Recurrent systemic embolism caused by thrombosis in a stenotic bicuspid aortic valve

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Thrombosis in a native aortic valve is a rare complication which may lead to systemic embolization. A few cases of aortic thrombosis in previously abnormal valves have been described. In this report, we describe a 42-year-old male who suffered two acute ischaemic attacks, one in the upper right limb and another in the cerebral territory supplied by the left sylvian artery, from a thrombus that developed in a bicuspid and stenotic aortic valve. The diagnosis was made with transthoracic and transoesophageal echocardiography, and the patient subsequently underwent surgery. In cases of bicuspid aortic valves, we should think of thrombosis as a possible complication with its resulting risk of embolism, and assess such patients with transthoracic and transoesophageal echocardiography, thus enabling their early detection and treatment.

KEYWORDS
Stenotic bicuspid aortic valve; Thrombosis; Transoesophageal echocardiography; Source of embolism

Background
The development of thrombi in native aortic valves with or without previous disease is a rare phenomenon which should be taken into account and diagnosed early, because it entails a high risk of systemic embolism.1–3 Here, we report a patient with aortic stenosis due to a bicuspid valve associated to thrombosis and complicated by recurrent episodes of systemic embolism.

Case presentation
A 42-year-old man with a history of hypertension, hypertriglyceridemia (216 mg/mL) and hyperglycemia (137 mg/mL) was referred for a diagnostic echocardiogram due to a heart murmur. At the time of the exam, he had no angina or dyspnoea during his usual activities, a ++++/4 systolic murmur was heard in the aortic area with a thrill, which radiated to the suprasternal notch. The electrocardiogram (ECG) showed sinus rhythm at 70 beats per minute, pathologic Q waves in inferior leads compatible with an old inferior infarction, and abnormalities of the anterolateral repolarization, but no arrhythmias.

The patient reported an episode of acute arterial occlusion of the right upper limb in 2001, which required emergency surgery (thromboembolectomy); reperfusion was successful with no long-term complications.

The transthoracic echocardiogram (TTE) showed preserved left ventricular diameters and global systolic function (diastolic diameter: 4.89 cm, systolic diameter: 2.77 cm; shortening fraction: 43%) and moderate concentric hypertrophy (septal thickness: 1.62 cm, posterior wall thickness: 1.15 cm), changes secondary to severe aortic stenosis (peak gradient: 77 mmHg, mean gradient: 45 mmHg) due to a bicuspid valve, with a mass on the ventricular side of the posterior leaflet. In view of these findings, the patient underwent a transoesophageal echo, which confirmed the presence of a highly mobile and friable mass of ~20 mm in size, adhered to the ventricular side of the posterior leaflet (Figure 1).

With a diagnosis of cardiac source of embolism originating in a stenotic aortic valve, a valve replacement was indicated. Although undergoing routine pre-surgical workup, the patient suffered a sudden right facio-brachio-cranial hemiparesis (1 April 2005). His brain computed tomography showed cortico-subcortical cerebral ischaemia in the left Sylvian territory, which required hospital admission and treatment (nimodipine 30 mg/day; biperidene 2 mg/day; and physical therapy) for 60 days. The patient recovered partially from his right facio-brachio-cranial hemiparesis.
**Figure 1** Echocardiographic studies. (A) Transthoracic echocardiography shows severe aortic stenosis (peak gradient 77 mmHg, mean gradient 45 mmHg) with a mass adhered to the ventricular aspect of the sigmoid leaflet. (B) Transthoracic echocardiography shows a longitudinal view of the aorta, and the zoom in the LV outflow tract shows a bicuspid aortic valve with a mass in the ventricular side of the non-coronary leaflet. (C) Transoesophageal echocardiography, short axis of the aortic valve, shows a bicuspid aortic valve with a mass in the non-coronary leaflet; *mass. LA, left atrium; LV, left ventricle; Ao, aorta; RVOT, right ventricle outflow track; PA, pulmonary artery; RA, right atrium; LVOT, left ventricle outflow track; RV, right ventricle; M, mass.

**Figure 2** Pathology. Macroscopic and microscopic exam. (A) View of the surgical specimen, showing the thrombus adhered to the ventricular side of the bicuspid aortic valve which is severely stenotic. (B) Vascular dilatations (green arrows) in the aortic valve, and granulation tissue with neovascularization (vertical channels marked with an asterisk) on the base of the thrombus. (C) Vascular dilatation and opening in the ventricular side where the thrombus has formed. Thickening of the aortic valve due to fibrosis (red arrow). Aortic surface (*); ventricular surface (**).
Laboratory exams, including antithrombin III levels, fibrinogen, protein C, protein S, and plasminogen, were normal. The Doppler-echo exam of the neck vessels and coronary angiography performed before surgery showed no abnormalities.

Since coronary angiography was normal and TTE and left ventriculography did not show wall motion abnormalities, the old inferior infarction showed in the ECG could be compatible with a mild and non-transmural infarction and could be explained by a silent coronary embolism.

Finally, on 24 August 2005, the definitive surgery was performed, which consisted of an aortic valve replacement with a #21 CarboMedics valve, a bileaflet mechanical prosthesis.

The surgical procedure confirmed the diagnosis of aortic stenosis due to a bicuspid valve and the presence of a multilobulated, vegetating mass, measuring ~1.5 by 2 cm in diameter, mainly adhered to the ventricular side of the non-coronary posterior leaflet. The diagnosis of a thrombus developed from the ventricular aspect of the valve was confirmed by Pathology, and no microorganisms were identified (Figure 2).

The patient suffered no post-operative complications and was discharged 7 days post-surgery, with a treatment of 50 mg/day of atenolol, acenocumarol, and 300 mg/day of ranitidine.

After 20 months of follow-up, the patient remains asymptomatic with International Normalized Ratio levels between 2 and 3.

Discussion

Aortic thrombosis may originate in valves with no apparent anatomic damage, in valves that have suffered endothelial damage during traumatic cardiac catheterization procedures, or as a complication of chronic deformities.1,3,4

In patients in whom no valve lesions are detected, the antiphospholipid syndrome or other haematological disorders which favour the development of autoimmune complexes with affinity for the endothelium may lead to formation of valve thrombi.5,6

Bicuspid aortic valves are a common congenital defect, with a frequency of ~2% in the general population and a higher prevalence (2:1) in males.7,8 It is associated to malformations such as aortic coarctation and Turner’s syndrome, entails a higher incidence of aneurysms and aortic dissection, as well as a higher risk of developing infective endocarditis.9–11

However, only a few authors have underlined the potential of bicuspid valves for developing valve thrombosis.

Clinical and pathological data obtained in cases of surgically resected aortic valves have defined a subgroup of post-infectious etiology, in which diffuse fibrosis and neof ormation vessels were identified. Bicuspid aortic valves with this pathological substrate comprise a subgroup that is prone to develop thrombosis in the valve surface or inside areas of calcification.12

Conclusion

In cases of bicuspid aortic valves, we should think of thrombosis as a possible complication with its resulting risk of embolism, and assess such patients with transthoracic and transoesophageal echocardiography, thus enabling their early detection and treatment.

Conflict of interest: none declared.

Authors’ contribution

R.J.M. and T.F.C. attended the patient, prepared the manuscript and figures and performed the echocardiographic images and participated in the manuscript description. C.E.P. acquired the pathological images. H.A.P. and L.A.V. participated in the design and review of the manuscript. All authors read and approved the final manuscript.

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