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Case Report

Three-Channeled Aortic Dissection in a Patient without Marfan Syndrome

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A 64-year-old man was admitted for evaluation of back pain. He did not have a Marfan syndrome (MFS)-like appearance, and had a history of a type B aortic dissection and total arch replacement. A connective tissue disorder had been suspected because of the histologic findings of the resected aortic wall. On admission, a computed tomography (CT) scan demonstrated a three-channeled aortic dissection (3ch-AD) measuring 63 mm in diameter. We planned to perform elective surgery during his hospitalization. On the fourth hospital day, he complained of severe back pain, and enhanced CT scan revealed an aortic rupture. The patients with 3ch-AD often have MFS. However, even if they do not have an MFS-like appearance, clinicians should consider fragility of the aortic wall in patients with 3ch-AD. If the aortic diameter is enlarged, early surgery is recommended. In particular, if a connective tissue disorder is obvious or suspected, emergent surgery is warranted.

Keywords: three-channeled aortic dissection, Marfan syndrome, Marfan-syndrome-related disease, rupture, connective tissue disorders

Introduction

Three-channeled aortic dissection (3ch-AD), which has two adjacent false lumens, is rare. Patients with 3ch-AD often have Marfan syndrome (MFS).[1)] In such patients, the 3ch-AD is histologically characterized by cystic medial necrosis, and the aortic wall readily dissects and ruptures. Therefore, urgent or emergent surgery for 3ch-AD has been recommended in some reports. However, elective surgical treatment of 3ch-AD has also been reported. Overall, the optimal therapeutic strategy for 3ch-AD has not been fully determined. We herein report a case of 3ch-AD with an enlarged aorta resulting in aortic rupture in a patient without MFS during hospitalization.

Case Report

A 64-year-old man was admitted to our hospital for evaluation of sudden back and lower abdominal pain in April 2012. He had a past history of a type B aortic dissection; however, its onset went unnoticed because of the absence of typical chest and back pains. The aortic dissection was incidentally revealed by a computed tomography (CT) scan performed in May 2007. The false lumen communicated with the true lumen by blood flow (**Fig. 1A**). In May 2008, he underwent aortic valve

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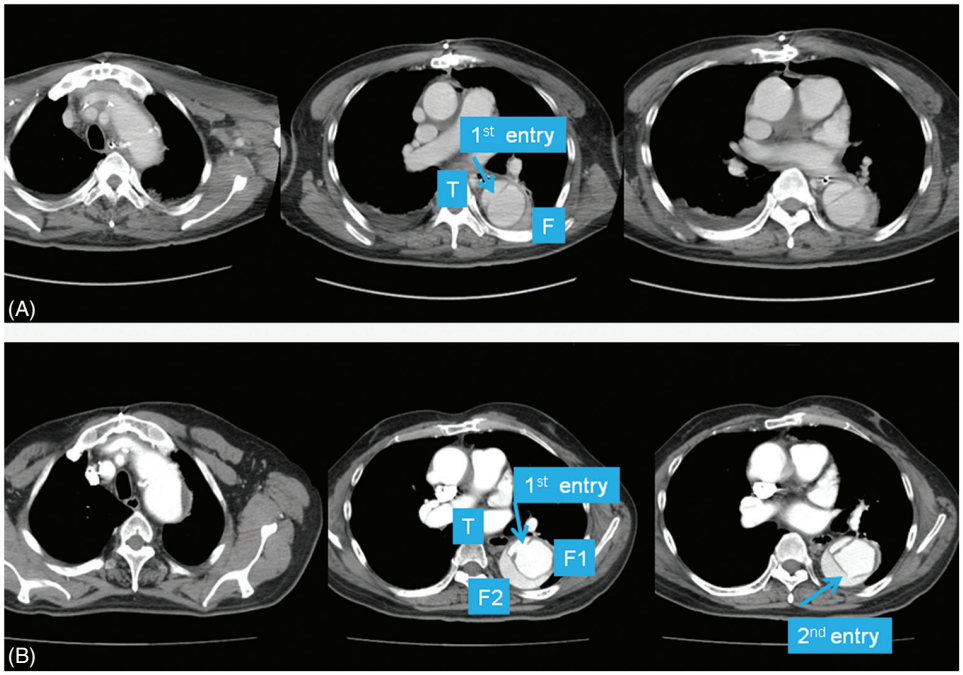

Fig. 1 (**A**) Computed tomography scan demonstrates post-total arch replacement in 2008 for aortic dissection in chronic period. The entry site of the first aortic dissection could not be removed at this surgery. (**B**) Computed tomography scan demonstrates the three-channeled acute aortic dissection on admission in 2012. The second entry site of three-channel aortic dissection was shown. T: true lumen; F1: previous false lumen; F2: new false lumen

replacement with total arch replacement for severe aortic regurgitation and distal arch dilatation. However, entry site could not be removed at this surgery. Histologic findings of the resected aortic wall revealed fragmented and decreased collagen fibers, and a connective tissue disorder was suspected.

On the patient's admission in April 2012, a CT scan demonstrated an acute type B aortic re-dissection with a three-channeled (3ch) lumen (**Fig. 1B**). The range of the 3ch-AD was from the distal arch to the left common iliac artery, which was almost the same range as the previous dissection. The re-entry of the second false lumen could not be found. The diameter of the proximal descending aorta had increased from 43 mm 4 years prior to 63 mm on this admission, which was suspicious of a rapid expansion of the aorta. The patient's systolic blood pressure was controlled within 130 mmHg during this hospitalization. Specific characteristics of MFS were not found during the patient's physical examination, and he had no family history of aortic disease at a young age.

We discussed the therapeutic strategy with a cardiovascular surgeon on admission and determined that elective surgical repair would be performed during this hospitalization. On the fourth hospital day, he complained of severe back pain and his systolic blood pressure decreased to 80 mmHg. Another emergent enhanced CT scan demonstrated leakage of contrast media from the aortic wall at the distal arch just distal to the replaced graft and proximal descending aorta level; bilateral hemothorax was also present (**Fig. 2**). We performed emergent descending aorta replacement for repair of the aortic rupture. A histologic specimen obtained from the surgery showed fragmented and decreased collagen fibers (**Fig. 3**), coincided with the previous histologic examination. Although he was rescued, his comatose state persisted. On postoperative day 56, he was transferred to another hospital.

Discussion

3ch-AD is rare, and its incidence has not been fully clarified. Few series of 3ch-AD among patients with AD have been published. Of 432 patients who underwent surgical treatment for AD, 31 (7.3%) had 3ch-AD, and 18 of these 31 patients (53.0%) had MFS.[1)] In contrast, in the subgroup of patients with both AD and MFS, the incidence of 3ch-AD varies. One report stated that 5 (4.2%) of 102 patients with MFS who underwent

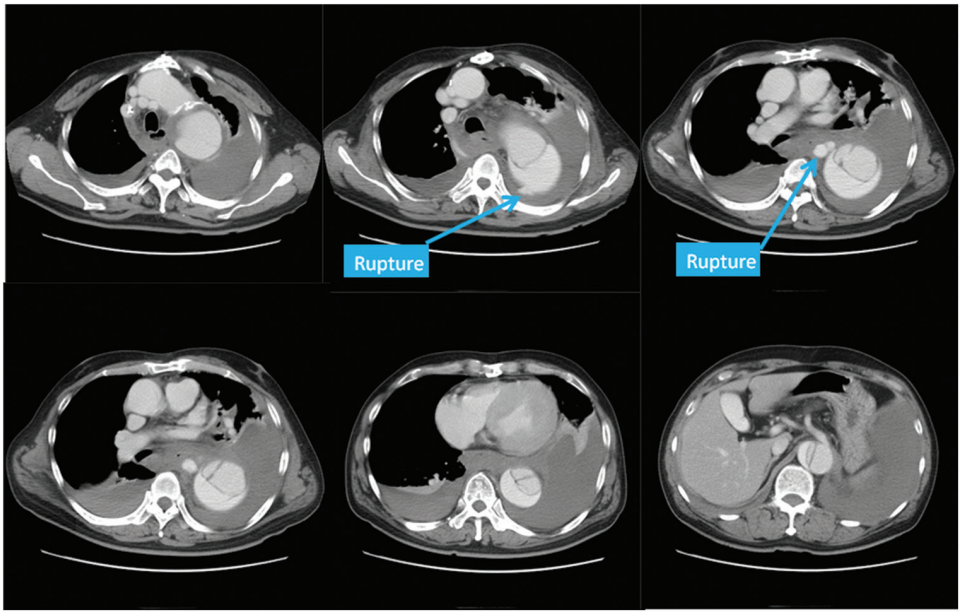

Fig. 2 Computed tomography scan demonstrates rupture of three-channeled acute aortic dissection on fourth hospital day. Contrast media is leaking out of the aortic wall.

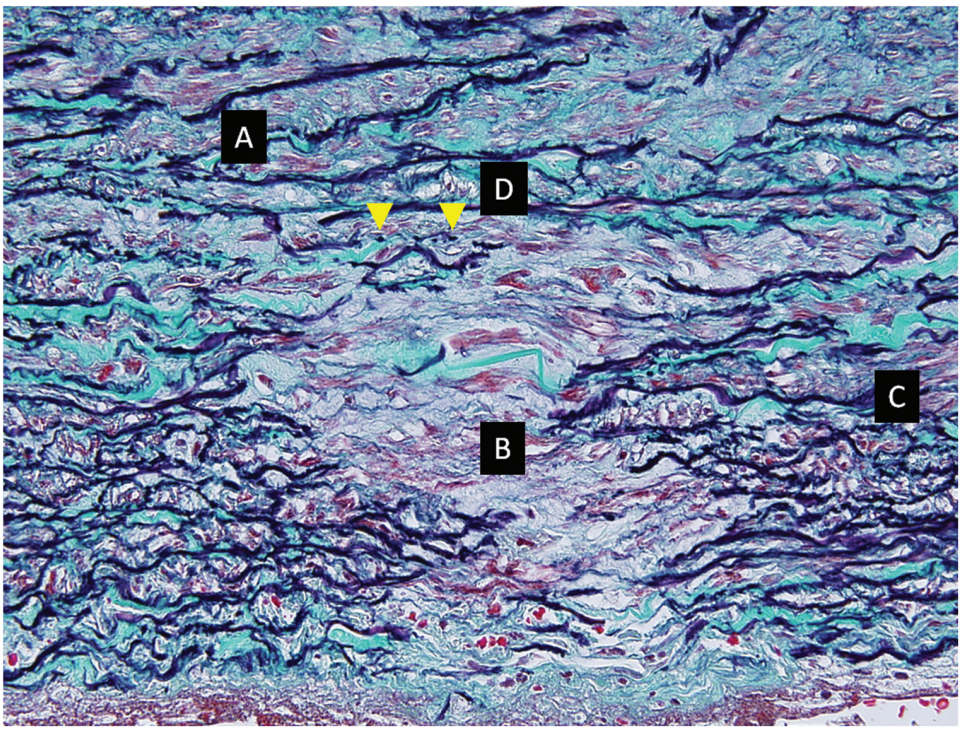

Fig. 3 Histologic examination of the excised tissue from the dissected aorta stained by Elastica Masson–Goldner shows elastic fiber (**A**) decrease and (**B**) discontinuation with (**C**) loss of smooth muscle cells and (**D**) accumulation of mucoid material without obvious atherosclerosis.

surgical treatment for AD had 3ch-AD,[2] and in another study, this rate was 15 (27.0%) of 55 patients.[3]

The etiology of 3ch-AD remains unclear. A connective tissue disorder is usually suspected in affected patients since many patients with MFS develop a 3ch-AD. Little is known regarding the causes of 3ch-AD other than the presence of connective tissue disorders. We speculate that some type of vascular fragility, due to a disease such as MFS, is present in almost all patients with a 3ch-AD. Recent studies have shown that patients who develop aortic disease at a young age often have MFS-related diseases other than MFS.[4] MFS-related diseases are known to be caused by mutations such as *TGFBR1* or *TGFBR2*, which is a cause of Loeys–Dietz syndrome;[5] *COL3A1*, which is a cause of vascular type Ehlers–Danlos syndrome;[6] *ACTA2*;[7] and others. Furthermore, patients with MFS-related disease often do not have an MFS-like appearance. Thus, an MFS-related disease other than MFS may be the cause of a 3ch-AD, even if the affected patient does not have an MFS-like appearance. However, MFS-related disease has not been previously reported in patients with 3ch-AD.

A 3ch-AD is often characterized by its occurrence in the descending aorta; a new false lumen is often found outside of a previous false lumen. Re-entry of a 3ch-AD often cannot be found,[1] and the aortic wall of the second false lumen is extremely thin because it was derived from the thin wall of the first false lumen. These characteristics must be associated with fragility of the aortic wall. A CT finding of a so-called "Mercedes Benz Mark Sign," with a balanced 3ch area, can be found in a 3ch-AD;[8–10] however, it may not necessarily be typical. In contrast, a CT finding of an unbalanced area in the three channels has also been reported.[11–13] The significance and frequency of the differences between these two types of CT findings have not yet been elucidated. We speculate that the area of the three channels may be unbalanced in the acute period, but that the 3ch area may become balanced during the time course. In addition, we should discriminate "aortic cobwebs," which is considered to be fibroelastic cords or bands projecting from the false lumen wall in aortic dissections,[14] from 3ch-AD.

The optimal treatment strategy for 3ch-AD has not been fully determined. One option is conservative treatment, such as the administration of anti-hypertensive drugs without emergent surgical repair. In one study, elective surgical repair was reportedly performed in 25 (81%) of 31 patients with 3ch-AD.[1] The other option is emergent surgical repair. For the reasons described above, the aortic wall of our 3ch-AD patient was strongly suspected to be fragile, and early surgical repair was consequently recommended.

As far as the reason of second aortic dissection resulting in 3ch dissection in this patient, aortic fragility would be associated. Although our 3ch-AD patient did not present with an MFS-like appearance, the histologic findings of a specimen obtained from past surgery showed fragmented and decreased elastic fibers, suggesting an aortic fragility caused by an MFS-related disease and pointing to the existence of a connective tissue disorder. The residual entry site of the first aortic dissection after previous total arch replacement should be also associated with second aortic dissection. As to the aortic rupture, in addition to the fragility to aortic wall, the CT scan did not reveal the re-entry of the second false lumen; the diameter of the descending aorta at this admission was also found enlarged to 63 mm with the suspicion of a rapid expansion. These findings were associated with the presence of an aortic rupture and indicated the need for emergent surgical repair on admission.

Conclusion

We have reported a case of rupture of 3ch-AD in a patient without MFS. The patients with 3ch-AD often have MFS. However, even if they do not have an MFS-like appearance, clinicians should consider fragility of the aortic wall in patients with 3ch-AD. If the aortic diameter is enlarged and re-entry site of the second aortic dissection could not be found, early surgery is recommended. And, in particular, if a connective tissue disorder is obvious or suspected, emergent surgical intervention is warranted.

Disclosure Statement

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