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MARFAN SYNDROME PRESENTED AS ASYMMETRICAL AORTIC ROOT ANEURYSM AND SPONTANEOUS ISOLATED AORTIC ABDOMINAL DISSECTION

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Introduction: Isolated abdominal aortic dissection (IAAD) refers to aortic dissection (AD) inferior to the diaphragm, which is very rare. Marfan syndrome (MFS) is an autosomal dominant connective tissue disorder, which is mostly presented as garlic-like aortic aneurysm in cardiovascular system. To the best of our knowledge, no such a case concerning MFS presented as IAAD has been reported before.

Case Report: A 37-year-old female with no history of hypertension was referred to our hospital for chest tightness. She had been initially diagnosed as spontaneous IAAD one month before with successful analgesic and antihypertensive treatment at a local hospital. On physical examination, the patient's blood pressure was 138/80 mmHg. Laboratory tests showed D-dimer of 2.13 ug/ml (<0.5 ug/ml), FDP of 8.42 ug/ml (0-5 ug/ml). Echocardiography revealed severe aortic regurgitation. CT showed an obvious compression of the left atrium by asymmetrical dilatation of non-coronary sinus. Abdominal aortic dissection originated distally to the superior mesenteric artery (SMA) ostium and extended downward to the bilateral common internal iliac artery. Then FBN1 mutation was found by gene analysis, thus the diagnosis of Marfan syndrome (MFS) was confirmed. She underwent Bentall procedure only and discharged on calcium channel blocker and warfarin. On follow-up at 1 year, the patient was doing well physically. CT scanning found no obvious extension of IAAD or aortic growth in diameter.

Lessons: First, this is the first case report of MFS combined with both asymmetrical aortic root aneurysm and spontaneous IAAD, which enriches our understanding of the clinical manifestations of MFS. Second, we should take aorta as a whole organ in which multiple levels of lesions may occur simultaneously, so it is necessary to assess the whole aorta in order to prevent serious missed diagnosis. Third, for MFS patient with IAAD, conservative treatment under careful surveillance seems satisfactory on a short-term follow up. Further follow-up is still needed to confirm the long-term effect.

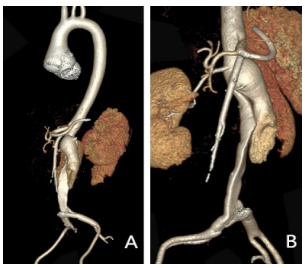

Figure 1: A: 3-D re-construction of CT scanning of the whole aorta. B: 3-D re-construction of CT scanning of the abdominal aorta.

Recent Publications

1. Cheng L, Huang F, Chang Q, Zhu J, Yu C, Liu Y, et al. (2010) Repair of extensive thoracoabdominal aortic aneurysm with a tetrafurcate graft: midterm results of 63 cases. The Heart Surgery Forum 13(1):E1-6.
2. Sun X, Zhang L, Yu C, Qian X and Chang Q (2014) One-stage repair of extensive aortic aneurysms: mid-term results with total or subtotal aortic replacement. Interactive Cardiovascular & Thoracic Surgery 18(3):278-82.
3. Zhang L, Yu C, Qian C, Luo X, Qiu J and Liu S (2016) Comparison of gene expression profiles in aortic dissection and normal human aortic tissues. Biomedical Reports 5(4):421-7.
4. Liu P, Qian C, Qian X, Sun X, Yu C, Tian C, et al. (2016) Early and mid-term results after hybrid total arch repair of DeBakey type I dissection without deep hypothermic circulatory arrest. Interactive Cardiovascular & Thoracic Surgery 23(4):608.

Biography

Cun Tao Yu is one of the most famous cardiovascular surgeons in China. He is especially good at all kinds of operation of large vessels and has completed over 1500 operations such as total arch replacement and thoracoabdominal aortic replacement. Jin Lin Wu is his doctoral candidate.

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