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Winslow pathway collaterals: an unusual arterial network in Takayasu arteritis

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Abstract

Takayasu arteritis which is reported more commonly from Asia and in females can present as middle aortic syndrome with lower limb claudication. We present a case of a young male with Takayasu arteritis with middle aortic syndrome and Winslow pathway collaterals with lower limb ischaemia, hypertension, coronary occlusion and stroke. The extensive collateral formation was visible as a clinical finding over the abdominal wall. The identification of these collateral pathways is essential in understanding the extent of haemodynamically significant disease and it alerts to the possibility of surgical injury during procedures like laparotomy or harvesting of internal thoracic artery for coronary artery bypass graft.

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Case presentation

A 20-year-old male presented with an eight-year history of bilateral lower limb claudication. His initial claudication distance of 30 minutes gradually worsened to 15 minutes. Six years ago he had a gangrene in the left fifth toe which was followed a year later by a gangrene in the left great toe. Evaluation had revealed hypertension with absent lower limb pulses. There was neither any bruit nor any asymmetry of pulses reported in the upper limbs. CT abdominal angiogram showed vessel wall thickening in the abdominal aorta along with long segment narrowing of the infrarenal abdominal aorta with collaterals reforming the external iliac arteries. He was prescribed antihypertensives, low-dose oral steroids and oral methotrexate but was non-compliant. Two years later he developed sudden onset weakness of the right hand. MRI brain revealed left parietal lobe infarction. He also noticed the development of dilated vessels over his abdomen starting from the flanks. He used nifedipine 10 mg daily as an antihypertensive for the next three years. At presentation to our department, he had a history of breathlessness, chest pain, palpitations and orthopnoea of ten days' duration. He reported persistent claudication pain in the lower limbs without any decrease in claudication distance. There was no history of fever, cough or haemoptysis. He denied any history of oral or genital ulcers, skin rash or ocular complaints. He was not a smoker. On examination, he had accelerated hypertension (BP = 220/110 mmHg), lower limb pulses were absent bilaterally while the upper limb and neck pulses were bounding and symmetric without any bruit or carotidynia. The first and the fifth toes on the left side had mild pulp loss at the tips. The abdominal wall had thick-walled pulsatile vessels both above and below the umbilicus (Figure 1). The rest of the systemic examination was unremarkable. Laboratory investigation revealed thrombocytosis (platelets 7.63 lakh/ml) and a raised erythrocyte sedimentation rate (ESR) of 50 mm in the first hour. The chest radiograph was suggestive of pulmonary oedema, the electrocardiogram (ECG) showed ST elevation in the anterior chest leads and troponin I level was elevated (10078 ng/ml). Echocardiography showed regional wall motion abnormality along the left anterior descending artery (LAD) territory with an ejection fraction of 30%. Coronary angiography revealed single vessel disease with type-3 90% mid-LAD stenosis. Magnetic resonance angiography (MRA) of the aorta showed narrowing of the distal descending thoracic aorta extending up to the origin of the superior mesenteric artery (SMA). Bilateral renal arteries and the SMA were reformed via collaterals. There was near-total stenosis of the infrarenal aorta up to the level of bifurcation. Right common iliac artery, left internal and external iliac arteries were reformed via collaterals. Extensive collateral formation was also noted between the superior and the inferior epigastric arteries in the abdominal parietal wall (Figures 2 and 3). Both kidneys were normal in size and signal intensity. MRA of the intracranial vessels

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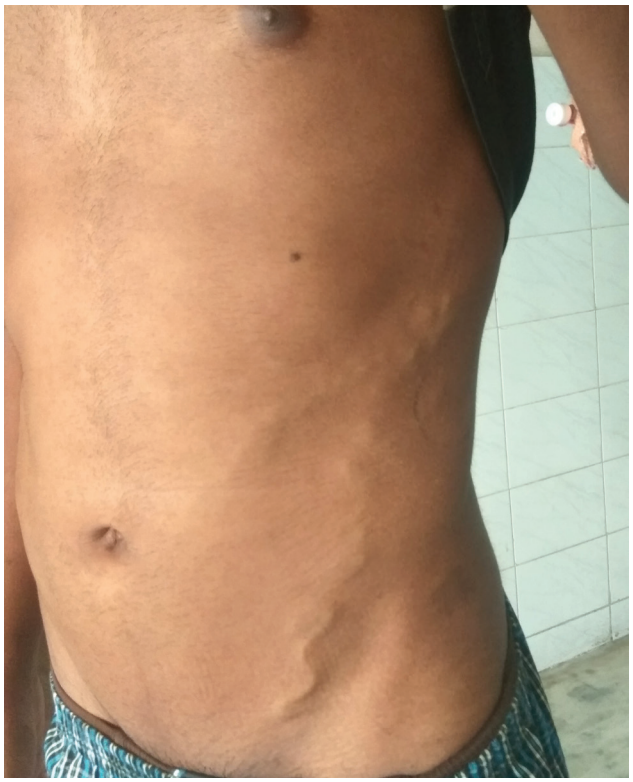
Figure 1 Prominent arterial collaterals over the abdominal wall in a patient with Takayasu arteritis with middle aortic syndrome (MAS)



showed normal intra- and extracranial vessels with a chronic infarct in the left parietal lobe. A diagnosis of Takayasu arteritis, Numano type III, with middle aortic syndrome and Winslow pathway collaterals with lower limb ischaemia, hypertension, anterior wall myocardial infarction secondary to LAD stenosis and left parietal lobe infarction was made. His Indian Takayasu Clinical Activity Score (ITAS) was ten. The patient was treated with high-dose oral steroids, antihypertensives, methotrexate and antiplatelets and aortic stent placement was planned after the resolution of systemic inflammation.

Discussion

Takayasu arteritis is a large-vessel vasculitis primarily affecting the aorta and its primary branches resulting in stenosis and aneurysms. It mainly affects young women with higher prevalence in Asian populations. Middle aortic syndrome (MAS) refers to a segmental or diffuse narrowing of the abdominal aorta (± descending thoracic aorta) and accounts for 0.5–2% of all the cases of aortic stenosis.[1] Sen et al. first described MAS in 16 patients with inflammatory aetiology in 1963.[2] MAS refers to obstructive lesions of the mid-aorta due to any aetiology. It causes renovascular hypertension in children and young adults with end-organ damage along with lower limb ischaemia and is associated with extensive collateral formation between various systemic and visceral arteries.[1]

Systemic-systemic collaterals develop between intercostal, internal thoracic, lumbar, deep circumflex iliac, inferior epigastric and obturator arteries in occlusion of the distal abdominal aorta. On the other hand, in more proximal abdominal aorta involvement the visceral-visceral or visceral-systemic collaterals form and are derived from the coeliac trunk, SMA and inferior mesenteric artery (IMA). One important connection is the marginal artery of Drummond connecting SMA with IMA. The basis for this collateralisation of the blood supply lies in the embryonic dorsal arterial system which has the potential to form interconnections.[3]

The Winslow pathway is one such unusual pathway between systemic arteries described for the first time in 1954 by Gottlob in obstructive atherosclerotic aortic disease.[4] This bypasses the descending thoracic and abdominal aorta and connects the internal thoracic artery and external iliac artery via superior and inferior epigastric arteries sequentially. This pathway is crucial for the blood supply to the lower limbs.

Babu et al. reported three patients with Takayasu arteritis with MAS and Winslow pathway collaterals. All three patients were females with a long duration of symptoms (6–15 years). Two had complete aortic occlusion and all had extensive systemic-systemic and systemic-visceral collaterals.[5]

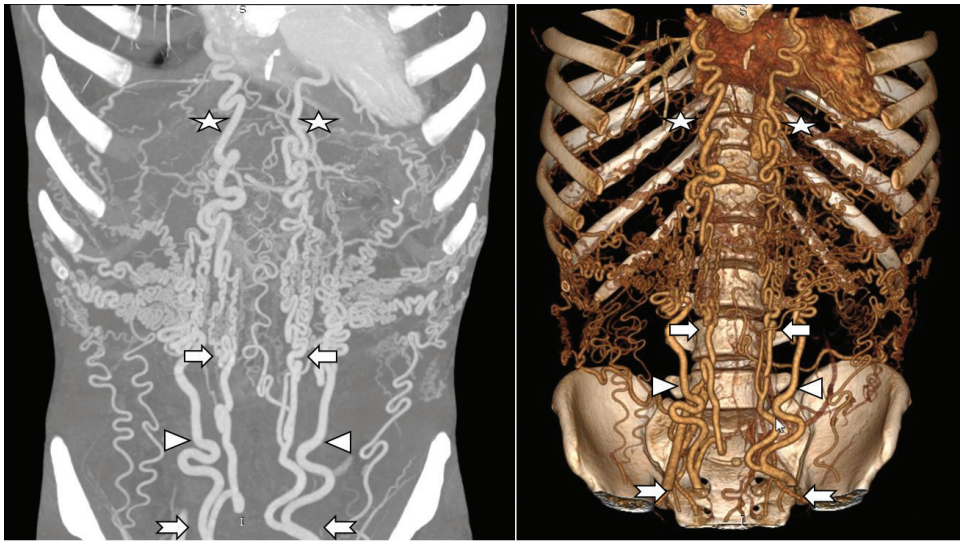
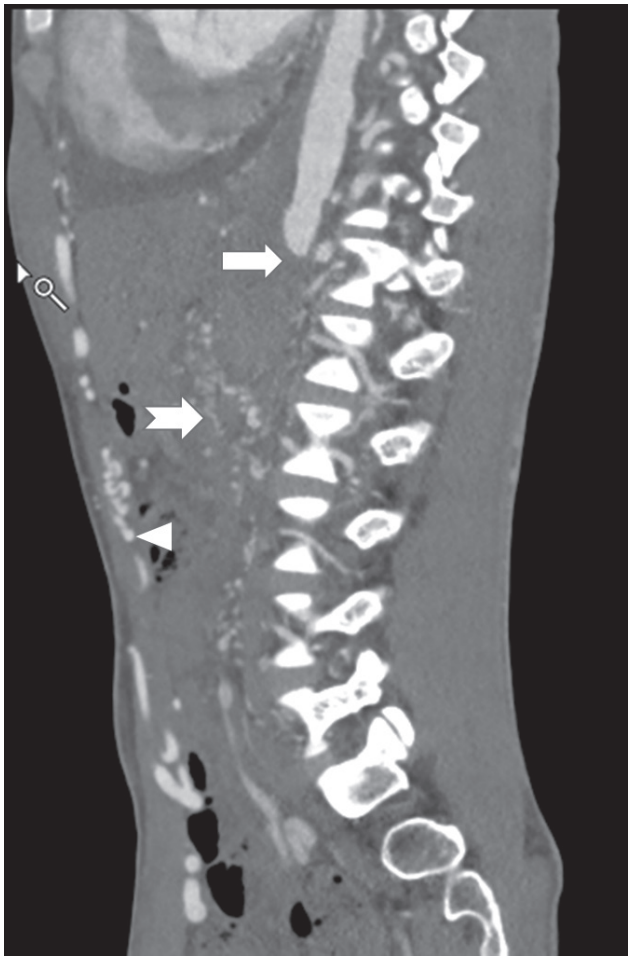


Figure 2 Computed tomography angiography MIP and VRT images showing well-developed Winslow pathway collaterals bilaterally that start as enlarged internal thoracic arteries (stars) in the thorax and continue as multiple vascular channels that anastomose the superior and inferior epigastric arteries (arrows and arrowheads respectively) and finally drain into the external iliac arteries (notched arrows).

MIP: maximum intensity projection; VRT: volume rendering technique

Figure 3 Sagittal contrast-enhanced computed tomography angiogram showing complete occlusion of the infrarenal segment of the abdominal aorta (arrow) with multiple collaterals seen in the mesentery (notched arrow) and in anterior abdominal wall (arrowhead)



CT angiography using the multi-slice technique provides excellent visualisation of the Winslow pathway collaterals and is preferred over conventional angiography which is invasive and tends to miss these collaterals unless ascending aortic angiography is performed and these collaterals are tracked down to the iliac level.[6]

Our patient had infrarenal aortic narrowing initially which progressively involved the suprarenal part of the abdominal aorta and the distal descending thoracic aorta making the Winslow pathway the most important collateral pathway. The external and internal iliac arteries were reformed on both sides. This case is unusual due to the presence of this rare collateral pathway. The clinical finding of arterial collaterals over the abdominal wall is also unusual in a patient of suspected systemic vasculitis. A close mimicker is a vena-caval obstruction secondary to Behçet's disease leading to venous collaterals over the abdomen which has a soft, collapsible wall and can be milked out to ascertain the direction of the flow. The arterial collaterals, on the other hand, have a thick wall and are often pulsatile.

Another unusual manifestation in this patient was digital gangrene. Only ten such cases have been reported in the literature so far with the majority of them being females with Numano type V followed by type I involvement and more frequent involvement of the lower limbs. The causative factors for the gangrene may be thrombus formation due to inflammation of walls of large vessels or presence of antiphospholipid antibodies and/or antiendothelial cell antibodies.[7]

Takayasu arteritis with MAS mainly manifests with refractory hypertension and its complications along with symptoms of lower limb ischaemia. Invasive options available for treatment are percutaneous stenting or surgical repair, the success of which mainly depends on the current disease activity and inflammation.[5,8] Intervention is planned for this patient after controlling inflammation with oral steroids and methotrexate to improve the outcome of surgical treatment.

The Winslow pathway demonstrates how extensively collateral formation can take place in occlusive arterial diseases like Takayasu arteritis how it helps in sustaining the function of the distal organs and tissues. The identification of these collateral pathways is essential in understanding the extent of haemodynamically significant disease and the blood supply to the tissues distal to the occlusion. We also need to be aware of any accidental surgical injury that may take place during procedures like laparotomy or harvesting of internal thoracic artery for coronary artery bypass graft.[9,10]

Conclusion

We present a case of Takayasu arteritis with Winslow pathway collaterals. The prominent arterial collateral network on the abdominal wall is a clinical pointer towards the severity of aortic occlusion and care should be taken to avoid any inadvertent injury during surgical procedures.

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