Aortic wall papillary fibroelastoma

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Papillary fibroelastomas are rare tumors usually involving the cardiac valves. We present an extremely rare case of a papillary fibroelastoma originating from the aortic wall. The patient was successfully treated with complete tumor resection and a valve-sparing procedure to restore the aortic root anatomy.

CLINICAL SUMMARY

A 77-year-old woman was admitted to the University Hospital of Salamanca with a 3-month history of progressive dyspnea and angina pectoris on exertion. On admission, physical examination laboratory findings and electrocardiogram did not reveal abnormalities. An echocardiogram showed a 2 × 2-cm mobile mass located immediately above the aortic valve in contact with the aortic root wall (Figure 1, A). The aortic valve looked normal and had a mild central regurgitation jet. A dilatation of the ascending aorta also was noted, and an enhanced computed tomography was indicated. Computed tomography images confirmed the presence of a small mass inside the aortic root lumen, apparently attached to the aortic wall next to the commissure between the right and noncoronary sinuses (Figure 1, B). An ascending aorta diameter of 50 mm was verified. Because of the unpredictable risk of embolism and the patient's symptoms, an urgent surgical correction was indicated.

A median sternotomy was performed, and standard cardiopulmonary bypass was established with cannulation of the proximal aortic arch and the right atrium. The distal ascending aorta was crossclamped, and the heart was arrested using antegrade and retrograde blood cardioplegia. The ascending aorta was resected from the sinotubular junction to 1 cm proximal to the brachiocephalic trunk. The aortic root was then exposed, and an irregular glistening yellowish-white 2 × 2-cm mass with jellylike appearance and small nodules on its surface was seen attached to the aortic wall of the noncoronary sinus, close to the top of the noncoronary right commissure (Figure 2, A and B). The aortic valve was tricuspid and structurally normal.

To achieve complete resection of the tumor, an arteriotomy-based triangular portion of the noncoronary sinus was excised with the mass (Figure 2, C). The wall root was reconstructed with a bovine pericardial patch slightly larger than the aortic defect using a continuous 4-0 polypropylene suture (Figure 2, D). To avoid suturing close to the commissure, a tissue patch was fixed in part to the wall of the right coronary sinus. The top of the commissure was then attached to the pericardial patch with an additional stitch buttressed in the autologous pericardium. Because of moderate root dilatation and central aortic insufficiency, subcommissural triangles were plicated with 3 Teflon-buttressed 2-0 Ticron stitches (Covidien, Mansfield, Mass). Finally, the ascending aorta was replaced using a 28-mm Dacron graft. Transesophageal echocardiography confirmed the competence of the aortic valve after surgical procedure.

The tumor excised was friable and had a yellowish and jellylike appearance (Figure 2, E). When the tumor was submerged in water, it acquired villous on its surface like a sea anemone (Figure 2, F). Histopathologic examination confirmed a papillary fibroelastoma. After the operation, the patient had an uneventful recovery and was discharged 10 days after surgery. Postoperative echocardiography showed a well-functioning aortic valve.

DISCUSSION

Papillary fibroelastomas are the second most common cardiac tumors, although they represent only less than 10%.¹ Although myxomas typically originate from the atrial wall, the papillary fibroelastomas usually grow on the valvular endocardium, mainly settled on the aortic, mitral, and tricuspid valves. The nonvalvular endocardial location is rare, and fibroelastomas are rarely found along the atrial or ventricular walls.² To the best of our knowledge, only 1 case of papillary fibroelastoma originating from the aortic wall has been reported.³

Because of higher-resolution imaging techniques and the widespread use of echocardiography, these cardiac tumors are increasingly found and usually discovered incidentally. Transesophageal echocardiography usually is sufficient to achieve a diagnosis. Symptoms associated with this tumor are variable. The majority of patients remain asymptomatic, but serious complications also could present in terms of transient ischemic attack, stroke, angina, myocardial infarction, or sudden death.^{2,4} Differential diagnosis includes infective vegetations, thrombi, and other cardiac tumors. Fibroelastomas are treated on the

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FIGURE 1. A, Transesophageal echocardiography focuses on the aortic root. A, Mobile mass 2×2 cm is attached to the aortic valve immediately over the aortic valve (arrow). B, Enhanced computed tomography scan showing the tumor inside the lumen of aortic root as an intraluminal filling defect (arrow). LA, Left atrium; LV, left ventricle; AV, aortic valve; AA, ascending aorta; RCS, right coronary sinus; NCS, noncoronary sinus; LCS, left coronary sinus.

basis of symptoms or to prevent potential complications. Surgical resection is strongly advocated when the tumor is located in the left side of the heart because of the risk of cardioembolic complications.^{4,5} Because of the tumor's benign nature, a simple excision usually is

enough, but careful resection is needed to avoid fragmentation of tumor tissue.

Additional surgical techniques may be required only in those cases with involvement of valves or other cardiac structures.^{1,4} An intraoperative finding of a gelatinous

FIGURE 2. A, Intraoperative findings of the yellowish mass over the aortic valve (arrow). B, Closer intraoperative view. C, Aortic wall defect remaining after complete resection of tumor with its implant surface. The aortic valve remains intact after resection. D, Triangle-shaped bovine pericardial used for restoring aortic root (arrow). E, Macroscopic view of surgical specimen showing the friable and glistening jellylike appearance of tumor. F, The tumor changed in water to its characteristic villous, arboreal, and sea anemone-like appearance, suggesting the diagnosis of papillary fibroelastoma. AA, Ascending aorta; AV, aortic valve; RC, right coronary; RCS, right coronary sinus; LCS, left coronary sinus; NCS, noncoronary sinus; LC, left coronary.

yellowish mass and its characteristic change of appearance when submerged in water to a villous sea anemone-like tumor suggests the diagnosis of papillary fibroelastoma. However, the definitive diagnosis is confirmed by histopathologic examinations after surgery.

CONCLUSIONS

We present an exceptional case of a papillary fibroelastoma originating from the aortic wall of the noncoronary sinus. The patient was successfully managed with a valve-sparing procedure. No aortic regurgitation was detected postoperatively.

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Open transcatheter tricuspid balloon expandable valve-in-valve implantation for failed bioprosthesis

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Transcatheter valve-in-valve replacement is a reliable therapeutic option for degenerated bioprosthetic heart valves in high-risk patients. Successful transfemoral and transapical techniques have been described for aortic, mitral, and pulmonary bioprostheses. To the best of our knowledge, few cases of transcatheter valve-in-valve tricuspid valve implantation have been reported.¹⁻³

Van Garsse and colleagues¹ described the deployment of the prosthesis via the right internal jugular vein with ventricular pacing under cardiopulmonary bypass (CPB) support via peripheral cannulation. Weich and colleagues² and Cerillo and colleagues³ delivered the device via the right internal jugular vein without ventricular pacing.

Hon and colleagues⁴ and Cheung and colleagues⁵ delivered the transcatheter prosthesis via a right thoracotomy and direct puncture of the right atrium. We present a case of an “open” transcatheter valve-in-valve replacement in a high-risk patient who required conventional cardiac surgery for severe dysfunction of the aortic, mitral, and tricuspid heart

valves with a high frailty of right-sided heart structures because of twice passed tricuspid valve replacement.

CLINICAL SUMMARY

The patient is a 66-year-old woman with rheumatic heart disease who underwent tricuspid valve replacement with a bioprosthesis (Hancock Medtronic, 27 mm; Medtronic Inc, Minneapolis, Minn) at 25 years of age. She underwent reoperation at 39 years of age, and the calcified tricuspid bioprosthesis was replaced with a new bioprosthesis (Hancock Medtronic, 29 mm) plus mitral commissurotomy. Six years later, she underwent percutaneous mitral valvuloplasty for recurrence of mitral stenosis. At that time, oral anticoagulation was begun for the onset of permanent atrial fibrillation.

During the past 4 years, her clinical condition worsened with shortness of breath and several hospital admissions for congestive heart failure despite optimal medical treatment. Transthoracic echocardiography revealed normal left ventricular systolic function, left and right atrium dilatation, a calcified tricuspid bioprosthesis with moderate stenosis (mean gradient 11 mm Hg), severe regurgitation, moderate mitral valve stenosis (mean gradient, 6 mm Hg), severe regurgitation, and moderate aortic valve stenosis (valve area, 0.66 cm²/m²).

A decision was made to replace the tricuspid bioprosthesis and the mitral and aortic valves. The European System for Cardiac Operative Risk Evaluation I was 31.55%, and European System for Cardiac Operative Risk Evaluation II was 36.73%. The patient underwent conventional cardiac surgery through a median sternotomy, CPB, and

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