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Preoperative Evaluation of Annuloaortic Ectasia in Marfan Syndrome Using ECG-gated MDCT Angiography

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Abstract : Aortic abnormalities in Marfan syndrome are represented pathologically by cystic medial necrosis leading to the dilatation of the aortic annulus. Multidetector-row computed tomography (MDCT) angiography is useful for the complete examination of the entire aorta in patients with acute aortic syndrome. We report preoperative evaluation of annuloaortic ectasia using electrocardiogram (ECG)-gated MDCT angiography in a patient with Marfan syndrome

Key Words : Annuloaortic ectasia, Aortic dissection, Marfan syndrome, Multidetector-row CT

Introduction

Aortic abnormalities in Marfan syndrome (MS) are represented pathologically by cystic medial necrosis leading to the dilatation of the aortic annulus. Characteristic cardiovascular features include aortic valve incompetence and regurgitation dilatation associated with dilatation of the aortic annulus and progressive dilatation of the proximal ascending aorta leading to aortic dissection or rupture. Echocardiography and magnetic resonance imagings have been commonly used for aortic screening such as in annuloaortic ectasia and cardiovascular complications. Multidetector-row computed tomography (MDCT) angiography is useful for complete examination of the entire aorta in patients with acute aortic syndrome. ECG-gated MDCT angiography, providing detailed cardiovascular anatomic information, may be a powerful diagnostic tool for the evaluation of cardiovascular complications and the planning of optimal surgical treatment in MS.

Case report

A 35-year-old man visited Keimyung University Dongsan Medical Center with chest discomfort for 2 days. His sister suddenly died of aortic rupture 15 years ago. Auscultation revealed regular heart beats with grade II/VI diastolic murmurs. A chest radiograph showed a large, elongated thoracic cage, and cardiomegaly

(not shown). Transthoracic echocardiography revealed dilated aortic annulus (47.5 mm in diameter), dilated ascending aorta (60.3 mm in diameter), moderate aortic regurgitation, left cardiac chamber enlargement, and mild mitral regurgitation (not shown).

ECG-gated MDCT angiography was performed by using a Sensation 16 scanner (Siemens, Forchheim, Germany). Data was retrospectively reconstructed at 10% (early systole) and 70% (mid-diastole) of the R-R interval. All images were postprocessed on a workstation (Syngo, Wizard; Simens Medical solutions). Sagittal oblique multiplanar reformation (MPR) image at mid-diastole showed left ventricular enlargement and annuloaortic ectasia with the precise assessment of diameter at the level of aortic annulus (37.4 mm), sinuses of Valsalva (58.7 mm) and sinotubular ridge (51 mm) (Fig. 1). Double-oblique MPR image at mid-diastole demonstrated a triangular coaptation defect of the aortic valve (Fig. 2). Sagittal oblique MPR image at mid-diastole showed annuloaortic ectasia, intimal flap beginning just distal to the origin of the left subclavian artery, and focally thrombosed false lumen in one plane (Fig. 3). The patient underwent aortic valve replacement, ascending aorta replacement, and coronary reimplantation.

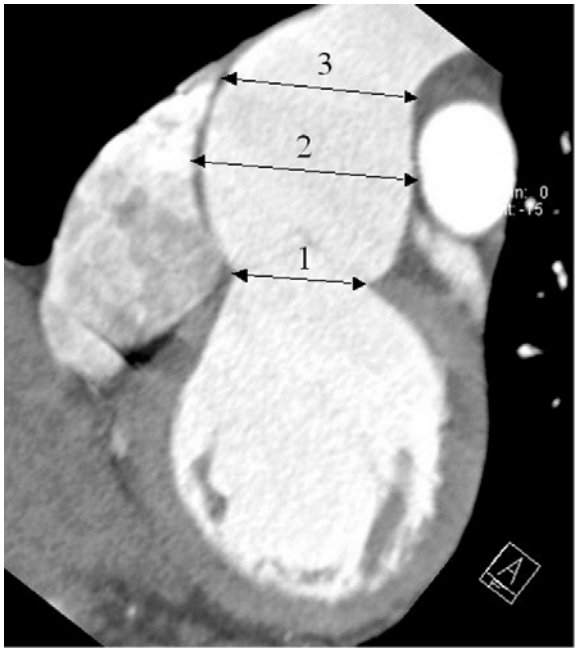

Fig. 1. A MPR image of the ascending aorta at mid-diastole shows left ventricular enlargement and annuloaortic ectasia. Diameters at the level of aortic annulus (1), sinuses of Valsalva (2), and sinotubular ridge (3) are 37.4 mm, 58.7 mm, and 51 mm, respectively.

Discussion

MS is an autosomal dominant connective tissue disease involving elastic tissue, which results in ocular, skeletal, pulmonary, and cardiovascular anomalies. The involvement of cardiovascular system occurs in more than a half of affected patients. Characteristic cardiovascular features include aortic valve incompetence and regurgitation dilatation associated with the dilatation of the aortic annulus and the progressive dilatation of the proximal ascending aorta leading to aortic dissection or rupture. Mitral regurgitation is the most common cardiac abnormality in children [1-3].

Conventional aortography is an invasive procedure and also less useful to evaluate cardiovascular complications in MS. Transthoracic echocardiography (TTE) is a primary imaging modality used for the noninvasive assessment of cardiovascular complications. In the situation which TTE is unsatisfactory, it is better to performed the transesophageal echocardiography (TEE) in order to detect aortic dissection accurately, to

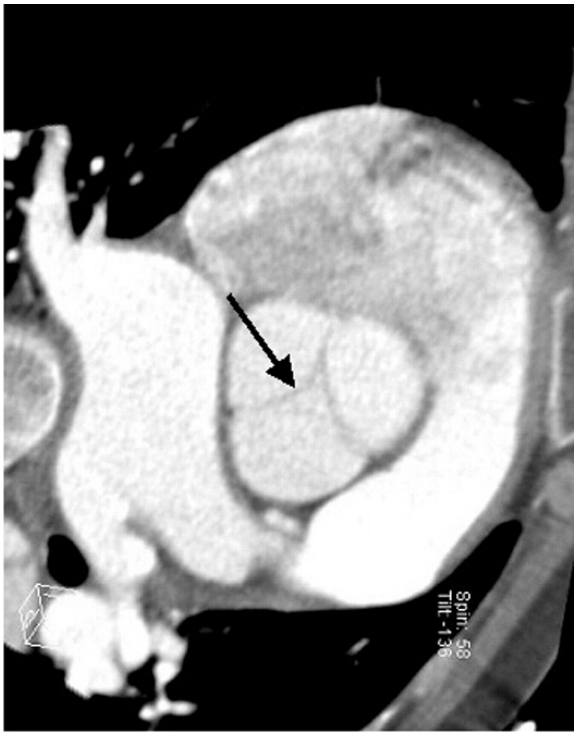

Fig. 2. A MPR image of the aortic valve at mid-diastole demonstrates a triangular coaptation defect (arrow).

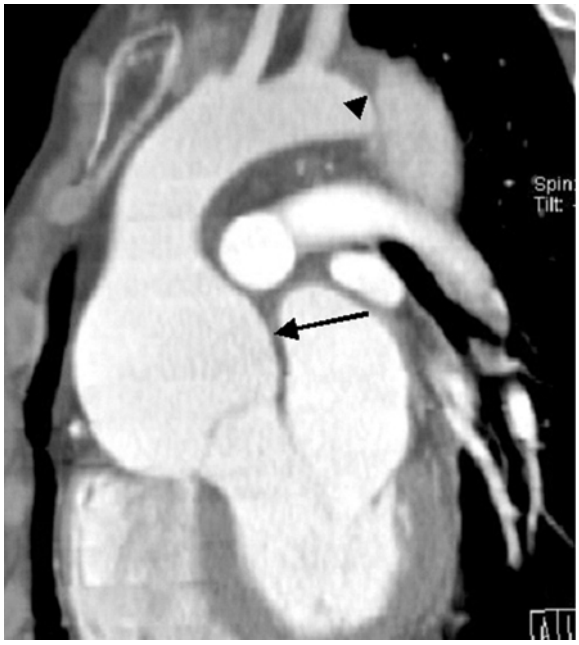

Fig. 3. A MPR image of the aortic arch at mid-diastole shows annuloaortic ectasia (arrow), intimal flap beginning just distal to the origin of the left subclavian artery and focally thrombosed false lumen (arrowhead).

evaluate aortic root complications, and to assess aortic dimensions. However, echocardiography is operator-dependent and less ideal for showing the full extent of aorta. Magnetic resonance imaging (MRI) is commonly used for aortic screening such as in annuloaortic ectasia in MS patients and their relatives. It is also useful for detecting cardiovascular complications with a wide field-of-view, multiplanar imaging and an ability to quantify the degree of valvular regurgitation[3-5]. However, this modality has several limitations including high cost, small number of scanners available, small number of trained radiologists who can operate and interpret the images, longer time for image acquisition, strong magnetic field to prohibit the presence of metal objects, and limit use in acutely ill patients.

CT angiography is the investigation of choice for the patients with acute aortic syndrome. It is more sensitive than catheter angiography for diagnosing aortic dissection, and comparable to MRI and TEE. With conventional CT angiography, however, the evaluation of torn site and of the involvement of dissection flaps to the aortic sinus and coronary arteries, is undesirable due to pulsating motion artifacts[5]. Recently introduced 16-slice MDCT allows faster acquisition, and higher spatial and temporal resolutions. By using ECG-gated MDCT technique, it is possible to suppress the cardiac pulsation artifact leading to improve the quality of cardiothoracic imaging[3, 4]. In an emergency situation, MDCT angiography may be the first-line imaging modality usually carrying out before TEE and MRI in case suspecting cardiovascular complications in patients with MS. This is due to

the widespread availability of CT scanners and less operator-dependent despite radiation exposure. The spatial resolution of MDCT is superior to that of MRI, allowing the investigation of acute visceral complications associated with high mortality of aortic dissection[4]. Furthermore, the multi-phase reconstruction of cardiovascular imaging data during systole and diastole with high quality 2D and 3D reformations may enable to assess the annuloaortic ectasia, the aortic dissection with coronary artery involvement and myocardial infarction, and the coaptation of aortic and mitral valves in MS.

There is no specific treatment for MS. The selection of surgical treatment is dependent on pathology of the aortic root, the severity of aortic regurgitation, and left ventricular function. The replacement of the aortic valve and as much of the ascending aorta as possible is the standard operation[1]. Using MDCT, the preoperative measurement of diameter at the level of aortic annulus, sinuses of Valsalva, sinotubular ridge and tubular portion of the ascending aorta may facilitate the planning of optimal surgical management of annuloaortic ectasis in MS.

Summary

ECG-gated MDCT angiography may be a powerful diagnostic tool for the evaluation of cardiovascular complications and for the planning of optimal surgical treatment in Marfan syndrome with annuloaortic ectasia and aortic regurgitation.

References

1. Treasure T. Cardiovascular surgery for Marfan syndrome. *Heart* 2000;**84**:674-8.
2. Lehmann ED. Cardiovascular screening in Marfan's syndrome. *Lancet* 1995;**345**:1501.
3. Miller SW. *Cardiac Imaging: the Requisites*. 2nd ed. Philadelphia: Elsevier Mosby; 2005, p.380-2.
4. Willoteaux S, Lions C, Gaxotte V, Negaiwi Z, Beregi JP. Imaging of aortic dissection by helical computed tomography. *Eur Radiol* 2004;**14**:1999-2008.
5. Hartnell GG. Imaging of aortic aneurysm and dissection: CT and MRI. *Thoracic Imaging* 2001 ;**16**:35-46.