormalities in vessel wall that imply inflammatory changes.

The most common site of stenosis in patients of TA is the subclavian and innominate arteries. Renal artery involvement is especially common in Indian patients, explaining the elevated incidence of hypertension.¹ Angiography is the gold standard for evaluation of vascular lesions. Angiography allows a topographic classification which correlates anatomic involvement, clinical manifestations and prognosis.⁶ TA is associated with substantial morbidity and its course usually extends for many years, with varying degree of activity. Approximately, 20% of patients have a monophasic and self limited disease. In others, TA is progressive or relapsing/remitting and requires immunosuppressive treatment. The 15 years survival rate has been reported to be 90–95%. Successful result of percutaneous angiography of aorta, renal and arch vessels approaches to 95–100% with good chronic outcome.

Early detection of the disease and appropriate management with proper follow-up is the key management of the disease as it requires multiple intervention. Almost all patients of TA improve when treated with high doses of corticosteroids; relapses are common with tapering

of corticosteroid therapy. Corticosteroid-resistant or relapsing patients may respond to the addition of daily cyclophosphamide (2 mg/kg) or weekly therapy with methotrexate (approximately 20 mg). Studies have demonstrated that treatment designed to block tumour necrosis factor γ dramatically improves most patients with TA who have relapsed or are refractory to conventional treatment.⁷

Learning points

- This report emphasises the importance of ensuring a thorough systemic evaluation in individuals with abdominal pain.
- In contrast to usual presentation, our patient presented with claudication of lower limbs with absent pulses.
- CT angiography showed abdominal aortic thrombosis without renal artery involvement.

Competing interests None.

Patient consent Obtained.

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