Letter to the Editor

Coinciding anomalous coronary artery and papillary fibroelastoma

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The primary cardiac tumors are extremely rare tumors that arise from the normal cardiac tissues. There are benign variants (mostly myxoma) and malignant tumors. Fibroelastoma (FE) is the most common primary tumor, of which myxoma is the most common subtype followed by papillary fibroelastomas (PFE). They account for the majority of primary heart valvular tumors that mainly involve the left (mitral and aortic) valves, and only few cases reported pulmonary valve PFE.[1] We present a case of 61 year old female who had incidental pulmonary valve papillary FE finding on echo along with anomalous coronary arteries.

A 61 years old diabetic female with a medical history of sarcoidosis presented with chronic stable angina. She underwent an exercise stress test in which the patient exercised 7 min of a modified Bruce protocol was terminated afterwards due to leg pain but not chest pain. Her ECG demonstrated ST segment changes consistent with ischemia (ST segment depression of more than 1 mm in lead II,III, aVF along with lead V5 and V6). The coronary arteries were found to be angiographically clean during left heart catheterization but the procedure showed an anomalous large, long left main coronary artery originating from the right coronary cusp. Cardiac computed tomography (CT) angiogram confirmed the anomalous origin of the left main coronary artery from a common ostium with the right coronary artery originating from the right coronary cusp. Cardiac computed tomography (CT) angiogram confirmed the anomalous origin of the left main coronary artery from a common ostium with the right coronary artery originating from the right sinus of Valsalva (Figure 1A and 1B). The left main coronary was crossing between the aorta and right pulmonary outflow tract consistent with an intra-arterial course. A Trans-Thoracic It also showed pulmonary valve FE (Figure 2). Echocardiogram showed an incidental finding of multiple mobile structures attached to the pulmonary valve, with the largest one measuring 0.7 × 0.7 cm, whose appearance was consistent with PFE. A Trans-esophageal echo showed a mobile mass attached to the pulmonary valve measured 0.5 × 0.6 cm (Figure 3 and Figure 4).

Given the anginal chest pain that was refractory to optimal medical management and because of the concomitant congenital coronary anomaly cardio-thoracic surgery was consulted and did not recommend relocating the left main coronary artery would not provide symptomatic relief due to the common ostia with the right and its intra-arterial course. After multi-disciplinary discussions among interventional cardiology, cardiothoracic surgery teams, the patient and her family, she underwent 2 vessel coronary artery revascularization (left internal mammary artery to left anterior descending artery, reverse saphenous vein graft to ramus with endoscopic vein harvest and excision of pulmonary valve mass). She had an uneventful post-operative course and was discharged after four cardiac rehabilitation, general cardiology, and cardiothoracic surgery clinic follow ups. The subsequent pathology report confirmed the diagnosis of PFE.

The case we present is unique given the coexistence of two rare cardiac conditions within the same patient. First, PFE is rare in general and even less common in the pulmonary valve or generally in the right side of the heart. An example of the rarity of this pathology is a 41 year single tertiary center experience from Austria that revealed only 12 cases of FE, of which only one arising from the right side of the heart (tricuspid).[2] In addition, the patient had an anomalous left coronary artery with common ostia with the right coronary artery and acute angle then took a septal course to the diagonal branch to the left anterior descending artery and the circumflex which is a very rare variant.

It was believed that the anginal pain in this patient was due to her anomalous coronary rather than the pulmonary valve FE as the first has been associated with ischemic events and sudden death, whereas other anomalies with a right-sided origin are considered to be harmless.[1]

In our case the FE was facing the pulmonary side and

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Figure 1. Cardiac computed tomography. (A): Anomalous origin of the left coronary artery arising with the right main coronary artery (arrow); (B): Top arrow: the left coronary artery coursing between the aorta and right ventricular outflow tract. Bottom arrow: typical location where the left coronary artery should arise from.

Figure 2. Cardiac computed tomography showing pulmonary valve fibroelastoma (arrow).

Figure 3. Trans-esophageal echo picture showing right ventricular outflow tract with pulmonary valve with a nodule in the pulmonary valve (Arrow).

Figure 4. Trans-esophageal echo picture showing pulmonary valve cusp with 5X6 mm nodule representing the fibroelastoma (Arrow).

was on the anterior pulmonary valve leaflet. In addition, our patient was diagnosed with sarcoidosis, for which, to our knowledge, there were no reported cases of sarcoidosis with pulmonary valve FE. Given that the pulmonary valve FE was mobile (which is the only independent predictor of papillary FE related death or nonfatal embolization) surgical removal was appropriate. Embolization of left-sided and tricuspid valve tumors has been well documented. By comparison, the first case of pulmonary embolization of a PFE arising from the pulmonary valve was reported in 2008. Having been timely diagnosed and appropriately surgically treated, the patient showed great improvement, and resolution of the symptoms. Recurrence after surgical resection has not been reported.

In addition to the extreme rarity of both these clinical entities this case raises the question about the possible underlying mechanism where disruption of normal connective tissue architecture and/or remodeling was the main etiology. Our case
models that in the triad of congenitally anomalous left coronary artery, PFE (of undetermined timing), and sarcoidosis.

Disclaimer

An abstract of this case was submitted to the Southern Medical Association (SMA) Annual Scientific Assembly (2014); a summary can be found in the Journal of Investigative Medicine.[4]

References