Papillary fibroelastoma of the aortic valve

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Cardiac papillary fibroelastomas (CPFs) are benign primary cardiac tumors and are most often found on the downstream side of cardiac valves [1–3]. The most often affected valve is aortic, but these tumors could implicate all valves. The clinical presentation varies from asymptomatic to embolic complications, which can lead to cerebral stroke, myocardial infarction, and sudden cardiac death [1, 2, 4, 5]. In approximately 30% of cases, they are found incidentally, during autopsies, echocardiography, or cardiac surgery [2, 5].

A 34-year-old male was referred to the cardiology department for further evaluation due to dizziness and blinking in the eyes. Neurological examinations (carotid ultrasound imaging, transcranial color-coded duplex ultrasonography, and head computed tomography) were without abnormalities. Transthoracic echocardiography revealed a 0.9 × 1.2 cm oval-shaped mobile mass attached to the left coronary cusp of the tricuspid aortic valve (Figure 1A–B, the arrows). The mass did not cause any outflow tract obstruction or aortic regurgitation. A contrast-enhanced computed tomography scan of the chest confirmed the presence of a hypodense nodular lesion of approximate size of 1.1 × 1.4 × 1.2 cm in the aortic root, adjacent to the origin of the right and left coronary arteries (Figure 1C, the arrow). Based upon the above findings, a differential diagnosis was made which included: papillary fibroelastoma, myxoma, thrombus, and inflammatory mass. A coronary computed tomography angiography was normal. The decision was made to perform an excision of the tumor under cardio-pulmonary bypass. The valve-sparing surgery was done. At the time of surgery, a gel-like tumor of 1.2 × 1.0 × 1.0 cm was found (Figure 1D). The tumor with a pedicle was attached at the base of the commissure between the right and left coronary cusps of the aortic valve.

The pathological analysis confirmed the nature of the mass and revealed multiple, branching fronds of paucicellular, avascular fibroelastic tissue lined by a single layer of the endocardium (Figure 1E–F). The postoperative period was complicated by hemorrhagic anemia and bacterial infection. The patient was discharged after 13 days. There was no recurrence seen on an echocardiogram 6 months after surgery.

The pathogenesis and risk factors of CPFs are unclear. Cardiac papillary fibroelastomas could be diagnosed at any age but most commonly occurs in middle-aged and older adults. Echocardiography is the principal diagnostic examination. Transesophageal echocardiography is more sensitive compared with transthoracic due to typically small sizes of these tumors and their attachment to the endocardial surface. Multimodality imaging (computed tomography and magnetic resonance angiography) could be helpful for differential diagnosis. A biopsy of the tumor is not usually needed. The treatment of CPFs is not clearly defined by guidelines. The surgery is recommended for larger than 1 cm left-sided papillary fibroelastomas, and it reduces the risk of thromboembolic complications [5]. CPFs can be safely excised with preservation of the native valve in experienced surgical centers [4]. Tamin et al. [4] demonstrated that the risk-to-benefit ratio of cardiac surgery is influenced by older age, comorbidities, and perhaps the uncertainty of embolic risk.

Usually, surgical resection is safe and has low perioperative mortality, and it is associated with perfect long-term outcomes [4, 5].

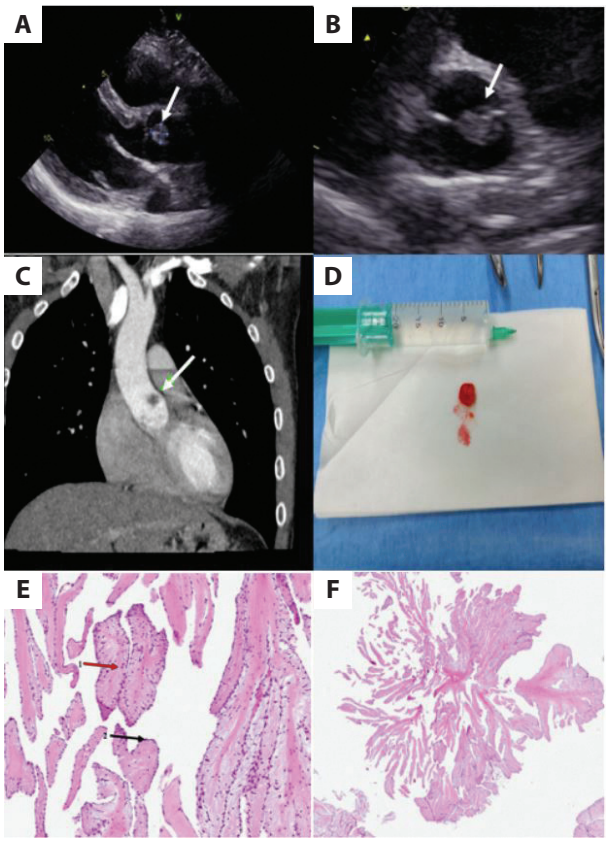

Figure 1. Papillary fibroelastoma of the aortic valve seen on transthoracic echocardiography (**A, B**) and contrast-enhanced computed tomography images (**C**) (the arrows). The microscopic image of the excised mass (**D**) and histo-anatomopathological analysis of the mass (**E–F**, the red — stroma, the black arrow — endocardium)

Article information

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