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A dissecting thoracic aortic aneurysm presenting with mild symptoms in a young woman

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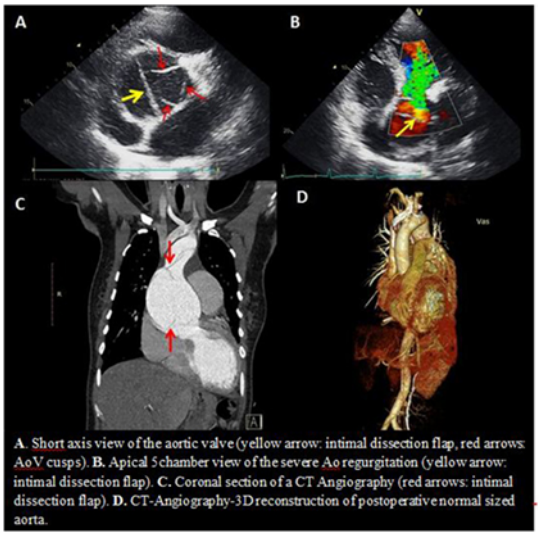
Introduction: Acute aortic dissection is a life-threatening condition associated with high morbidity and mortality rates, and it remains a challenge to diagnose and treat.

Case presentation: We present the case of a 29 years old young woman, brought to the emergency department with mild chest and back pain, shortness of breath on exertion with an onset of 12 days prior to admission. The patient has a history of 3 pregnancies delivered by natural birth and C-section (last one 2 years ago), but no personal cardiovascular disease history. She relates a family history of sudden cardiac death at a young age (mother, grandmother, great-grandmother) and a sister with diagnosed aortic aneurysm associated with a genetic mutation. Physical examination revealed a marfanoid habitus, diastolic heart murmur, low diastolic blood pressure (BP of 90/25 mmHg). Transthoracic echocardiography performed in the ED, discovered an anuloaortic aneurysm with a maximum diameter of 8.7 cm at the aortic root and a dissecting intimal flap of the ascending aorta up to the aortic arch. We also detected a dilated left ventricle with a low ejection fraction (30%) and a severe aortic regurgitation. The findings were confirmed by contrast-enhanced CT Angiography, showing the aneurysm of 8.4/8.7 cm and the intimal dissection flap with patency of both true- and false lumens. At this point we diagnosed an anuloaortic aneurysm with Stanford type A acute aortic dissection and a Marfan syndrome (according to the Ghent Nosology criteria). She immediately underwent surgery, the Bentall procedure was performed, consisting of a composite graft replacement of the entire ascending aorta and aortic valve, followed by direct implantation of the coronary arteries. Histopathological examination of the excised aortic wall reinforced the diagnoses of a connective tissue disorder. The early postoperative evolution was marked by the development of a left-sided hemiparesis and seizures, caused by an acute ischemic stroke. The patient was discharged after the complete resolution of the neurological symptoms under appropriate medication. Recommendations were made for periodic follow-up, genetic testing and the screening of her children after the age of 10 years old.

Conclusion: Discussions evolve around the interesting fact that she managed to survive 3 pregnancies, natural births and a C-section, without developing an aortic dissection or even a possible fatal rupture up until this presentation. It is imperative to mention that the lack of a follow-up from a general practitioner, who could have notice the signs of a possible connective tissue disorder, have possibly led to the development of such aortic sizes. The current guidelines sustain the importance of surgical approach in all types of ascending aortic dissection, the only treatment proven to increase the short- and long term mortality.

Abstract P723 Figure. Imaging studies



A. Short axis view of the aortic valve (yellow arrow: intimal dissection flap, red arrows: AoV cusps). B. Apical 5chamber view of the severe Ao regurgitation (yellow arrow: intimal dissection flap). C. Coronal section of a CT Angiography (red arrows: intimal dissection flap). D. CT-Angiography-3Dreconstruction of postoperative normal sized aorta.

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