A young male patient with Marfan syndrome suffered from acute type B aortic dissection with visceral organ malperfusion. The thoracic stent grafting was urgently performed with a successful outcome. This study reports a potential endovascular approach to treat complicated acute type B aortic dissection in a Marfan syndrome patient.

Keywords: Aortic dissection, endovascular stent graft, malperfusion, Marfan syndrome

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ABSTRACT

A 22 year old man presented with severe epigastric pain 11 days ago experienced during sexual activity. He had visited many hospitals and received medication for peptic ulcer, but his pain still persisted. He had no history of abdominal injury, no smoking and no relevant family history.

Physical examination showed a tall, thin man with 190 cm in height, 192 cm in arm span. BP 130/90 mmHg., pulse 90 beats/minute. He had no cardiac murmur, no abnormal visual acuity, mild abdominal tenderness, no guarding and no rigidity. All peripheral pulses were full. His long fingers can surround his wrist. Blood chemistry did not show any abnormality.

Chest X-ray revealed widening of the mediastinum and CT angiography of his thoracic and abdomen demonstrated a descending aortic dissection extended to the aortic bifurcation. Aortic dissection is characterized by an intimal flap which divides the aortic lumen into true and false lumen (Fig 1-3). The aortic sinus was 4.6 cm in maximal diameter. Both aortic lumens were persistent with large false channel and compressed true channel. The celiac artery, superior mesenteric artery and right renal artery were taken off from the true lumen except the left renal artery came from the false lumen. No pleural effusion was demonstrated.

His clinical features match with Marfan syndrome appearance. The patient was diagnosed acute aortic dissection Stanford type B with visceral malperfusion. He was decided to operate because of the complicated acute type B dissection.

Intraoperative transesophageal echocardiography (TEE) was performed and showed the intimal tears at many levels of the descending and abdominal aorta with the major tear at the proximal descending aorta. The true lumen was compressed. An endovascular stent grafting was planned to get rid the proximal communication between true and false lumen and enhance the blood flow to the true lumen. The procedure was accomplished under general anesthesia and the TEE guide. The stent graft was deployed in the true lumen at the proximal descending aorta to close the major tear and to intentionally cover the left subclavian artery. The postoperative angiography revealed an enlarged size of the true lumen with decreased flow in the false lumen. His abdominal symptom gradually improved without left arm claudication and vertebral steal syndrome. At 3 months postoperatively, computerized tomographic angiography (CTA) revealed the patent stent graft occupying in the true lumen and disappearance of the false lumen in the descending thoracic aorta (Fig 4,5). The dissection persisted below the stent graft level and both lumens were perfused.

DISCUSSION

The acute aortic dissection is an emergent cardiovascular condition. The symptom of severe sudden onset of chest or abdominal pain carry a high index of suspicion. The other symptoms could be from complications of vascular compromise for example syncope, stroke, myocardial infarction, spinal cord ischemia, visceral or limb ischemia. The majority of the patients have a history of hypertension. The connective tissue
diseases such as Marfan syndrome or Ehler Danlos syndrome are the risk of aortic dissection. The patient had fulfilled clinical diagnosis for Marfan syndrome by 2 major Ghent criteria (aortic dilatation and dural ectasia). The standard clinical Ghent criteria are established (Table 1). The definite diagnosis of Marfan syndrome can be made when at least 2 major criteria are presented with an involvement of the third system (either major or minor). Dural ectasia is characterized by dilatation of dural sac or nerve root sleeves of the spinal canal (Fig 6), which has been observed in 56-65% of patients with Marfan syndrome.

The physical signs may reveal pulse deficit, murmur of the aortic valve regurgitation and signs of organ ischemia. Chest X-ray finding of a widening mediastinum or abnormal aortic contour is seen in up to 75% of patients. Associated clues may include left pleural or pericardial effusion. A 12 lead electrocardiogram (ECG) is useful prognostic information and may differentiate acute coronary syndrome from acute aortic dissection. This issue is very important because treatment of acute coronary syndrome by antiplatelets and thrombolysis may have catastrophic consequences in the presence of aortic dissection.

The CTA is most often selected as the initial imaging study due to its convenience, high sensitivity (>95%) and it offers great visualization of the vessel involvement, pericardial and pleural effusion. It demonstrates the extension of dissection, how collapsed the true lumen is and the origin of involvement of visceral arteries (celiac, superior, inferior mesenteric and renal arteries). Transthoracic echocardiography (TTE) may be best reserved to screen for dissection in patients presenting with unexplained shock or syncope. Transesophageal echocardiography (TEE) offers excellent views of the aorta from the root to the descending aorta with high sensitivity (99%) and specificity (89%). Aortography has been considered as a gold standard in diagnosing aortic dissection, but is currently replaced by non invasive modalities such as CTA and echocardiography. MRI offers superior imaging resolution, but is not used as an initial imaging study due to impractical issues in an unstable patient.

It is essential to classify in acute aortic dissection (within 14 days) whether ascending aorta is involved (Stanford type A) or not (Stanford type B) because the emergent surgery is needed in acute ascending dissection. For the patient with acute aortic dissection without ascending aorta involvement (Stanford type B), the principle of management is anti-impulse therapy especially beta-blocker, since the surgical mortality exceeds that for medical management by factor of 3. The overall in-hospital mortality was 29% in type B aortic dissection who required surgery. However, surgery has an important role in acute type B dissection with complications such as rupture, organ malperfusion.
**TABLE 1. Diagnostic Ghent criteria for Marfan syndrome.**

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<th>Organ system</th>
<th>Major criteria</th>
<th>Minor criteria</th>
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| Skeletal     | Presence of at least four of the following components:  
  - Pectus carinatum, or pectus excavatum requiring surgery  
  - Reduced upper-to-lower segment ratio for age (<0.85 for older children or adults) or arm span-to-height ratio (>1.05)  
  - Wrist and thumb signs  
  - Scoliosis of >20° or spondylolisthesis  
  - Reduced extension at the elbow (<170°)  
  - Medial rotation of the medial malleolus causing pes planus  
  - Protrusio acetabulae (abnormally deep acetabulum with accelerated erosion) of any degree (ascertained on radiographs) | Two major components or one major component and at least two of the following:  
  - Pectus excavatum of moderate severity  
  - Joint hypermobility  
  - Highly arched palate with tooth crowding  
  - Facial appearance (dolichocephaly, malar hypoplasia, enophthalmos, retrognathia, down-slanting palpebral fissures) |
| Ocular       | Ectopia lentis | At least two of the following:  
  - Abnormally flat cornea (as measured by keratometry)  
  - Increased axial length of the globe (as measured by ultrasound)  
  - Hypoplastic iris or hypoplastic ciliary muscle causing decreased pupillary miosis |
| Cardiovascular | At least one of the following:  
  - Dilatation of the ascending aorta involving the sinuses of Valsalva  
  - Dissection of the ascending aorta | At least one of the following:  
  - Mitral valve prolapse with or without mitral regurgitation  
  - Dilatation of the main pulmonary artery, in the absence of obvious cause, before the age of 40 years  
  - Calcification of the mitral annulus before the age of 40 years  
  - Dilatation or dissection of the descending thoracic or abdominal aorta before the age of 50 years |
| Dural        | Lumbosacral dural ectasia (ascertained by CT or MRI) | |
| Pulmonary    | | At least one of the following:  
  - Spontaneous pneumothorax  
  - Apical blebs (ascertained by chest radiography) |
| Cutaneous    | | At least one of the following:  
  - Striae without obvious cause  
  - Recurrent or incisional hernia |
| Familial/Genetic | At least one of the following:  
  - Having a parent, child, or sib who meets these diagnostic criteria independently  
  - Presence of a mutation in FBN1 known to cause Marfan syndrome  
  - Presence of a haplotype around FBN1, inherited by descent, known to be associated with Marfan syndrome in the family (ascertained by linkage analysis) | |

and uncontrolled pain or hypertension. The algorithm for the treatment of acute aortic dissection is shown in Fig 7.\textsuperscript{6-8} This patient had a sudden onset of severe epigastric pain in which imaging study supported the diagnosis of acute aortic dissection type B and visceral malperfusion, so an operation is indicated.

The conventional surgery for organ malperfusion in aortic dissection is the fenestration by cutting the intimal flap to provide blood flow into the true and false lumens. The procedure is hazardous due to fragile tissue in an acute period. The new modality of treatment is endovascular surgery which is less invasive, with less morbidity and mortality by putting the stent graft into the true lumen and landing to cover the large proximal communication between the true and false lumen.\textsuperscript{9,10} The objective is to restore blood flow to the true lumen and obliterate the false lumen. The proximal landing zone is one of the essential issues in the stent grafting procedure. At least 2 cm. neck length is needed for the proximal part of the stent graft to fix in the normal aorta. In the descending aortic dissection, a proximal tear usually starts just distal to the left subclavian artery origin. The artery is frequently covered by a proximal part of the stent graft intentionally during the procedure. The stroke from the left vertebral artery occlusion is a concern especially in the patient who has a dominant left vertebral artery or incomplete circle of Willis. However, pre-operative left carotid-left subclavian artery bypass is still a controversial issue. Our strategy is performing left subclavian artery bypass in all elective cases who have a dominant left vertebral artery, but not in urgent or emergent situations unless the patient developed ischemic symptoms post-operatively.

We report the successful treatment of a complicated acute type B dissection by endovascular stent graft. The patient still needs to have close observation and to follow a CTA protocol at 3, 6, 12 months and yearly. The long term outcome of stent grafting is important to be monitored, especially in the connective tissue disease patient who has a tendency of progressive aortic dilatation and might have a chance of proximal endoleak or retrograde aortic dissection in the future.

REFERENCES