Paraplegia as a symptom of failure after endovascular therapy of type B aortic dissection in Marfan syndrome

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This report describes successful treatment of an unusual case of concomitant paraplegia and type 1 endoleak during the early postoperative course of endovascular therapy of type B dissection in a patient with Marfan syndrome. (J Vasc Surg 2009;49:478-82.)

Spinal cord ischemia is now a well-known complication after endovascular therapy of thoracic aortic aneurysms and dissections.[1] This report describes an unusual case of concomitant paraplegia and type 1 endoleak during the early postoperative course of endovascular therapy of type B dissection in a patient with Marfan syndrome.

CASE REPORT

A 29-year-old man from Africa with Marfan syndrome was referred for a type B aortic dissection. He met Ghent criteria and his brother had died of aortic complications. He had no past history of acute symptoms. Transthoracic cardiac echography showed a normal left ventricular ejection fraction. A CT scan (Fig 1) showed an aneurysm of the aortic root and of the aortic arch measuring 50 mm in diameter, associated with an aortic dissection starting 10 mm after the origin of the left subclavian artery. A 20 mm tear was visible at that level, which widely communicated with the false lumen. The consequence of this wide communication was a marked narrowing of the true lumen. The dissection extended down to the level of the aortic bifurcation. Both celiac axis and right renal artery arose from the false lumen, whereas the superior mesenteric artery (SMA) and left renal artery arose from the true lumen. Maximum diameter of the descending thoracic aorta was 70 mm, and maximum diameter of the infrarenal aorta was 43 mm. Multiplanar reconstruction (MPR) of CT scan slices (Fig 2) precisely located the origin of the Adamkievicz artery at the level of the left T9 artery, from the true lumen.

Considering the extent of the aortic disease, we planned a staged surgical repair using the elephant trunk technique.[2] The first stage of the repair was performed under cardiopulmonary bypass with deep hypothermic circulatory arrest. We performed replacement of the ascending aorta by a mechanical valve conduit (Gelweave Valsalva, Vascutek Terumo, Renfrewshire, Scotland) using the modified Bentall technique, total aortic arch replacement with separate re-implantation of the supra-aortic trunks, and construction of a distal elephant trunk using a 24 mm Dacron tube graft. Due to angulation of the aortic isthmus and to the marked narrowing of the true lumen of the dissection, the elephant trunk had to be inserted into the false lumen.

The patient was weaned from ventilatory supply on the first postoperative day. At day 9, he experienced brutal chest pain, with concomitant dyspnea, and hypotension. Emergency CT scan (Fig 3) showed a 15 mm enlargement of the descending thoracic aorta and a left pleural effusion: emergency endovascular intervention was thus considered, because of the short interval from the first intervention, and of the precarious respiratory status of the patient.

Cardiac arrest upon anesthetic induction resolved rapidly after resuscitation maneuvers. Endovascular therapy was performed. The elephant trunk graft was catheterized via a right brachial approach. As there was a discrepancy between the size of the elephant trunk (24 mm) and the size of the distal aorta (false lumen 34 × 22 mm, overall lumen 34 × 33 mm), three Valiant (Medtronic, Minneapolis, Minn) stent-grafts (28 × 100 mm, 34 × 200 mm, 37 × 170 mm) were inserted via a left femoral approach, without evidence of endoleak on control angiogram. The distal end of the endograft was located in the false lumen at the thoracoabdominal junction, 2 cm above the celiac axis. An angiogram showed proper filling of the visceral arteries and did not show the true lumen at the level of the thoracic aorta. Intraoperative transesophageal echography (TEE) confirmed the absence of endoleak. No early postoperative paraplegia was noted, and spinal cord fluid drainage was left in place for 2 days, although it did not show evidence of any significant pressure elevation. The patient was weaned from ventilatory support after 48 hours, after a control TEE showed the absence of endoleak.

At day 4 of this second intervention, the patient suddenly presented acute paraplegia, followed a few minutes later by recurring chest pain, dyspnea, and hypotension. Emergency CT scan (Fig 4) showed a type 1 endoleak at the distal end of the endograft and of the celiac aorta, with a 10 mm increase of the size of the false lumen. Three-D reconstruction did not show evidence of any patent Adamkievicz artery at the level of T9 artery.

Lumbar spinal cord fluid drainage was started again, initial pressure being around 20 mm Hg. Emergency aortic replacement was performed via a left posterior thoracotomy through the sixth

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Competition of interest: none.

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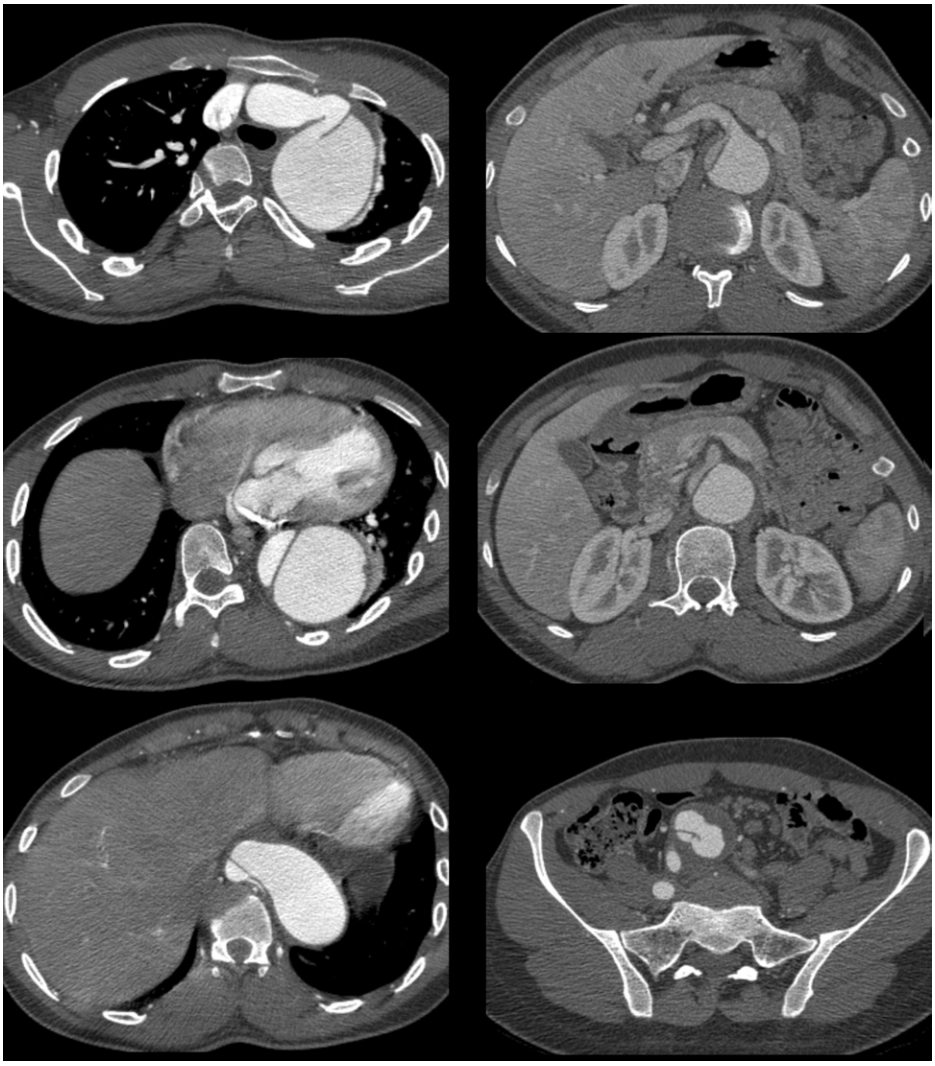

Fig 1. Preoperative CT scan shows - **Left (top to bottom):** aortic dissection starting 10 mm after the origin of the left subclavian artery, a 20 mm tear which widely communicates with the false lumen, .aneurysm of the aortic root and of the aortic arch, compression of the true lumen by an enlarged false lumen; **Right (top to bottom):** origin of celiac axis from the false lumen, superior mesenteric artery from the true lumen, extension of the dissection to the aortic bifurcation.

intercostal space under arterio-venous femoro-femoral cardiopulmonary bypass: the three stent-grafts were removed, a 30 mm Dacron graft was anastomosed to the true lumen at the level of T12 and to the posterior aortic wall with T8 to T12 arteries; aortic wall with T4 to T6 arteries was re-implanted laterally in the 24 mm elephant trunk graft, and end-to-end anastomosis of both grafts were performed. We performed complementary fenestration of the abdominal aorta from the anastomosis to the level of the renal arteries, as two out of four visceral arteries were arising from the false lumen, as documented by the preoperative work-up.

The patient was weaned from ventilatory supply after 48 hours, without evidence of residual paraplegia, and spinal cord fluid drainage was removed. Respiratory function impaired again due to right upper lobe infection. The patient was placed again under respiratory support for 8 days. As the infection resolved, he was permanently weaned from ventilatory supply on postoperative day 14. Control CT scan (Fig 5) showed patency of the re-implanted intercostal arteries, and of the Adamkievicz artery.

The patient was discharged 8 weeks after the first intervention, without evidence of recurring neurological symptoms, and without respiratory sequelae.

DISCUSSION

Paraplegia is a well-known complication of both surgical and endovascular therapy of the thoracic aorta.[1] Reimplantation of intercostal arteries and spinal cord

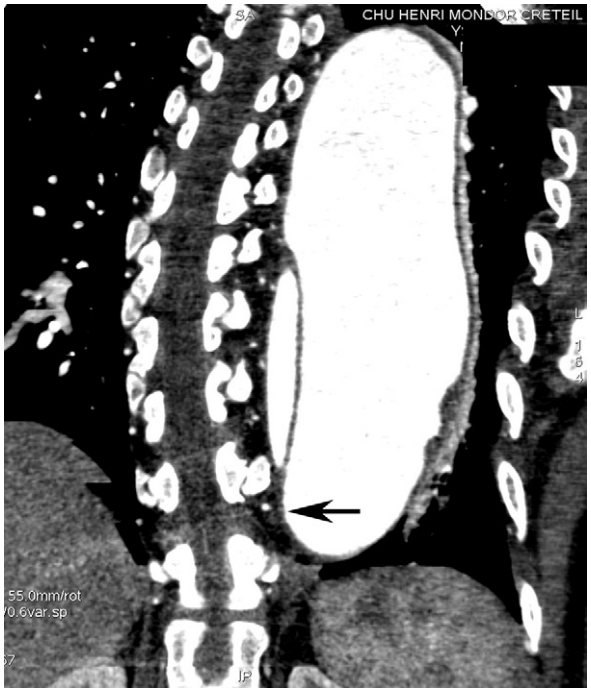

Fig 2. Preoperative identification by multiplanar reconstruction (MPR) of CT scans of Adamkievicz artery arising (*arrow*) from left T9 artery.

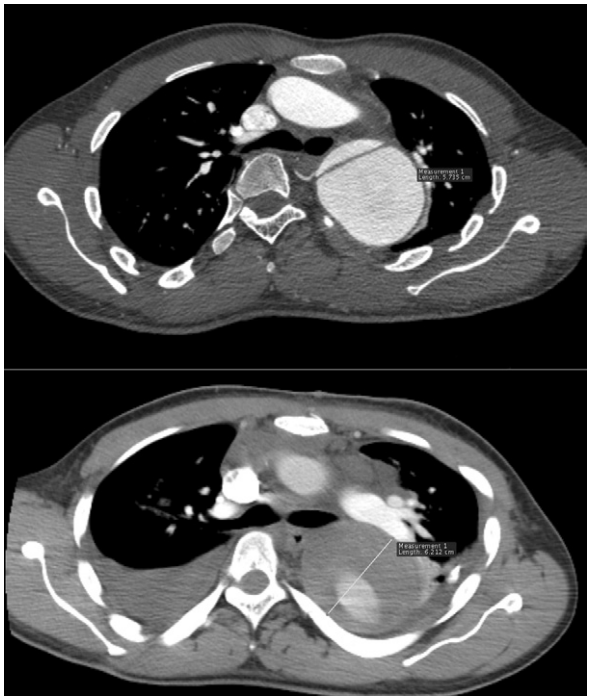

Fig 3. Comparison of CT scan slices at the level of the tracheal bifurcation show enlargement of the descending aorta and pleural effusion **(below)** as compared to preoperative work-up **(above)**. The elephant trunk graft can be seen in the false lumen.

fluid drainage are deemed the best options to prevent this complication after conventional surgery. Preservation of the patency of the left subclavian artery and avoidance of extensive coverage of the thoracoabdominal aorta are the only recommendations that can be made after endovascular therapy. Whether occlusion of the Adamkievicz artery by the stent-graft leads to a higher rate of paraplegia is still controversial.[3]

The case history of this patient is unusual. Our initial attempt – ie, endovascular – to treat the aortic dissection, aimed at the exclusion of both the false and true lumen by the endograft on the entire height of the descending aorta down to T12 level.

There was no endoleak on intraoperative control angiogram and TEE, as well as on repeated TEE at day 2. However, as the patient was asymptomatic, we were reluctant to document the status of intercostal arteries by early CT scan. Thus, we cannot exclude the persistence of a retrograde perfusion of the true lumen from a more distal entry site, maintaining patency of T9-T12 arteries.

The underlying mechanism of acute spinal cord ischemia occurring on day 4 after endovascular therapy is not clear. The rapid onset of symptoms, the elevated spinal fluid pressure, and the evidence of a distal type 1 endoleak on CT scan are consistent with the diagnosis of a sudden endoleak, increasing the pressure within the false lumen. The consequence was a compression of the true lumen and an occlusion of intercostal and of Adamkievicz arteries. Thus, the Adamkievicz artery was visible on all CT scans but the one emergently performed after the onset of paraplegia. Furthermore, patency of T9 to T12 arteries and full recovery of neurological symptoms after surgical repair and re-implantation of intercostal arteries are also consistent with our hypothesis. To our knowledge, there have been no previous reports of spinal cord complications induced by the acute onset of an endoleak.

Marfan syndrome has been identified as an independent factor of surgical conversion after endovascular therapy of the thoracic aorta.[4] Although debatable, our first choice to treat the threat of impending rupture was an emergency endovascular procedure as the patient's condition was deemed precarious. Endovascular therapy of aortic dissection in patients with Marfan syndrome has been reported in patients with rapid evolution and poor general condition.[5]

The onset on day 4 of paraplegia and of a type 1 endoleak was to us a clear indication for conventional surgical replacement of the thoracic aorta, although successful treatment of delayed paraplegia by translumbar cerebral fluid drainage alone has been reported.[6] Although compression of the true lumen is the most probable hypothesis, we preferred to provide direct vascular supply to intercostal arteries, together with cerebral fluid drainage and temporary intraoperative distal aortic perfusion as recommended by Safi,[7] in order to provide the best chance of recovery to the patient.

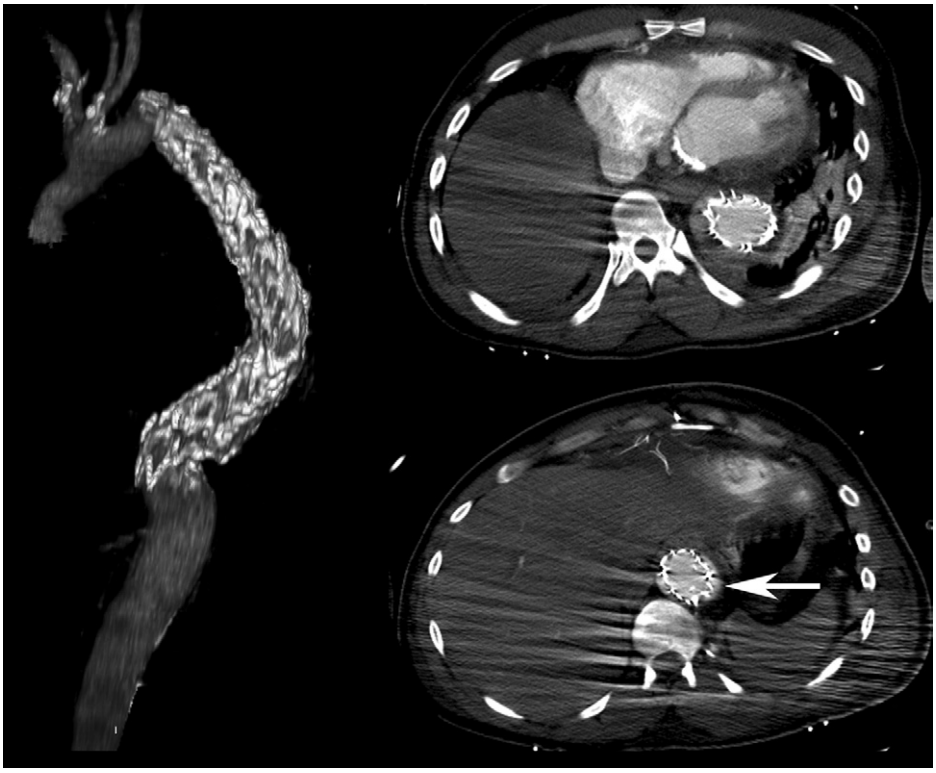

Fig 4. CT scan at day 4 after endovascular repair - **Left:** 3-D reconstruction shows stent-graft in the false lumen from the aortic isthmus to the level of the thoracoabdominal junction; **Right:** contrast (*arrow*) around the stent-graft **(below)** identifies a distal type 1 endoleak, filling the false lumen **(above)**, thus compressing the true lumen.

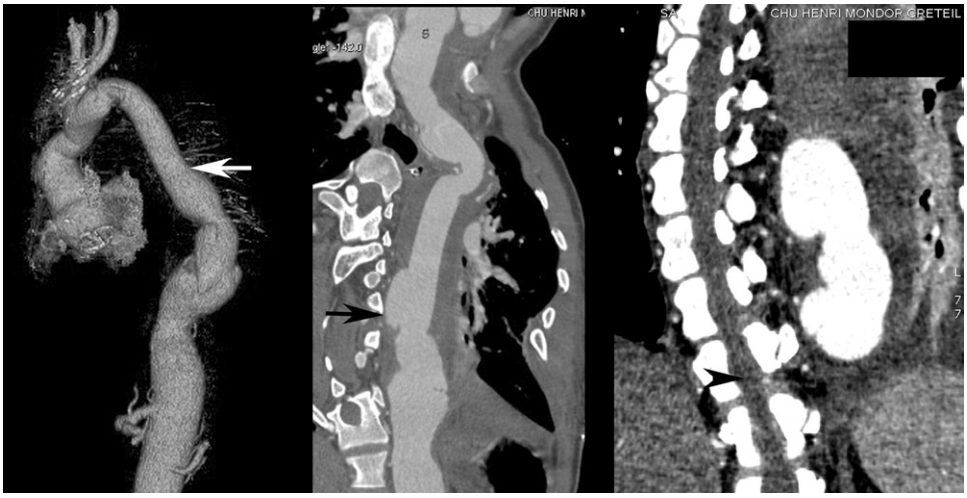

Fig 5. Control CT scan after surgical repair: **Left:** 3-D reconstruction shows graft replacement of the descending thoracic aorta by elephant trunk graft anastomosed (*arrow*) with a larger graft preserving vascularization of T9-T12 intercostal arteries; **Center:** centerline MPR reconstruction shows enlargement (*arrow*) of the graft at the level of the intercostal arteries; **Right:** MPR identifies a patent Adamkievicz artery at T9 level (*black arrowhead*).

CONCLUSION

Our study confirms that the mechanisms of spinal cord ischemia after endovascular therapy of the thoracic aorta are various and complex. Paraplegia was not a direct consequence of exclusion of intercostal arteries, but was delayed and contemporary with the onset of a distal type 1 endoleak. As endovascular therapy in patients with Marfan syndrome is still controversial, with limited data and follow-

up, surgical treatment remains the method of choice in patients in good general condition.

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