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Letter to the Editor

Malignant hypertension, dissection of aorta, cardiac tamponade and monoparesis—Unusual presentation of Takayasu's arteritis: Clinicopathological correlation

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Abstract

We report a case of a 24-year-old woman with an unremarkable past medical history who was presented to the emergency department with acute onset of breathlessness and weakness of right lower limb. Clinical examination was suggestive of malignant hypertension with acute left ventricular failure and acute ischemia of right lower limb. Colour Doppler and CT findings were consistent with dissection of entire aorta with extension into its major branches. She died in less than 18 h after admission following cardiac tamponade. Autopsy revealed left renal artery stenosis with features of Takayasu's arteritis with intimal rupture in the abdominal aorta with Stanford type A dissection of aorta extending to all the major branches of aorta, and hemopericardium. This case demonstrates a rare example Takayasu's arteritis involving left renal artery leading to secondary hypertension presenting as malignant hypertension with fatal dissection of aorta with cardiac tamponade and emphasizes the varied presentations of this disease and importance of early diagnosis and interventions to prevent these fatal complications.

Keywords: Aortic dissection; Takayasu's arteritis; Malignant hypertension

1. Case report

A 24-year-old housewife was admitted to the emergency department with complaints of progressive breathlessness and palpitation of 15 days duration and sudden onset of weakness of right lower limb. The breathlessness rapidly progressed to orthopnoea at admission. The weakness of right lower limb was associated with severe pain. She had no past history suggestive of hypertension, cardiac illness or connective tissue diseases. She gave a history of significant weight loss and loss of appetite over 6 months. She had four children and no significant events during antenatal or postnatal period noted.

At admission to the emergency department, general physical examination revealed a thin built underweight young female who was tachypnoeic at rest. No characteristic signs of Marfan's syndrome were found. She was pale and cyanosed. Pulse rate was 126/min. All pulses in right lower limbs were absent and limb was cold to touch. All other peripheral pulses were palpable. Thrill was felt in the epigastrium just above the umbilicus. Blood pressure was 220/130 mm of Hg. Optic fundus examination showed grade IV hypertensive retinopathy changes. Examination of cardiovascular system was suggestive of congestive cardiac failure with pulmonary edema. Central nervous system examination was suggestive of ischemic paresis of right lower limb.

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Primary laboratory parameters were as follows: Hemoglobin was 7 G/dl with microcytic normochromic anemia. ESR – 80 mm at 1 h, RBS – 125 mg/dl, blood urea – 184 mg/dl, serum creatinine – 12 mg/dl, serum electrolytes were within normal range. Lipid profile was normal. Urine routine showed albumin in traces. ECG was suggestive of sinus tachycardia with gross LVH. Chest radiograph showed LV type cardiomegaly with pulmonary edema. 2D echocardiogram showed concentric LVH with global hypokinesia. LVEF was 47%. Aortic valve was tricuspid. There was no evidence of pericardial effusion. USG-abdomen revealed bilateral shrunken kidneys with features of grade III nephropathy. Emergency colour Doppler of abdominal aorta documented dissection of abdominal aorta arising at the level of subdiaphragm extending up to the right common iliac artery (Fig. 1A,B). It also demonstrated the intimal tear below the level of renal arteries. Other serological investigations like VDRL, HIV, RA factor, and ANA were negative. Emergency CT angiogram documented dissection of entire length of aorta extending up to the bifurcation of the abdominal aorta (Fig. 1C,D). Cardiothoracic and vascular surgery opinion was taken and since patient's general condition was poor attempt was made to stabilize the patient. In view of no response to intravenous antihypertensive medications she was subjected for haemodialysis. During this process patient developed sudden onset of pulmonary edema and patient was in shock, heart sounds muffled attempt for emergency pericardiocentecis failed and patient expired within 18 h of admission.

At clinical autopsy heart weighed 540 g with gross left ventricular hypertrophy. There was haemopericardium of about 175 ml. Intimal tear was noted 2 cm below the opening of renal arteries. The perforation dissected along the wall of the aorta both in a retrograde and antegrade fashion to involve the entire length of aorta. The intima was completely transected circumferentially and had formed a free cylinder inside the aorta (Fig. 2A). Dissection extended to all major branches of aorta. There was no aneurysmal dilatation. Distally the aorta showed extensive atherosclerotic changes involved by thickening and cobblestoned with tree bark appearance. Bilateral kidneys were shrunken in size. There was total stenosis of left renal artery. Histopathological examination of autopsy specimen showed extensive atherosclerosis of aorta with dissection (Fig. 2B). Multiple sections of left renal artery showed chronic inflammation, fibrosis and thickening of the intima and adventitia with multiple Langhan's type giant cells suggestive of granulomatous arteritis (Fig. 2C). Sections from the kidney showed malignant

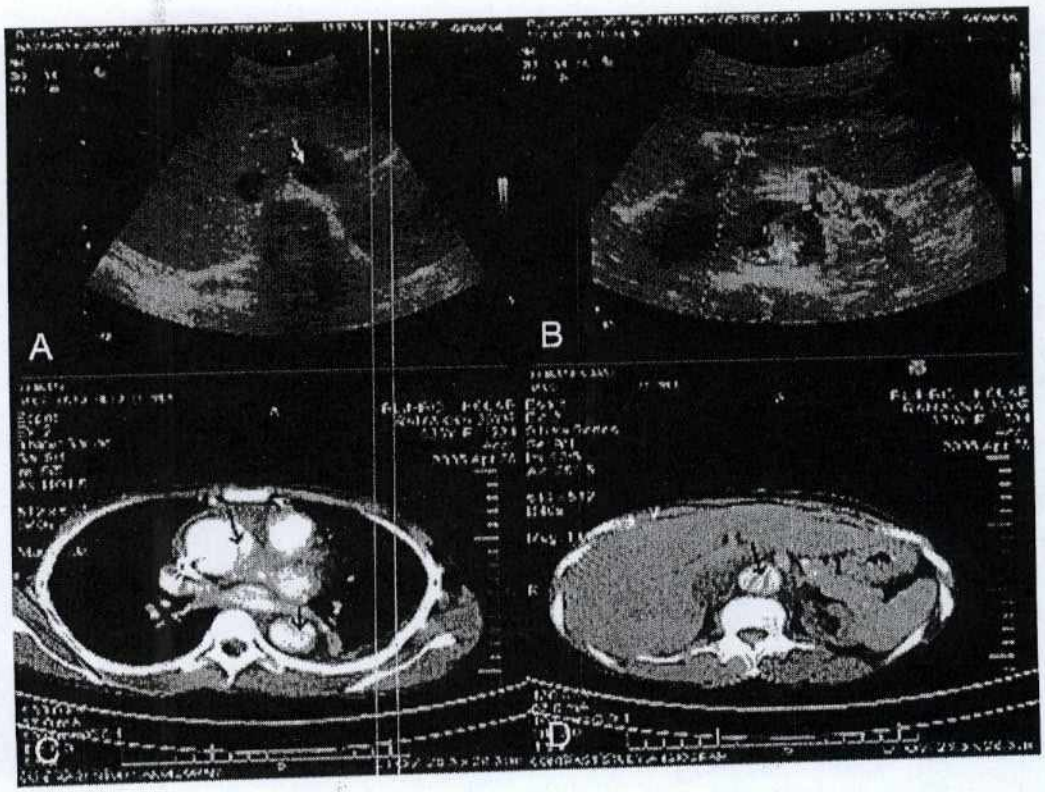

Fig. 1. A: 2D Echo image showing dissection (arrow) of abdominal aorta. B: Doppler image showing dissection of abdominal aorta with true and false lumen. C: Contrast CT of thorax showing dissection (arrow) of both ascending and descending aorta. D: Contrast CT of abdomen showing dissection (arrow) of abdominal aorta.

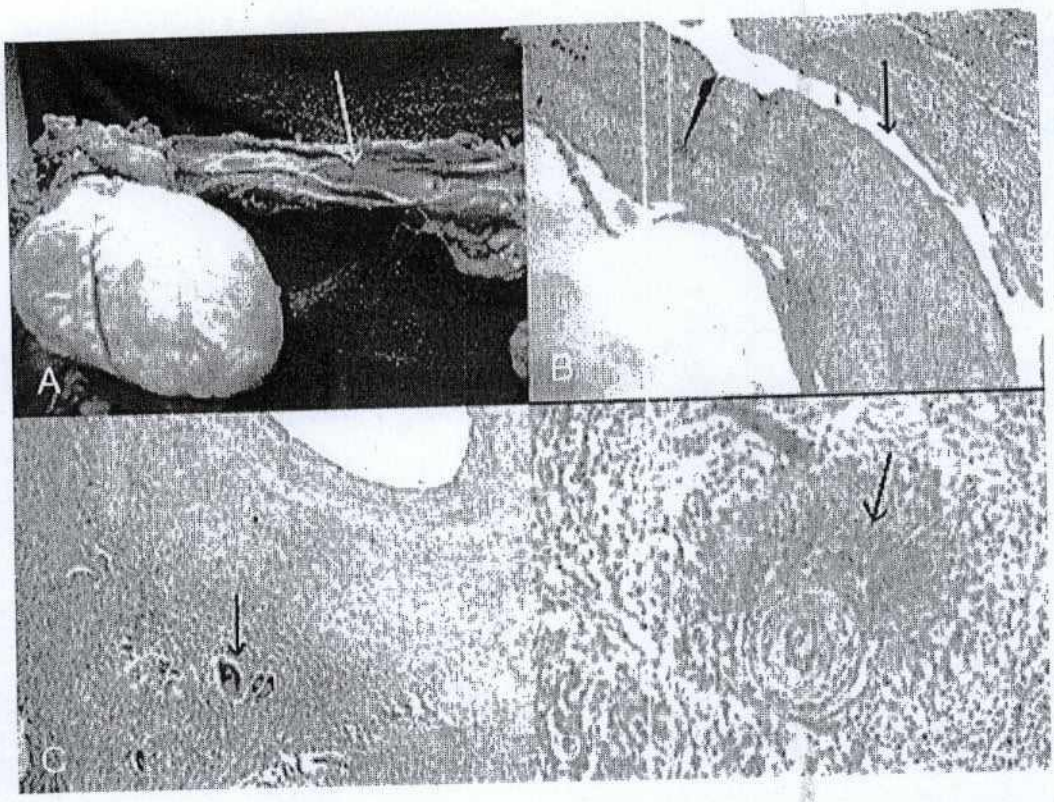

Fig. 2. A: Autopsy specimen showing cardiomegaly with circumferential dissection of aorta (arrow). B: Histopathological examination of aorta showing extensive atherosclerosis with intimal thickening and dissection (arrow). C: Histopathological examination of left renal artery showing chronic inflammation, fibrosis and thickening of the intima and adventitia with multiple Langhan's type giant cells (arrow). D: Histopathology of renals showing onion skin proliferation with necrotising arteriolitis (arrow).

nephroscerotic changes suggestive of malignant hypertension (Fig. 2D).

2. Discussion

Takayasu's arteritis (TA) is a chronic vasculitis mainly involving the aorta and its main branches. It induces clinically varied ischaemic symptoms due to stenotic lesions or thrombus formation. More acute progression causes destruction of the media of the arterial wall, leading to the formation of aneurysms or rupture of the involved arteries [1]. TA commonly affects patients in 2nd and 3rd decade of life. Females are affected more commonly than males. The onset of symptoms occurs before 30 years of age in most patients. Comparative studies of different ethnic groups and the clinical manifestations of Takayasu's arteritis have shown that many Japanese patients present with weak or absent radial pulse, disturbed vision and aortic regurgitation. In contrast, Indian patients presented with hypertension probably due to abdominal aortic lesions, including involvement of the renal arteries [2]. The major causes of morbidity and mortality in Indian patients with TA is due to severe uncontrolled hypertension and its effect on heart, kidney and brain.

Isolated abdominal aortic aneurysm with perforation, dissection, and rupture is probably the rarest manifestation of Takayasu's arteritis. There are only few cases of spontaneous dissection of aorta in granulomatous arteritis reported in the literature, but most of them were in giant cell arteritis and in above 45 years old [3–6]. Our case report is unique in many aspects. Dissection of aorta is the rarest complication of Takayasu's arteritis. A case of aortic dissection beginning in the abdominal aorta is considerably rare. Previous reports have shown only a 1% to 2% incidence of primary spontaneous abdominal aortic dissection among cases of aortic dissection [7]. There is only one case report of spontaneous retrograde acute abdominal aortic dissection resulting in hemopericardium as its fatal complication in the literature [8]. In our case, the spontaneous dissection of abdominal aorta extended in retrograde direction to cause cardiac tamponade and in antegrade direction resulting in ischemic monoparesis of right lower limb. Monoparesis is very rare presentation of Takayasu's arteritis and to best of our knowledge this is the only case report in English literature.

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Appendix A. Supplementary data

Supplementary data associated with this article can be found, in the online version, at doi:10.1016/j.ijcard.2007.12.042.

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