Abdominal Aortic Dissection with Acute Mesenteric Ischemia in a Patient with Marfan Syndrome

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Marfan syndrome is an autosomal dominant inherited disorder of connective tissue, with various complications manifested primarily in the cardiovascular system. It potentially leads to aortic dissection and rupture, these being the major causes of death. We report a patient who complained of acute abdominal pain, which presented as acute mesenteric ischemia combined with abdominal aortic dissection. Echocardiography showed enlargement of the aortic root and mitral valve prolapse. Abdominal computed tomography scan revealed acute mesenteric ischemia due to abdominal aortic dissection. Finally, the patient underwent surgery of aortic root replacement and had a successful outcome. Therefore, we suggest that for optimal risk assessment and monitoring of patients with Marfan syndrome, both aortic stiffness and the diameter of the superior mesenteric vein compared with that of the superior mesenteric artery are useful screening methods to detect acute mesenteric ischemia secondary to abdominal aortic dissection. Early diagnosis and early treatment can decrease the high mortality rate of patients with Marfan syndrome. [*J Chin Med Assoc* 2006;69(7):326–329]

Key Words: abdominal aortic dissection, Marfan syndrome, mesenteric ischemia

Introduction

Marfan syndrome is an autosomal dominant inherited disorder of connective tissue, with various complications manifested primarily in the skeletal, ocular, and cardiovascular systems.[1] The estimated prevalence is 2–3 per 10,000 population.[2] Prognosis is mainly determined by progressive dilation of the aortic root, potentially leading to type A aortic dissection and rupture, these being the major causes of death.[3–5]

There are no clear guidelines for intervention of descending aortic complications in patients with Marfan syndrome.[6–8] The risk of aortic dissection rises with increasing aortic diameter, but it may also occur in non-dilated aortas.[7,9] Thus, aortic stiffness as a potential predictor of aortic dissection and aortic dilatation has been investigated in patients with Marfan syndrome.[6,7]

The in-hospital mortality rate of patients with acute aortic dissection has been reported to be 15–25%;[10–13] however, treatment of patients with the complication of acute mesenteric ischemia secondary to aortic dissection is still challenging due to the difficulty of early diagnosis.[14,15] Acute mesenteric ischemia has been reported to be a risk factor of early death, and the overall mortality rate is >60%.[14]

We report a case of Marfan syndrome with acute mesenteric ischemia secondary to abdominal aortic dissection.

Case Report

A 34-year-old woman visited our emergency room due to acute cramping abdominal pain. She had a history

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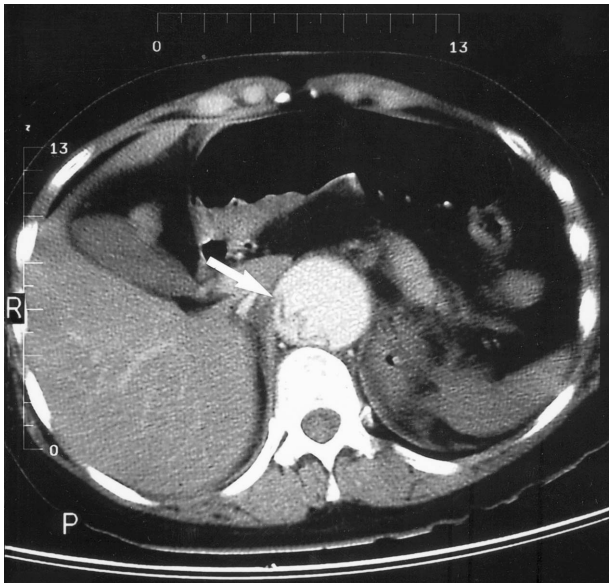

Figure 1. Abdominal computed tomography scan revealed abdominal aortic dissection with no sign of rupture.

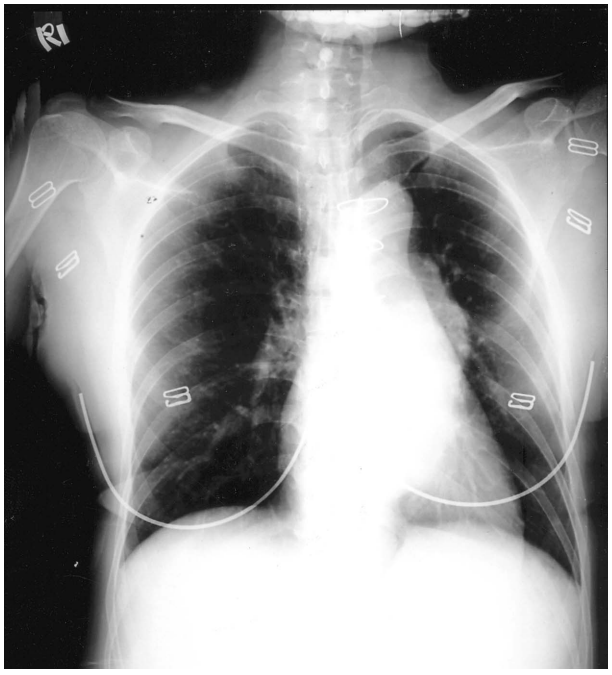

Figure 2. Chest X-ray of the patient.

of aortic aneurysm with Marfan syndrome and had received aortic root replacement surgery 2 years before this visit. Abdominal computed tomography (CT) scan revealed abdominal aortic dissection with no sign of rupture (Figure 1), and she had received coumadin therapy for 2 years.

On arrival, the patient's blood pressure was 120/54 mmHg, body temperature was 36.5°C, pulse rate was 112/minute, and respiratory rate was 24/minute. Physical examination showed an acutely ill-looking patient. The conjunctiva was pale with superior lens dislocation (ectopia lentis). There was maximal impulse lateral to the mid-clavicular line with visible carotid pulsations. Heart sounds of the murmur of mitral valve prolapse and the sound of aortic regurgitation murmur could be heard loudest along the left sternal border at the 3rd and 4th intercostal spaces and at aortic and apex locations. The blowing and faint murmur began immediately after the aortic sound and ended before the first sound. Anterior chest deformity (pectus excavatum) required surgery (Figure 2). Joint hypermobility with spondylolisthesis were also noted. The abdomen was distended, with severe epigastric tenderness but without muscle guarding or rebound tenderness. Murphy's sign and Courvoisier's sign were both negative. There was no palpable mass. The bowel sounds were hyperactive with bruit. Digital rectal examination was negative.

The hemogram showed a white blood cell count of 11,000/μL (4,000–10,000/μL), hemoglobin level of 8.5 g/dL (14.0–18.0 g/dL), and platelet count of 100,000/μL (130–400 × 10^3/μL). Serum biochemistry showed alanine aminotransferase (ALT) of 27 IU/L (0–40 IU/L), aspartate aminotransferase (AST) of 42 IU/L (5–34 IU/L), creatine phosphokinase (CPK) of 233.4 IU/L (25–190 IU/L), and MB isoform of 43.8 IU/L (5–34 IU/L). The prothrombin time was 21.4 seconds, with an INR of 1.54. D-dimers were positive, and troponin I was negative. Other tests were unremarkable.

The electrocardiogram (ECG) showed severe left ventricular hypertrophy. In the precordial leads, we found that S waves in V1 and V2 and an R wave in V5 and V6 were >25 mm. Chest radiographs showed cardiomegaly with left ventricular prominence. Echocardiography demonstrated enlargement of the aortic root and mitral valve prolapse. Doppler scan showed severe mitral valve regurgitation. According to the De Bakey classification, this patient had type III dissection involving the entire aorta,[16] which originated in the descending aorta and extended distally down the aorta and retrograde into the aortic arch and ascending aorta.

CT scan was performed to detect the acute aortic dissection from the levels of aortic arch branches to the common iliac arteries (Figure 3). The diameter of the superior mesenteric vein compared with that of the superior mesenteric artery reflected that this patient suffered from mesenteric ischemia secondary to acute aortic dissection.

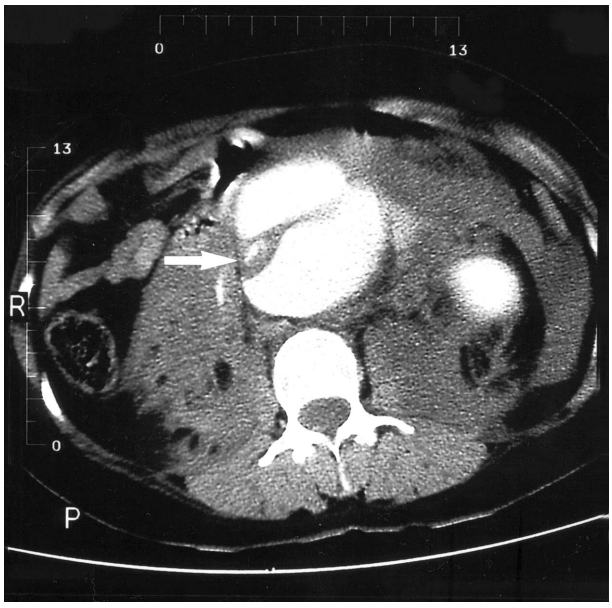

Figure 3. Computed tomography scan showed the acute abdominal aortic dissection from the level of the common iliac arteries.

After conservative treatment, the patient felt better, and surgical intervention was suggested. On the second day, she was transferred to the surgical section, and her condition showed progressive improvement as she underwent surgery for acute mesenteric ischemia. Finally, she was discharged from our hospital.

Discussion

In our patient, the aortic root and ascending aorta had previously been replaced with a tubular Teflon graft containing a mechanical valvar prosthesis before admission; the coronary arteries were reimplanted in the graft. This has now become the standard surgical procedure for patients with aneurysm of the aortic root in the setting of Marfan syndrome.[4] However, after replacement of the aortic root, patients should have continued attention because of complications that may eventually develop in the descending aorta beyond the aortic arch,[13] because replacement of the aortic root is usually associated with a considerably higher risk of redissection and recurrent aneurysm, or mesenteric ischemia secondary to acute aortic dissection.[16] Thus, once a patient presents with dissection, either acute or chronic, at the time of the first surgery, there is a significant necessity to predict subsequent need for reoperation.[17–19]

The majority of patients with Marfan syndrome die prematurely due to rupture of the aorta, often by the third decade of life. Whenever possible, patients with the syndrome should be under the care of professionals with specific training and experience.[11] Ideally, this should be performed in a multidisciplinary setting. All patients with Marfan syndrome are advised to take β-adrenergic blocking agents,[18] and to remain on this therapy unless intolerable side effects preclude their use. This is especially true, usually in association with other blood pressure-lowering agents, if dissection has occurred.[13] During follow-up, the aortic root and the entire aorta should be regularly evaluated with echocardiography, CT, and/or abdominal ultrasound examinations, especially if a dissection remains and its stability is being monitored.[11,13,20] Patients with mitral valve prolapse, and more than moderate mitral regurgitation, should also be followed with yearly echocardiographic examinations.[6,17] Prophylaxis against endocarditis is recommended for 6 months following the replacement of the aortic root,[19] or for life if any residual gradient or lesions persist, or in the presence of prosthetic valvar or mitral regurgitation.[4]

Life expectancy has increased significantly in patients with Marfan syndrome, due to advances in medical and surgical treatment.[13] In an unknown number of patients, nonetheless, diagnosis is established only after development of an aortic aneurysm, dissection, or even after death.[11] Early diagnosis, therefore, should be improved by increased awareness in the general population and further education of physicians. Because aortic complications are not always predictable exclusively on the basis of dimensions of the aortic root, further research should be focused on the functional properties of the aorta, and on molecular genetics to identify the patients at greatest risk for acute aortic dissection.[1,5,13]

Although the diagnosis of acute mesenteric ischemia secondary to aortic dissection in patients who complain of abdominal pain is not so difficult,[14] the treatment of this kind of patient is still challenging due to the difficulty of early diagnosis in painless abdominal aortic dissection, which accounts for 5–10% of aortic dissections.[14] Further, its clinical presentation consists mainly of neurologic symptoms.[3] Acute mesenteric ischemia has been reported to be a high risk factor of early death, and the overall mortality rate is >60%.[14,15] Our patient underwent surgery after clinical diagnosis, and recovered well after surgery; due to early diagnosis, finally, she had a successful outcome.

Marfan syndrome is a disease mainly located in the ascending and descending aortas, but rarely in the abdominal aorta. The potential relationship between the presence of abdominal aortic dissection and Marfan syndrome has been reported.[6] Whether abdominal

aortic dissection could be related to the previous surgery for Marfan syndrome in this particular patient remains unknown. Recently, many methods have been suggested for careful surveillance of aortic aneurysm formation and timely surgical intervention for patients with Marfan syndrome,[6,10,20] to prevent the eventual occurrence of aortic dissection, and to reduce the risk of acute mesenteric ischemia secondary to aortic dissection, which is uncommon but a potentially deadly disorder. For optimal risk assessment and monitoring of patients with Marfan syndrome, both the severity of aortic stiffness and the diameter should be assessed at least annually.[6] The diameter of the superior mesenteric vein compared with that of the superior mesenteric artery and lactate concentration are also useful screening methods to detect mesenteric ischemia secondary to acute abdominal aortic dissection.[14] Further, patients with acute aortic dissection show significant systemic inflammatory reactions.[16] Recently, D-dimers have been found to be highly elevated in acute aortic dissection; thus, the measurement of D-dimers may be a valuable addition to the current diagnostic work-up of patients with acute abdominal aortic dissection and Marfan syndrome.[21] However, more investigations of these kinds of patients are still required.

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