Different Clinical Course of Ascending Aortic Aneurysm In Monozygotic Twins

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Case Summary:
A 45-year-old woman without medical history of cardiovascular disease came to the clinic. She complained of dyspnea on exertion with cold sweating and dull aching pain on the left lower back for the past 6 months and she has suffered from more aggravating pain for the recent 1 month. Chest PA showed enlarged cardiac silhouette and prominent
ascending aorta and aortic arch and electrocardiogram showed -20 degree left axis deviation and T-wave inversion in the precordial leads. Echocardiogram demonstrated visible dissecting flap and wide false lumen from the root of ascending aorta to hepatic level of descending aorta (Figure 1A, 1B). It also showed markedly dilated sinus of Valsalva (diameter 50.4mm), sinotubular junction (diameter 53.5mm), and ascending aorta (diameter 58.1mm) with grade III/IV of aortic valve regurgitation (Figure 1A, 1B). Conventional computed tomography (CT) (Figure 2A) and ECG gated 64-multidetector computed tomography (MDCT) (Figure 2B) revealed annuloectasia and aneurysm of ascending aorta and dissected aorta from above the aortic valve to renal artery level. There are no laboratory abnormalities including CBC, biochemistry, and serology tests etc. She does not have any Marfanoid features. Surprisingly, when we reviewed her family history, we realized her monozygotic twin sister performed Bentall operation because of grade III/IV of aortic regurgitation due to annuloectasia and aneurysm of ascending aorta shown at the echocardiogram (Figure 3A, 3B) and ECG gated 64-MDCT (Figure 4) in our hospital 2 years ago. Her twin sister had no previous medical history and had suffered from exertional dyspnea with vague chest discomfort at that time. That time the current patient took a chest X-ray at the health center and she heard it was normal and so, she did not take any further work-up. Now she underwent modified Bentall operation (Figure 5).

Discussions
There have been very few case reports about identical ascending aortic abnormalities including aneurysmal changes, annuloectasias, and dissection in monozygotic twins. Typically, they have no previous medical problems and known predisposing factors of the aortic aneurysm with aortic regurgitation. The predominant inheritance pattern is autosomal dominant, with varying degrees of penetrance and expressivity. The familial group is significantly younger than the sporadic group, but not as young as the Marfan syndrome group (mean ages, 58.2 versus 65.7 versus 27.4 years). Aortic growth rate is highest for the familial group (0.21cm/y), intermediate for the sporadic group (0.16 cm/y), and lowest for the Marfan group (0.1cm/y) [1]. Thereafter, when encountering a patient with thoracic aortic aneurysm and the patient has no considerable previous medical problem, physicians should
meticulously check-up the first-order relatives of probands before dissection happens because familial thoracic aortic aneurysms tend to grow at a higher rate.

- **References**:

- **Key Pictures**

**Figure Legends**

Fig. 1A. Echocardiogram demonstrated visible dissecting flap (arrow) and wide false lumen from the root of ascending aorta to hepatic level of descending aorta.

Fig. 1B. Echocardiogram also showed markedly dilated sinus of Valsalva (diameter 50.4mm), sinotubular junction (diameter 53.5mm), and ascending aorta (diameter 58.1mm) with grade III/IV of aortic valve regurgitation.

Fig. 2A. Conventional computed tomography (CT) of chest revealed annuloectasia of aorta and dissected aorta from above the aortic valve extending to renal artery level.

Fig. 2B. ECG gated 64-multidetector computed tomography (MDCT) also revealed the dissected flap in the aorta which was shown at the conventional chest CT.

Fig. 3A, 3B. Echocardiogram of her twin sister also demonstrated the grade III/IV of aortic regurgitation due to aortic aneurysm and annuloectasia of aorta.

Fig. 4. ECG gated 64-multidetector computed tomography (MDCT) of twin sister.

Fig. 5. Op findings.
로 제출할 계획이 있으신 경우 아래사항을 기재하여 주시기 바랍니다.

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