Clinical features of aortic dissection associated with Takayasu’s arteritis

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Takayasu’s arteritis (TA) is a chronic and nonspecific inflammatory vessel disease that involves the aorta and its major branches. TA results in stenosis, occlusion or aneurysmal degeneration of large arteries pathologically. Although TA is an unusual disease, it is more commonly observed to occur in Asian females compared to the general population worldwide. Aortic dissection is a very rare complication of TA, and only a limited number of TA cases presenting with aortic dissection have been published so far.[1–3] This study presents the clinical characteristics, treatment, and prognosis of patients with aortic dissection associated with TA based on a case series and intends to serve as a reference.

Seven cases were recorded to have aortic dissection associated with TA in our hospital from 2000 to 2016, of which six patients were female. The clinical characteristics, medical examination findings, and follow-up details of these patients are shown in Table 1. All patients had the history of hypertension. A clear history of tearing chest pain and tightness, which is characteristic of aortic dissection, was recorded only in two patients; the remaining five patients revealed no history of chest or back pain but complained of fatigue or breathless, recurrent chest discomfort, or pulselessness, and hypertension. Angiography demonstrated a linear, low-density intimal flap dividing the aorta into a true and false lumen (Figure 1A) in patient #1. The diameter of the false lumen was greater than that of the true lumen, with the former almost completely occluding the latter (Figure 1B). Bare-metal stents were implanted. Antihypertensive drugs (angiotensin converting enzyme inhibitors, calcium channel blockers, beta blockers, and diuretics) and prednisone were continued after the invasive intervention. Follow-up examinations were conducted; her blood pressure was noted to have returned to normal levels at one month, and the left ventricular ejection fraction was improved from 45% to 55% at six month.

Patient #2 was managed by endovascular stent-grafting for dissection of the descending aorta. Subsequently, the blood pressure was controlled within the normal range using a beta blocker and a calcium channel blocker. Aortic graft replacement was performed at for a dissection at the descending aorta in patient #7, after that, the patient was weaned off the antihypertensive drugs, and the blood pressure was maintained in the normal range on follow-up. The remaining four patients were treated with oral antihypertensive drugs and steroid. Among these four patients, one patient classified as Stanford type A subsequently died of aortic dissection rupture, while one patient died of congestive heart failure. The other two patients were treated with antihypertensive drugs and prednisone 10–30 mg per day because they were hemodynamically stable with no serious symptoms; the blood pressure was controlled well and the dissection remained stable during follow-up (details shown in Table 1).

Although TA primarily involves vascular damage to the aorta and its branches, aortic involvement leading to aortic dissection is relatively rare. TA affects all the layers of the arterial wall: in the acute inflammatory phase, lymphocytic infiltrate with giant cells of the media and fibroblast proliferation cause thickening of the intima in large vessels; in the chronic fibrotic phase, collagen replacement of elastic tissue in vessel walls results in thickening of all vascular layers leading to diffused rigid intimal fibrosis. It is possible that such fibrosis leads to a lowered risk of aortic dissection in TA patients.[1,4] In the seven cases of aortic dissection described here, it is possible that delayed or insufficient fibrosis might have resulted in the thinning of the arterial wall, eventually leading to arterial dilation or aortic dissection under the influence of local hemodynamics (such as hypertension).

Of the seven patients included in this study, five patients did not display any of the classic symptoms of aortic dissection, such as sudden-onset tearing chest pain. Moreover, variations of erythrocyte sedimentation rate (ESR) implied...
Table 1. Clinical characteristics, medical examination, and follow-up of the seven patients with aortic dissection associated with TA.

| Case | Age, yrs | Gender | History of TA, yrs | ESR, mm/h | LV, mm | LVEF, % | Aortic dissection type | Treatment | Follow-up, months | Clinical results |
|------|----------|--------|--------------------|-----------|--------|--------|------------------------|-----------|-------------------|-----------------|
| 1    | 31       | F      | 6                  | 15        | 56     | 45%    | B                      | BMS+medication | 52                | Stabilized      |
| 2    | 39       | F      | 20                 | 12        | 42     | 62%    | B                      | Stent-grafting+medication | 12            | Stabilized      |
| 3    | 30       | F      | 9                  | 46        | 54     | 54%    | B                      | Medication     | 13                | Stabilized      |
| 4    | 35       | F      | 15                 | 29        | 65     | 25%    | B                      | Medication     | 5                 | Death           |
| 5    | 36       | M      | 15                 | 86        | 62     | 42%    | A                      | Medication     | 1                 | Death           |
| 6    | 30       | F      | 13                 | 30        | 46     | 65%    | B                      | Medication     | 24                | Stabilized      |
| 7    | 59       | F      | 28                 | 14        | 59     | 50%    | B                      | Surgery+medication | 48            | Stabilized      |

BMS: bare metal stents; ESR: erythrocyte sedimentation rate; F: female; LV: left ventricular end-diastolic diameter; LVEF: left ventricular ejection fraction; M: male; TA: Takayasu’s arteritis.

Figure 1. Angiography showed a Stanford type B aortic dissection (A); and CT showed the false lumen was greater than the true lumen (B).

that ESR may not be correlated with possibility of complicated dissection. Hence, the clinical diagnosis of aortic dissection associated with TA may be delayed if only based on clinical symptoms and laboratory examinations. CT is an excellent tool for differentiating TA from atherosclerosis. In particular, when diagnosing dissections, it can reveal lacerations in the aortic dissection, identify the intimal flap, and differentiate between the true and false lumen. Further, it can also be used to determine the involvement of the aortic branches in such dissection. CT angiography can also clearly reveal narrowing, occlusion, dilation, calcification, and other pathological changes in the aorta as well as determine the thickness of the arterial wall.[5,6]

The treatment of aortic dissection associated with TA requires a combined therapeutic needs of TA and dissection both. A typical course of steroid treatment for TA comprises 1 mg/kg of oral prednisone, generally starting at 30–40 mg/d. ESR needs to be monitored regularly; if it reduces to within normal levels, the dosage of prednisone can be reduced by 5 mg every 3–4 weeks, until it reaches 20 mg/d. Subsequently, the dosage can be reduced by 2.5 mg every 3–4 weeks, until it reaches 10–15 mg/d. This course of treatment can lead to stabilization of TA for up to 15–20 years with no obvious side effects. Further, interventional or surgical treatment can be considered based on the patient’s clinical condition.[7] The aim of such invasive treatment is to reshape the damaged aorta and improve blood supply to the distal vessels.

The treatment for aortic dissection includes antihypertensive drugs, surgical treatment, or invasive intervention. At present, surgery is advised in acute type A aortic dissections with aortic reconstruction and restoration of antegrade flow into the true lumen, while type B aortic dissections are usually treated by medical or interventional treatment in stable patients.[8] Patients generally refrain from opting for surgical and interventional treatments due to the high levels of surgical trauma and higher rates of postoperative complications and mortality. However, with improvements in technology and increasing medical efficacy, the levels of trauma and the rate of complications caused by interventional treatment are expected to be greatly reduced.[9,10] Type B aortic dissection cases have been successfully treated by percutaneous stent-graft placement covering the entry tear in the descending aorta and even in the aortic arch, and suggest that percutaneous stent-graft placement in the dissected aorta is safer and produces better results than surgery for dissection.[11] In the present study, the majority of cases of aortic dissection associated with TA were classified as Stanford
type B. The mortality rate of untreated type B aortic dissection has been reported to be 91% within one month of onset or diagnosis and 89% within one year.\footnote{12} Moreover, 20%–28% of patients with type B aortic dissections develop thoracic aortic aneurysms or abdominal aortic aneurysms within 40–50 months, and fatal aortic ruptures occur in 18% of patients.\footnote{12,14} Therefore, any aortic dissections that are diagnosed should be actively treated in order to reduce the dialation of the false lumen and to avoid the risk of rupture.

In general, the occurrence of aortic dissection associated with TA is relatively low. It is typically not manifested by any classic symptoms, and Stanford type B aortic dissections are more common than type A. Patients with a long-term history of TA and poor control of hypertension should be monitored for aortic dissection using imaging studies. CT angiography is crucial for earlier diagnosis of dissection. The overall condition of the patient and the specific anatomical features of the aortic dissection would determine the most appropriate way and timing of the treatment. Conservative pharmacotherapy should be applied in cases of chronic aortic dissection associated with TA, wherein the hemodynamic status is stable, and there are no complications, such as distal limbic ischemia or ischemia of the kidneys and/or other organs. Appropriate treatment, focusing on strict blood pressure control, reduction in the heart rate, and prevention of aortic dilation, along with regular follow-up, can be help for controlling extension of dissection. Complicated aortic dissections, such as those involving severe aortic stenosis, poor perfusion of involved organs, or increasing size of the false lumen, should be actively treated by endovascular stenting or surgical intervention.

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