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Case report

Successfully treated contained circular rupture of the ascending thoracic aorta in a patient with a bicuspid aortic valve

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Introduction

Few cases of spontaneous rupture of the ascending thoracic aorta have been reported [1–3]. A spontaneous rupture eludes definition, but can be described as a sudden event not associated with aortic aneurysm, dissection or trauma, inflammation of the aortic wall or erosion from a neoplastic mass [1–3]. Spontaneous rupture of the ascending aorta should be considered in patients with clinical signs of aortic dissection [2]. The incidence of thoracic aortic rupture is much higher in patients with bicuspid aortic valve as well as in patients with connective tissue disease. When BAV is combined with dilatation or aneurysm of ascending aorta, it is called bicuspid aortopathy [4,6,8,10].

Here, we present the rare case of a patient with contained circular rupture of the ascending thoracic aorta, bicuspid aortic valve and histologically declared cystic medial necrosis, who was successfully treated surgically and survived.

Case report

A 71-year-old male patient with a history of arterial hypertension was referred to a comprehensive cardiology evaluation. The patient was planned for a surgical treatment for recently

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Table 1 – The diameters of cardiac chambers and aorta obtained from TTE.

Variables	Diameter
Left ventricular end-diastolic diameter (PLAX[a])	57 mm
Left atrium antero-posterior diameter (PLAX[a])	40 mm
Right ventricular diameter	
PLAX[a]	33 mm
A4C[b] (RVD1[c])	38 mm
Interventricular septum thickness	12–13 mm
Posterior wall thickness	13 mm
Ascending aorta	47 mm
Aortic bulbus	45 mm
Aortic arch	30 mm

[a] PLAX – parasternal long axis.
[b] A4C – apical four chamber.
[c] RVD1 – right basal ventricular diameter.

diagnosed prostatic adenoma. A systolic murmur at the site of the aortic valve was detected within the preoperative clinical examination by an outpatient internist, who sent him into the hospital. The patient declared one single attack of 10 min duration, with severe chest pain accompanied with vegetative symptoms two weeks prior to the elective admission to the hospital. The pain had sudden onset, resolved spontaneously and had not relapsed. The patient did not seek any medical care. In the family history, there was no mention about aortic dissection or sudden cardiac death and the patient did not have any previously known morphological abnormalities or genetic disorder.

Bicuspid aortic valve (BAV) with severe stenosis (mean gradient 45 mmHg, V_{max} 3.9 m/s) and moderate regurgitation (PHT 450 ms, vena contracta 6 mm) were found during the initial transthoracic echocardiography (TTE) (Table 1). Additionally, dilatation of aortic bulb and ascending aorta (AA) (47 mm) were present. A circular pericardial effusion (PE) without the echocardiographic evidence of cardiac tamponade was another pathological finding.

The next day, a cardiac catheterization was performed as a part of the complete cardiologic evaluation. Coronary angiography demonstrated a diffuse coronary atherosclerosis without significant stenosis. Aortography showed a massive calcification of the aortic valve and a dilatation of the AA with an aortic lumen contour irregularity (Fig. 1). There was no presence of aortic coarctation in the proximal part of the descending aorta. According to those findings, a computed tomography (CT) angiography was performed the day after the cardiac catheterization, where the dilatation of the AA (47 mm) and a short membrane-like structure of unknown aetiology located approximately 50 mm distal to the aortic valve was confirmed. The CT scan also verified the presence of PE (Fig. 2). Very short atypically located aortic aneurysm or atherosclerotic plaque or aortic dissection was assumed in differential diagnosis. To clarify the diagnosis, transoesophageal echocardiography (TOE) was performed and showed a discontinuousness of the aortic wall approximately 3 mm wide localized 50 mm distally from the aortic valve and communicating with the pericardial cavity (Fig. 3). The examination showed no evident dissection membrane. The TOE also confirmed the morphology of the aortic valve – a bicuspid valve with one raphe.

Based on the findings listed above, the patient was indicated for an urgent cardiac surgery. A contained circular aortic rupture of the AA approximately 50 mm distally from the aortic valve and 20–30 mm from ST junction without any signs of dissection, as well as the presence of haemopericardium, was confirmed perioperatively (Fig. 4). The patient underwent the aortic valve replacement with bioprosthesis and the AA replacement. Histological findings corresponded to the clinical manifestation of the rupture of the aortic wall, which had probably arisen in association with the presence of bicuspid aortic valve and regional changes due to the cystic medial necrosis (Fig. 5). Furthermore, the post-operative period passed without any complications and the patient was discharged from the hospital the seventh day after the surgery. At one-year follow-up, the patient had not reported any cardiac difficulties.

Discussion

A spontaneous aortic rupture without previously presented aortic aneurysm, dissection, trauma, inflammation of the

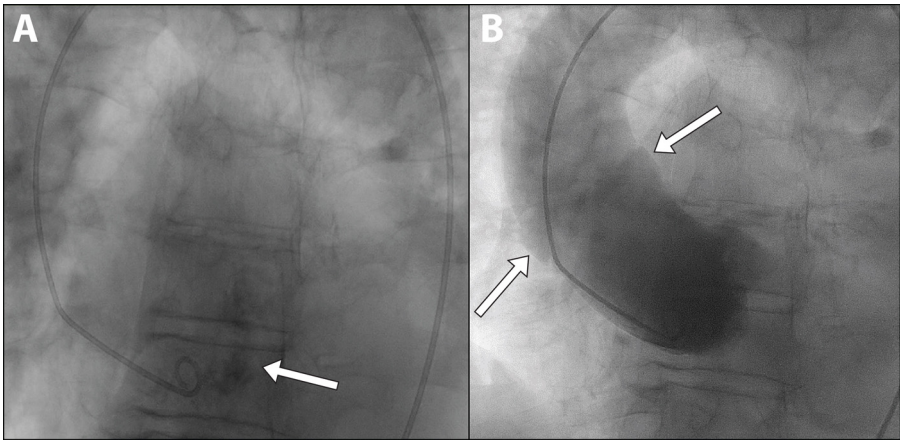

Fig. 1 – The aortography before the contrast agent injection (A) showed the massive calcification of the aortic valve (labelled with arrow). The ascending aorta was without calcifications. The aortography with contrast agent (B) showed the dilatation of the AA with the aortic lumen contour irregularity (labelled with arrows).

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