Multiple papillary fibroelastoma originating from 3-leaflets of the aortic valve

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Informed Consent Statements

Written patient’s informed consent was obtained.

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Glossary of Abbreviations

LCL; left coronary leaflet
NCL; non-coronary leaflet
PFE: papillary fibroelastoma
RCL; right coronary leaflet

Central Message
We report a successful surgical removal of a papillary fibroelastoma originating from 3-leaflets of the aortic valve.

Central picture legend
Papillary fibroelastoma originating from the 3-leaflets of the aortic valve.
Cardiac papillary fibroelastoma (PFE) is a rare, but not uncommon, benign cardiac tumor that occurs mainly on the valvular endocardium. We report an extremely rare case of an 81-year-old woman, who underwent surgical removal of 3 masses arising simultaneously from each leaflet of the aortic valve; which were confirmed as cardiac PFEs. Ethics review is not required for case report implementation at our institution. Written informed consent for publication was signed by the patient.

Case report
An 81-year-old woman was referred to our department for surgical intervention of aortic valve attached mass, discovered incidentally during pre-operative assessment for cataract surgery. The most recent echocardiography checkup revealed a mass, increasing in size during the last 3 months, suspicious of a cardiac tumor.

Preoperative transthoracic echocardiography showed no aortic stenosis, trace aortic insufficiency and a relatively luminous cordiform, mobile tumor of 15 mm on the left coronary leaflet (LCL) toward the sinus of Valsalva. (Figure 1A) Attachment of a relatively high luminosity structure was suspected on the right coronary leaflet (RCL) and non-coronary leaflet (NCL) as well. No obvious anomalous structures were observed adhering to the mitral, tricuspid, or pulmonary valves. Other than the imaging tests, the patient showed no abnormal physical findings or laboratory data.

Although asymptomatic, the patient was scheduled for surgery to prevent embolism. The surgery was performed through a median sternotomy. We initiated cardiopulmonary bypass with
ascending aortic cannulation and bi-caval venous drainage. Upon transverse aortotomy, observation of the aortic valve revealed a stalked joint mass at the Arantius body of the NCL, RCL and LCL. (Figure 1B1-3, Video 1) The apex of each mass was covered with fine hair-like fibers, visually reminiscent of a "sea anemone". Each mass was removed without injuring the aortic valve leaflet. The remainder of the aortic valve and sinus of Valsalva appeared grossly normal. The sinus of Valsalva was filled with saline to ensure that there was no obvious regurgitation, and the aortotomy was closed.

The specimens removed were each approximately 10 mm long, and were all whitish, stalked papillary lesions. (Figure 2A) Postoperative tumor histopathology (Figure 2B-D) revealed, that the tumors were fibrous connective tissue with branching and eosinophilic unstructured deposits, covered by a single layer of squamous epithelium, which is compatible with cardiac PFE.

The patient's postoperative course was uneventful and postoperative echocardiography also revealed favorable results without evidence of any residual tumor on the aortic valve and aortic insufficiency. The patient was discharged 12 days after surgery. The 1- year postoperative follow up was also uneventful, and we are now carefully monitoring the progress.

**Discussion**

PFE is a rare benign cardiac tumor. However, with the development of imaging techniques such as echocardiography, incidental discovery in previously asymptomatic patients is very likely. [1] Cardiac PFE occurs most commonly on the cardiac valves (78%) and is predominantly located on the aortic valve. While most PFEs are solitary, according to the Mayo Clinic series, 21% of resected cases were reported to be multiple (range 2 to 40). [2] However, multiple lesions in the same valve are extremely rare and few cases have been reported previously. [3] In the present case, we report
the case of a patient who had 3 masses arising simultaneously from each leaflet of aortic valve, which were then confirmed as cardiac PFEs. Each tumor was removed by direct excision, preserving the native aortic valve.

There are several hypotheses about the cause of PFE formation, but none have been scientifically proven. Whether they are reactive growths, hamartomatous processes, or neoplastic lesions remains unknown. However, their propensity to develop on damaged endocardial surfaces seems to support the notion that they are reactive growths. [4] Given that PFE occurs at the contact surfaces of the three leaflets, reactive growth may be a possible cause due to continuous endothelial damage on valvular surfaces.

In patients with echocardiographically suspected PFE, who do not undergo surgical removal, rates of cerebrovascular accident and mortality are thought to be increased; [2] therefore operative indication was considered reasonable for this asymptomatic elderly woman. For multiple lesions surgery, Touati et al. warned of the existence of unresected "hidden tumors"; [3] and although the abovementioned Mayo Clinic series reported 1.6% recurrence after the tumor removal, [2] the most recent data from the Mayo Clinic showed that 12% had a recurrence of PFE, with the first occurrence being an aortic valve. [5] Needless to say, continued follow-up is necessary for this benign tumor. Furthermore, it may be necessary to reconsider surgical strategies for similar cases in the future.

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References


Figure legends

Figure 1

(A) Preoperative transthoracic echocardiography shows a relatively luminous, cordiform, mobile tumor of 15 mm (red arrowhead) attached to the aortic valve.

(B) Surgical view through aortotomy demonstrates tumor on each coronary leaflet (blue arrowhead). 1: NCL, 2: RCL, 3: LCL, *; caudal side, **; cranial side, NCL; non-coronary leaflet, RCL; right coronary leaflet, LCL; left coronary leaflet.

Figure 2

(A) Macroscopic image of the tumor, characteristic gelatinous appearance of papillary fibroelastoma, looked like “sea anemone” shape.

(B-D) Hematoxylin and eosin stains of the tumor. (B) x 20; histologically, the tumor consists of narrow papillary fronds showing complex branching patterns (C) x 200, (D) x 400; avascular fibroelastic tissue lined by a single layer of endocardium.

Video legend

Video 1

Head camera video from left side of patient; PFEs attaching to NCL, RCL, and LCL, in sequence.

PFE; papillary fibroelastoma, NCL; non-coronary leaflet, RCL; right coronary leaflet, LCL; left coronary leaflet.
Figure 1

A

B