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Clinical Case Report

Aortic fibromuscular dysplasia complicated by dissection: a case report and review of literature☆,☆☆

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ABSTRACT

Fibromuscular dysplasia (FMD) is an idiopathic, segmental, nonatherosclerotic, non-inflammatory vascular disease, which is often complicated by the occurrence of dissection. Although it is known to occur in all arteries, aortic involvement is relatively rare. To date, 33 cases of aortic FMD have been reported in available English literature, among which only three cases have been complicated by the occurrence of dissection. We describe the case of a 40-year-old woman diagnosed with aortic FMD complicated by the occurrence of a type A aortic dissection. Non-invasive imaging revealed an ascending to descending thoracic aneurysm measuring 8 cm in diameter associated with dissection. Histopathologically, a segment of the wall of the aneurysm showed architectural disorganization of the aortic wall with loss of elastic fibers, collagen deposition, and irregular proliferation of smooth muscle cells in the intima and media—features suggesting FMD. No atheromatous plaque or medial cystic degeneration was observed in the aorta. Although aortic FMD is sometimes fatal, it is often very difficult to diagnose using imaging techniques. Therefore, performing a histopathological diagnosis is very important and should be emphasized.

1. Introduction

Fibromuscular dysplasia (FMD) is an idiopathic, segmental, nonatherosclerotic, non-inflammatory vascular disease occurring primarily in middle-aged women. Although it commonly affects medium-sized arteries, especially the renal arteries, extracranial carotid arteries, and vertebral arteries, it can be occurred in almost all arteries, and involvement of two or more arteries at different anatomical sites is not rare [1,2]. Stenosis, aneurysms, and dissection are common complications associated with this condition. Reportedly, a dissection or aneurysm has been found in 14.3%–19.7% and 17.0%–26.2% of FMD patients, respectively [2,3]. Aortic involvement is rare, and to the best of our knowledge, among the 33 cases reported in English literature [4–28], only 3 cases have demonstrated the occurrence of dissection as a complication [15–17].

Histopathological findings of FMD are loss of elastic fibers, collagen deposition, and proliferation of smooth muscle cells and/or myofibroblasts leading to architectural disorganization in the arterial wall. Based on the arterial wall layer (intima, media, or adventitia) in which the lesion predominates, FMD is classified into three categories. Medial FMD is subclassified into medial fibroplasia, perimedial fibroplasia, and medial hyperplasia [1,29]. The most common subtype is medial fibroplasia characterized by stenosis and poststenotic dilatation caused by alternating areas of thinned media and thickened collagen-containing medial ridges. Perimedial fibroplasia is characterized by the presence of a homogenous fibrous collar at the junction of the media and adventitia. The intima and internal elastic lamina appear normal. Intimal fibroplasia demonstrates an intimal thickening due to collagen deposition without lipid accumulation and inflammation. Medial hyperplasia and periarterial fibroplasia are the rarest subtypes. These subtypes correlate well with clinical imaging; therefore, almost all patients with FMD are diagnosed using catheter-based angiograms, computed tomography angiogram (CTA), or magnetic resonance

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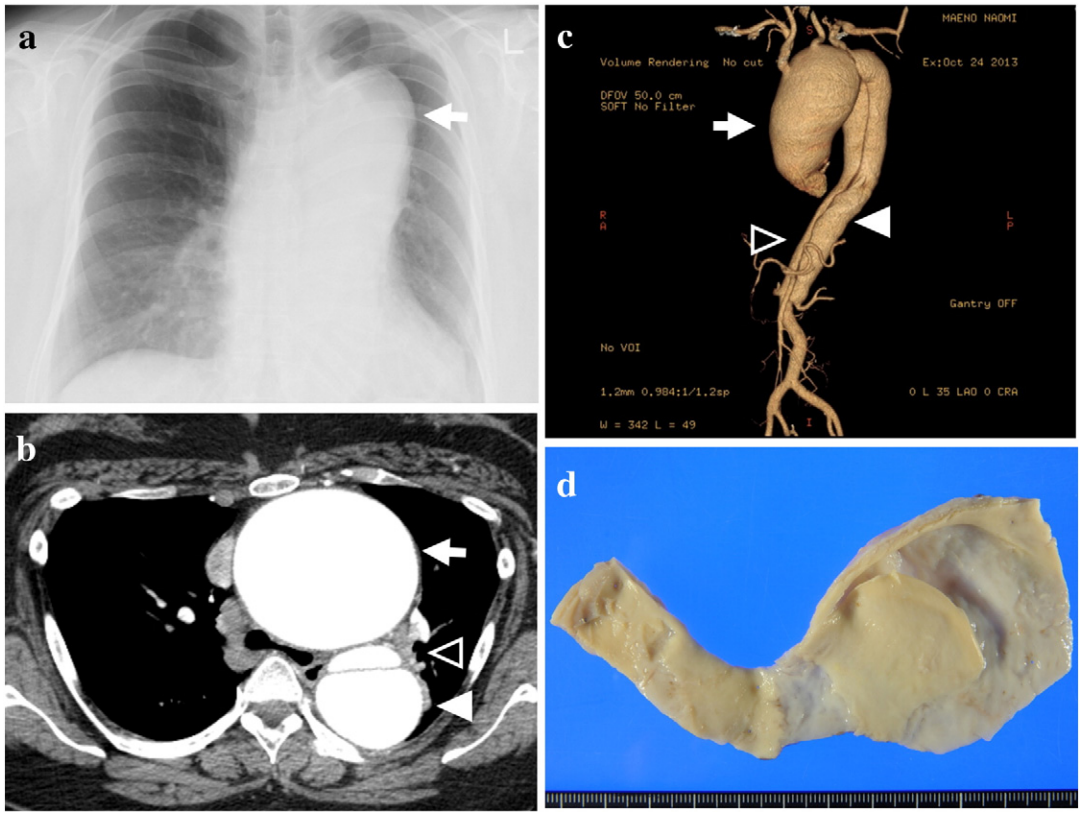

Fig. 1. Radiographic and macroscopic findings of the resected aorta. (a) A chest x-ray showing widening of the aortic silhouette (arrow). (b) Axial image from a CT scan showing an 8-cm thoracic aortic aneurysm of the ascending aorta (arrow) and dissection (closed arrow head: true lumen, open arrow head: false lumen). (c) CTA demonstrates a thoracic aortic aneurysm (arrow) and aortic dissection (closed arrow head: true lumen, open arrow head: false lumen). (d) Macroscopically, the aneurysmal wall of the dilated ascending aorta shows a rough luminal surface and no atheromatous plaque.

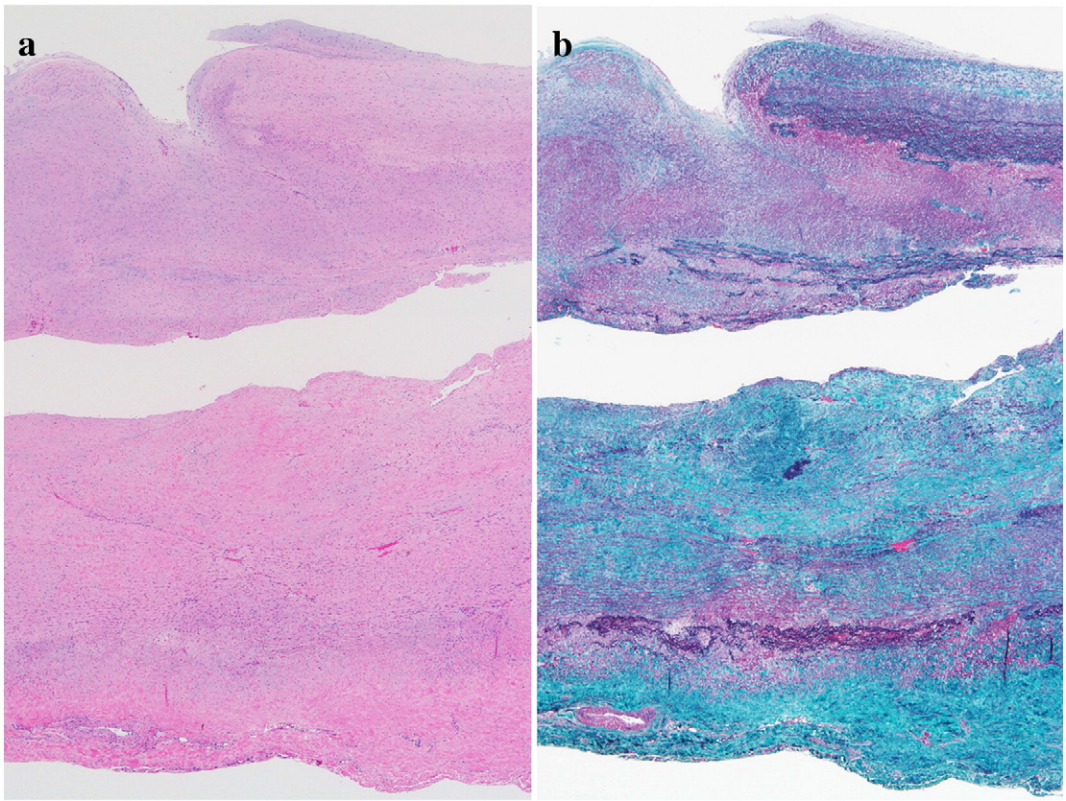

Fig. 2. Microscopic findings of aortic dissection. (a) Separate layers of the arterial wall are not clearly distinguishable, and dissection is observed in 1/2 to 1/3 from the luminal side. Extensive myxoedematous changes are noted in the media. (b) Loss of elastic fibers in the media and collagen deposition are clearly observed using elastica Masson's trichrome stain.