

Familial Type A Aortic Dissection in Two Sisters within One Week in a Family without Marfan's Syndrome, Suggestive of Familial Aortic Aneurysm

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Abstract

We describe a large family in which 13 members over two generations developed aortic dissection or aortic aneurysm at a young age. Two sisters of this family presented with type A aortic dissection within one week with no previous medical history, Marfan's syndrome or other connective tissue disorders. Their 2D echocardiographic images were unique with floating tissue in the false lumen. Hereditary disorders, familial aortic aneurysms cause the individuals to develop an enlargement of the aorta. The aneurysm may occur anywhere, dilation at the level of the ascending aorta or the sinus of Valsalva, and it may progress over time to eventually tear or rupture. In both of our cases, the intimal flap started from the annulus and extended to the abdominal aorta, with severe aortic regurgitation. The Bentall procedure was performed for both of our patients; one of them died during the operation due to massive hemorrhage, but the other one tolerated surgery well. All their family members were subsequently screened; there was no medical history of Marfan's syndrome or other connective tissue diseases. Echocardiography, however, showed varying degrees of dilatation of the ascending aorta in most of the first and second-degree relatives with mild to moderate AI, suggestive of familial aortic aneurysms (*Iranian Heart Journal 2007; 8 (4): 57-59*).

Key words: aneurysm ■ aorta ■ dissection

Case Report

A 63-year-old woman with no medical history presented with acute, tearing chest pain with radiation to the back. On admission, she complained of severe pain in the back of her chest with a sensation of tearing hollow viscus.

Physical examination showed BP=100/60 mmHg, PR=80/min. and RR=15/min. Extremity pulses were symmetric, and cardiac examination revealed a holodiastolic murmur. She had no medical history, nor had she ever been admitted to hospital before.

She and her family had no criteria for Marfan's syndrome. Transthoracic (TTE) and transesophageal echocardiography (TEE) showed type A aortic dissection (intimal flap beginning from 6mm of the annulus and extending to the abdominal aorta with the possibility of right coronary artery involvement). There was severe aortic valve insufficiency (AI) with diastolic mitral regurgitation (MR) and aneurysm of the ascending aorta (6.6cm). Cardiac catheterization having confirmed the above data, the patient underwent emergent surgery

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