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Clinical Case Report

Vascular malformation in a bicuspid aortic valve

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ABSTRACT

We report here a case of capillary vascular malformation developed in a bicuspid aortic valve incidentally detected during valve replacement in a 67-year-old male patient. The International Society for the Study of Vascular Anomalies (ISSVA) classification was used to classify this vascular lesion instead of using the term hemangioma. The differential diagnosis and the literature are reviewed.

A 67-year-old male patient was admitted for radiofrequency catheter ablation of recurrent atrial fibrillation. He had previously known bicuspid aortic valve (BAV) and mild aortic valve regurgitation. Before the procedure, transthoracic echocardiography showed worsening of the aortic valve regurgitation, dilatation of the left ventricle (LVEDD: 70 mm), decreased LVEF at 37%, and ascending aorta aneurysm. The CT-scan and MRI confirmed a 54 mm-aneurysm of the tubular ascending aorta. Surgical replacement of aortic valve and ascending aorta by Bentall procedure was performed as well as radiofrequency pulmonary epicardial veins isolation. Given a bluish area on a cusp, the cardiac surgeon suspected endocarditis.

The bicuspid aortic valve, non-raphe type, was composed of two cusps measuring 5×1.3 cm and 3×0.8 cm mildly thickened and fibrous without calcifications. A brown 0.5 cm area was observed on both aspects in the middle of the larger cusp (Fig. 1). At histology (Fig. 2) this area corresponded to a cluster of abnormal vascular blood vessels containing red blood cells. They were limited by a thin wall covered by a flat endothelium positive for CD31 and CD34, and negative for podoplanin. The abnormal vessels were looking like dilated capillaries or postcapillary venules. This lesion was consistent with a capillary malformation according to the International Society for the Study of Vascular Anomalies (ISSVA) classification [1]. Focal disruption of the fibrosa layer was observed. The lesion was covered by a thin thrombus on the ventricular aspect. Endocarditis was ruled out.

The ISSVA stratified vascular anomalies into true vascular tumors and vascular malformations. Although ISSVA proposed since 1996 a classification adequate for clinical and pathological diagnostic purposes, old nomenclature persists and can lead to confusion. Updated ISSVA classification [1] recognizes the present case as a capillary malformation, withdrawing the term hemangioma commonly used in many papers. The term hemangioma is pointing a true proliferative vascular tumor whereas capillary malformations are congenital persistent lesions with no or slow progression.

In the literature cardiac hemangiomas, which should be now classified as vascular malformations, are included in benign cardiac tumors and represent less than 5% of cardiac tumors [2]. Cardiac valve localization is extremely rare and is not mentioned in WHO classification [2]. It has been reported in a few cases mainly in the tricuspid and mitral valves [3]. In the aortic valve, only two cases have been previously published [4,5]. In a case [5] the lesion was incidentally discovered as in our case in the valve during valve replacement. In the other case [4] preoperative transesophageal echocardiography detected a nonspecific small mass attached to the noncoronary cusp suggesting an endocarditis. In all the cases histopathological examination of the valve affords to make the diagnosis.

For the differential diagnosis, histology easily rules out true tumors, mainly myxomas and angiosarcomas, organized thrombus, and blood cysts. Blood cysts are described in infants and young children as endocardial invaginations mainly along the line of closure of valves. They are endocardial diverticula connected to the cardiac cavities. Cardiac varices are dilated veins and do not involve valves.

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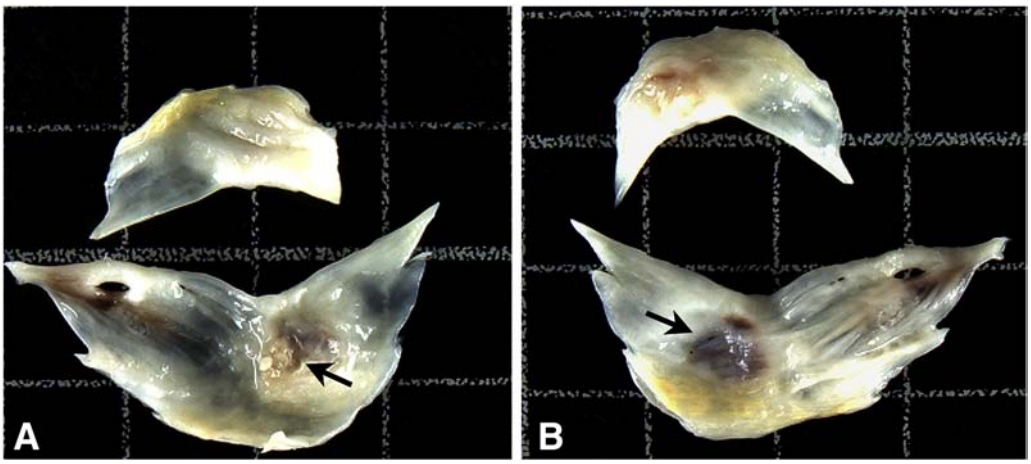

Fig. 1. A–B: Bicuspid aortic valve: Panel A: Ventricular aspect; Panel B: Aortic aspect. Both cusps are moderately thickened, fibrous, and non-calcified. Note on both aspects a brown hematic area (arrow) with a rough pattern due to mural thrombus on the ventricular aspect mimicking a vegetation during surgery. (Centimetric scale)

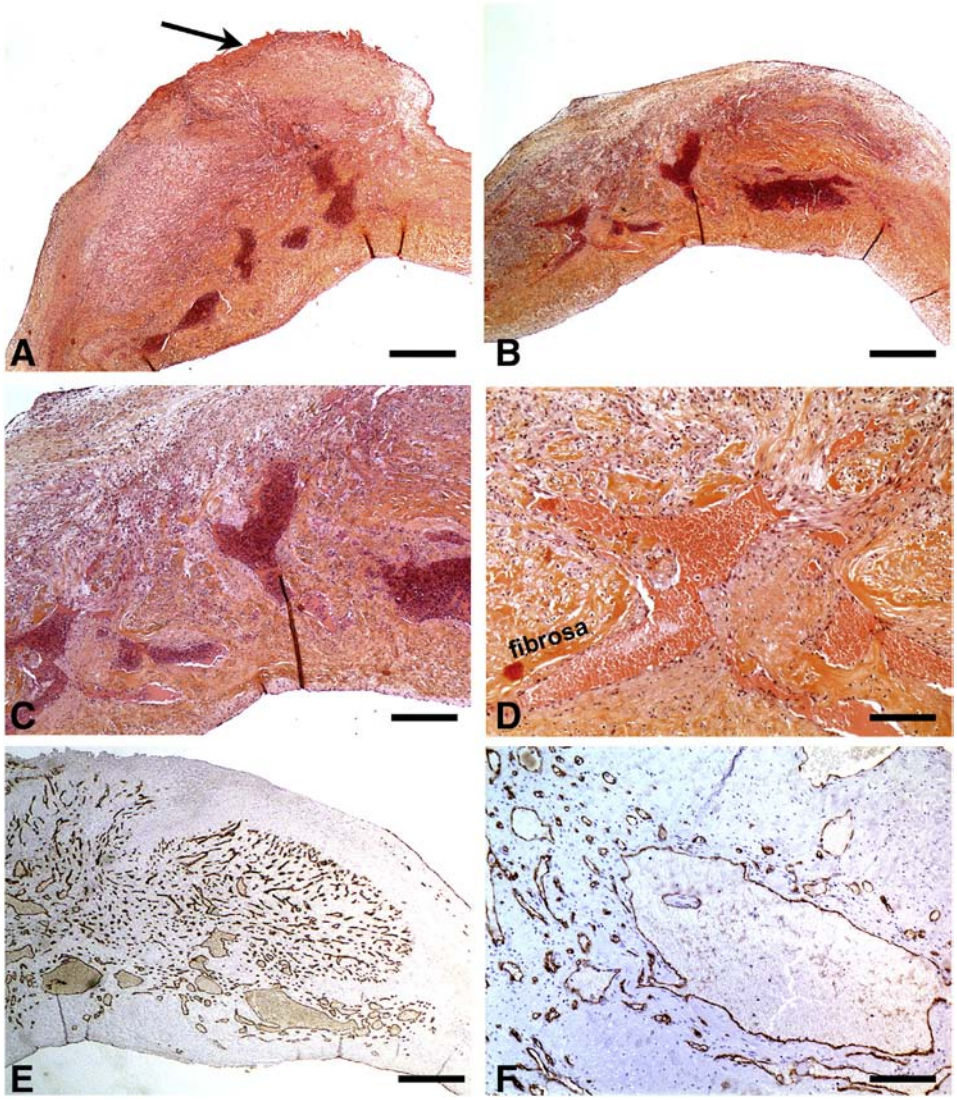

Fig. 2. A–F: Histology of the valve: Panels A, B, and C: Low magnifications show abnormal blood vessels looking like dilated capillaries and/or postcapillary venules throughout the thickness of the cusp. Note mural thrombus (arrow) in panel A. H&E stain; Panels A and B: Bar=0.5 mm; Panel C: Bar=0.25 mm. Panel D: The fibrosa is disrupted by the vascular malformation. H&E stain; Bar=130 μm. Panels E and F: The CD34 labeling of the endothelial lining clearly shows both smaller and larger abnormal vessels throughout the cusp. CD31 immunohistochemistry; Panel E: Bar=0.5 mm; Panel F: Bar=130 μm.

We have described a rare subclinical valve lesion which can mislead the diagnosis to endocarditis unless histology is performed.

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