Manipal Alumni Science and Health Journal 2021; 6(1): 10

CASE REPORT

Necrotic Bowel In A Male With Takayasu Arteritis: A Case Report

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Keywords: Disseminated Takayasu Arteritis, case report, intestinal gangrene, hypertension, Asian

ABSTRACT

Takayasu Arteritis is one of the two large vessel vasculitis according to 2012 revised International Chapel Hill Consensus Conference definition of vasculitis. Disseminated Takayasu Arteritis in a male patient presented as intestinal gangrene is very rare and potentially fatal. Here, we have a case of a 40-year-old male of Asian ancestry with disseminated Takayasu Arteritis, came with acute abdomen and acute renal failure. No associated hemoptysis and no prior history of limb claudication. He was hypertensive with absent bowel sounds. Clinically he was hypertensive with raised acute phase reactants. No bruits were present. CT angiography showed total occlusion of abdominal aorta. Histopathogical analysis of resected bowel showed involvement of small vessels as well. Absence of history of ischemic symptoms of upper and lower limb ischemia is unique in this case. Early diagnosis is important for early targeted therapy to avoid further ischemic events and even mortality.

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INTRODUCTION

Takayasu Arteritis is one of the two large vessel vasculitis according to 2012 revised International Chapel Hill Consensus Conference definition of vasculitis [1]. Takayasu Arteritis presentation as intestinal gangrene is very rare. In fact, the recent 52 Canadian Takayasu Arteritis cohorts (a study over 27 years) [2] showed none of the ischemic bowel cases. Here, we reported a case of a male Takayasu Arteritis, which according to our local cohort study in 2016[3], rarely occurred among males because of 12:1 female preponderance.

CASE REPORT

RESULTS

A 40-year-old gentleman, active smoker (20 pack-years), admitted with one-day history of diffuse abdominal pain and vomiting. He had hypertension four weeks prior, requiring three antihypertensive agents. Careful history revealed absence of oro-genital ulcers or limbs claudication. No preceding noxious stimuli including illicit drug use or trauma. No history of prior altered bowel habit. No history of prior malaise or constitutional symptoms. Systemic review reported absence of headache, red eyes, skin rashes, skin nodules, chest pain, breathlessness, nasal discharge or crusting, epistaxis, or limb claudication. There was no history of diabetes or significant family history of premature cardiovascular disease.

On physical examination, patient was afebrile. Tenderness was felt over the entire abdomen with absent bowel sounds. No bruits were noted while peripheral limbs were not cyanosed. Laboratory data revealed white blood cell count between 20,000/mm3 to 30,000/mm3 and platelet count between 500,000/mm3 to 600,000/mm3. Erythrocyte sedimentation rate was 92 mm/hr and C-reactive protein was 77 mg/L. Cultures from blood and urine did not show pathogenic organism. Liver function was normal while renal function was impaired with creatinine of 275 umol/L (normal range = 62-106 umol/L).

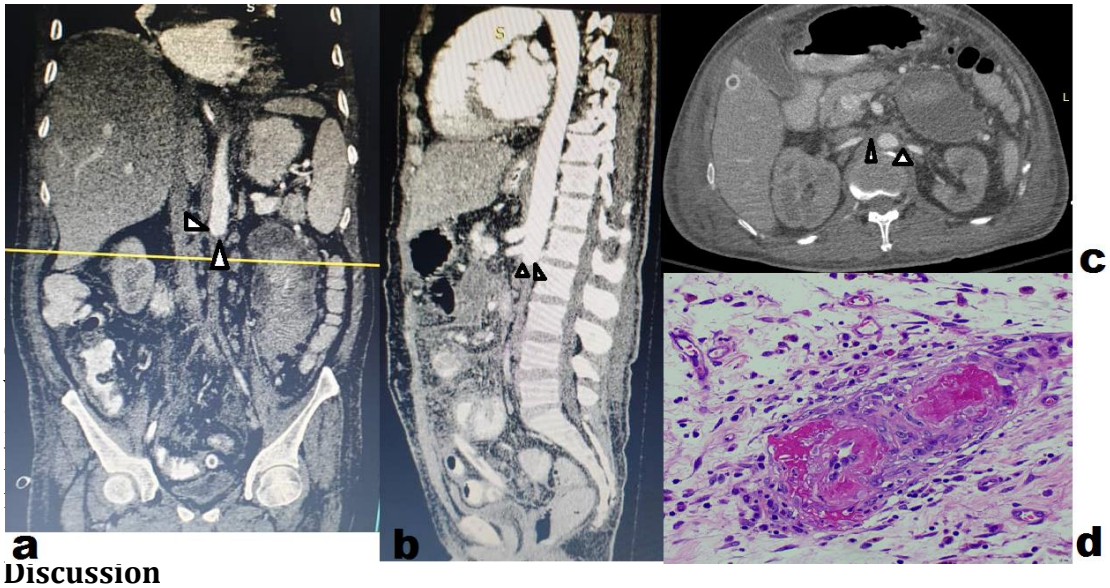
Computed tomography (CT) of abdomen showed small bowel obstruction. Urgent exploratory laparotomy was done and small bowel from duodenojejunal junction until terminal ileum was resected with end to end anastomosis. Subsequent CT angiography denoted multiple long segment small bowel thickening and non-opacification of aorta distal to the superior mesenteric artery (Fig. 1a,b), with common iliac arteries and bilateral renal artery stenosis (Fig. 1c).

The proximal aorta appeared to be chronically narrowed. No atherosclerotic plaques were found along the vessel walls and patient lipid profile was normal. With this imaging finding, Takayasu Arteritis was suspected. Blood pressure was then measured retrospectively and the difference in systolic blood pressure was noted to be more than 10 mmHg between the upper arms. Blood pressures of both lower limbs, however, were the same. Upper limbs

pulses were present. Bilateral popliteal arteries were difficult to palpate due to muscular limbs. But distal pulses (posterior tibial and dorsalis pedis arteries) were absent.

Subsequent autoimmune and antiphospholipid antibody screenings were negative. There was no thrombus identified from echocardiography. Further imaging of thoracic aortic arch was offered but the patient was unable to afford the cost. Histological examination of the excised specimen revealed leucocytoclastic vasculitis (Fig. 1d) with no obvious thrombus identified during serial sectioning. No crypt distortion or lamina propria cellular infiltrates noted in this histopathological specimen as well as no evidence of mucosal base plasmacytosis. Apart from that, no fistula and no fat wrapping were noted in the bowel segments intraoperatively and no evidence of granuloma was found.

Pulse methylprednisolone followed by prednisolone of 1 mg/kg/d, azathioprine and antiplatelet were administered. He was discharged well with no recurrence of abdominal symptoms and no further ischemic events. Unfortunately, follow-up was not feasible as he decided to fly back to his native country for continuation of care due to the financial constraint.



lete filling
a, b). Axial
heads) (c).
al vessel
utrophilic

Discussion

The differential diagnosis of this case included Buerger's disease (is a medium to small vessel disease but in this case it involved abdominal aorta as large vessel); fibromuscular dysplasia (no imaging features of this differential on computed tomography); giant cell arteritis involving aorta (but clinically he did not have any visual symptoms or headache); syphilis or lupus aortitis (but autoimmune and syphilis screening were negative); and neurofibromatosis (but clinically no such skin nodules in this patient). The clues for the diagnosis of Takayasu Arteritis in this case relied upon age of 40 years at disease onset, Asian ancestry, arterial hypertension, blood pressure discrepancy between arms, abdominal aortic involvement and high acute phase reactants.

Specifically, our diagnosis of Takayasu Arteritis was made based on the American College of Rheumatology 1990 criteria[4]. This patient fulfilled three of the six needed criteria: age at onset equal to 40 years; blood pressure difference > 10 mmHg systolic pressure between arms; and computed tomography angiography evidence of occlusion of abdominal aorta, not due to arteriosclerosis or fibromuscular dysplasia).

Further follow-up recommendations in this case would include imaging of thoracic aorta and its branches (to determine extent of large vessel involvement) as well as disease monitoring of response to treatment with positron emission tomography.

Th1 and Th17 cells which played pivotal role in cellular mediated inflammation have been implicated in its pathogenesis[5]. High-dose glucocorticoid is often the cornerstone management of active Takayasu Arteritis with methotrexate or azathioprine as suitable second line therapeutic options. JAK/STAT pathway[6] may rejuvenate our future treatment approach in abating inflammatory activity of Takayasu Arteritis.

CONCLUSIONS

The most striking finding in this case is that of disseminated Takayasu Arteritis involving intestinal gangrene with rare histopathological finding of leukocytoclastic vasculitis. Arterial hypertension, abdominal aorta involvement and high acute phase reactants in this case provided clues for the diagnosis. Early recognition can be life-saving in this case as immunosuppression is the first line therapy here. Without treatment, it could lead to other serious ischemic events involving myocardial infarction and stroke because it is a systemic disease, and could further result in disability and even death.

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