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Clinical Case Report

Uncommon associations and catastrophic manifestation in Takayasu arteritis: an autopsy case report

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ABSTRACT

Takayasu arteritis, a chronic inflammatory vasculitis affecting aorta and its major branches, is complicated by stenosis, occlusion, and aneurysm formation. The aneurysm formation and subsequent complications such as heart failure, aortic regurgitation, and aneurysm rupture can be fatal. The aortic aneurysm rupture is a rare and fatal complication with only a few cases reported in the English literature. The involvement of coronary artery in Takayasu occurs in about 10% patients, and the coronary artery aneurysm is the least common manifestation. Here, we describe a case of Takayasu arteritis with abdominal aortic aneurysm rupture and coronary artery aneurysm. This patient also had associated systemic inflammatory diseases like sarcoidosis and Hashimoto's thyroiditis.

1. Introduction

Takayasu arteritis (TA), first reported by Dr. Mikito Takayasu, is a chronic inflammatory vasculitis affecting aorta and its major branches characterized by stenosis, occlusion, and aneurysm formation. The cases have been reported worldwide; however, it has the highest prevalence in Asian countries, and females younger than 40 years are affected more commonly than males. The disease is characterized by an acute phase followed the chronic phase which is dominated by vascular symptoms such as hypertension, diminished or absent pulses, limb claudication, or localized pain. The long-term prognosis of patients with TA is good; however, the aneurysm formation and subsequent complications such as heart failure, aortic regurgitation, and aneurysm rupture can be fatal [1]. In addition to involvement of aorta and its major branches, small percentages of patients have coronary artery involvement in different forms of which aneurysm formation is rarest [2].

The present case describes the simultaneous occurrence of abdominal aortic aneurysm with rupture and coronary artery aneurysm. In addition, the patient had immune mediated associations in form of sarcoidosis and Hashimoto's thyroiditis.

2. Case report

A 38-year-old female presented in the emergency in unconscious state. Three hours prior to the admission, she complained of sudden abdominal pain followed by abdominal distension and loss of consciousness. Her blood pressure and vitals were unrecordable with low Glasgow score. Cardiopulmonary resuscitation was carried out; however, the patient could not be revived and was declared dead.

At the age of 23 years in 1998, the patient first presented with pedal edema and was diagnosed to have hypertension and was started on clonidine. On imaging, she was found to have Takayasu disease based on asymmetric dilatation of aorta. Subsequent angiography showed bilateral renal artery stenosis for which left renal artery balloon dilatation and stenting was done. In year 2010, she was hospitalized for lower abdominal pain along with backache. On examination, brachial pulse was weak with absent left radial pulse. Both side popliteal and posterior tibial pulses were also absent. There was pressure difference of 20 mm of mercury in both upper limbs. Per abdomen examination revealed prominent aortic pulsations with aortic and left renal bruit. Noncontrast-enhancing computerized tomography scan showed presence of fusiform dilatation and thrombosis at the bifurcation of abdominal aorta and small-sized right kidney with multiple calculi (Fig. 1A). Magnetic resonance angiography showed fusiform dilatation with tortuosity of abdominal aorta with significant luminal narrowing in the proximal part. She was managed on antihypertensives and analgesics. Two years later on a routine outpatient follow up, a visible pulsatile lump below umbilicus was noticed which was extending to the left side. She was counseled for potential complications, and endovascular repair surgery was

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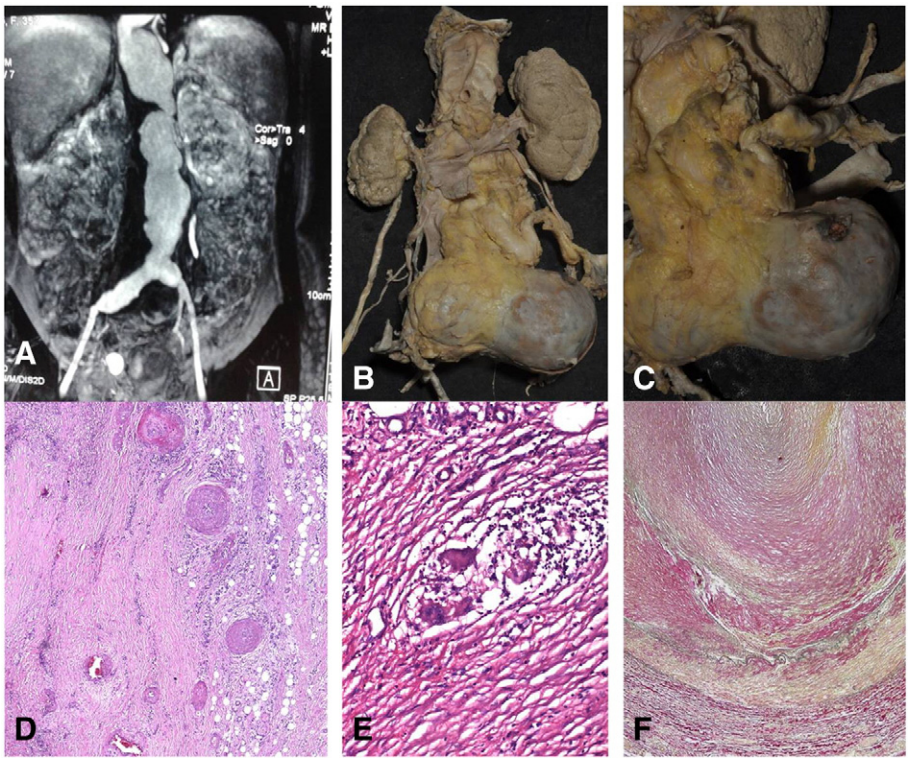

Fig. 1. (A) Noncontrast computed tomography showed fusiform dilatation of abdominal aorta and thrombosis at bifurcation of aorta and both iliac arteries; (B) Gross photograph showing stenosis and dilatation of abdominal aorta with large aneurysm at the bifurcation. Adventitia is markedly thickened with adhesions to surrounding structures; (C) Closer view to show the ruptured site on posterior wall. Photomicrographs showing (D) marked fibrosis of aortic adventitia and vasa vasorum showing concentric hypertrophy with perivascular lymphoplasmacytic cell infiltrate; (E) Giant cells in the media; (F) EVG stain highlighting break in internal elastic lamina.

advised. However, because of financial constraints, patient lost to follow-up. In 2013, she was brought in unconscious state and died after a brief hospital stay. Partial autopsy was performed after written consent from the patient's relatives.

2.1. Autopsy findings

Peritoneal cavity on opening yielded 2 L of blood with blood clots. Whole length of aorta and its main branches showed dense fibrosis with adhesions. The intima showed marked wrinkling with tree bark-like appearance with superimposed atherosclerotic plaques, which were more prominent at the origin of branches. Skip lesions with relatively uninvolved areas in between were also seen in descending thoracic aorta. There was stenosis at the level of renal arteries while the post stenotic segment was markedly dilated. A large fusiform aneurysm measuring 10×7×5 cm was present at the bifurcation which was extending into proximal part of right common iliac artery (Fig. 1B). The posterior surface of aneurysm showed a rupture site measuring 1.5×1.4 cm (Fig. 1C). The aneurysmal sac showed a large fresh thrombus almost occluding the whole lumen. Microscopically the involved segments showed dense adventitial fibrosis with prominent vasa vasorum showing intimal hyperplasia, medial thickening, and moderate lymphoplasmacytic infiltrate (Fig. 1D). Few giant cell granulomas were also seen (Fig. 1E). The inflammatory infiltrate was extending to the media which was also showing marked fibrosis. The loss of elastic fibers and thinning of media was highlighted on elastic von Gieson (EVG) stain (Fig. 1F). Intima showed proliferation with luminal narrowing and superimposed atherosclerotic changes and areas of calcification. All the major branches showed changes of chronicity in the form of adventitial fibrosis, moderate lymphoplasmacytic cell infiltrate, and replacement fibrosis in the media.

Heart weighed 425 g, and the epicardial surface on the right side showed a bulge at atrio-ventricular groove. On dissection, it showed a fusiform aneurysm of right coronary artery starting from its origin and measuring 2 cm in length. The aneurysmal sac showed a thrombus; however, the lumen was patent (Fig. 2A–C). The left coronary artery and its branches showed mild luminal narrowing with atherosclerotic changes. There was significant left ventricular hypertrophy with reduction in the volume of cavity. Microscopically, the wall of the coronary artery showed similar changes as seen in aorta (Fig. 2D–E). The heart showed patchy areas of interstitial fibrosis; however, no fresh myocardial infarction (MI) was noted.

Lungs together weighed 675 g with focal areas of pleural dullness. Cut surface did not show any focal lesions, and pulmonary arteries were unremarkable. Histologically, many compact noncaseating interstitial granulomas were seen in the subpleural, paraseptal, and bronchocentric location (Fig. 3A). These were composed of epithelioid histiocytes, Langhans and foreign body type of giant cells, lymphocytes, and scattered plasma cells. The Ziehl- Neelsen stain for acid-fast bacilli was negative. These granulomas were T-cell rich with dominant CD4 positive helper T-cells. The right kidney was small and granular with dilated pelvicalyceal system containing few irregular calculi. Left kidney was of normal size with granular outer surface. A metallic stent was present at the origin of left renal artery while right renal artery showed atherosclerotic narrowing at the origin. Microscopically, the renal parenchyma showed changes of benign arterionephrosclerosis.

Thyroid was firm, fibrotic, and weighed 21 g. Microscopically, there was extensive lymphocytic infiltrate with lymphoid follicles causing destruction of thyroid follicles and dense fibrosis (Fig. 3B). An incidental adenoma was also noted in right superior parathyroid gland composed of clear cells. The superior and inferior mesenteric arteries were extensively involved by the disease process causing ischemic changes in the intestines in the form of submucosal fibrosis and fat infiltration.