An unusual case of localization of papillary fibroelastoma on the upstream side of the tricuspid valve

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Accepted: January 17, 2023 Early publication date: January 29, 2023 A 73-year-old woman was hospitalized at the Clinic for Gastroenterology for acute pancreatitis manifested by abdominal pain, nausea, and vomiting. During hospitalization, transthoracic echocardiography (TTE) was performed showing a round soft tissue formation in the right side of the heart. The patient denied having any heart problems earlier. The TTE test performed at our Institute of Cardiovascular Diseases (Figures 1A, B; Supplementary material, Video S1, Figure S1) confirmed the presence of a soft tissue formation in the right side of the heart, which gave the impression of being attached to the interventricular septum by a small pedicle, directly below the septal cusp of the tricuspid valve (TV), 14×15 mm in size, and floating freely. The structure was regular and round. In the differential diagnosis, it resembled a benign tumor rather than a thrombus or endocarditis. Subsequently, computed tomography (CT) of the heart was performed (Figure 1C) showing an oval hypodense change $(13 \times 13 \times 15 \text{ mm})$ in the subvalvular segment, attached to the interventricular septum by a small pedicle. The described change corresponded to a fibroma or fibroelastoma of the right ventricle. Based on multi-imaging diagnostics, the Heart Team recommended surgical removal of the tumor.

Transesophageal echocardiography (TEE) in the operating room (Figure 1D, Supplementary material, *Video S2*) established the presence of a round hyperechoic mass, 17 × 15 mm in size, above the septal cusp of the TV. This result did not correspond with TTE and CT findings, which indicated that the change is related to the pedicle below the TV. The operation was performed under general endotracheal anesthesia through total median sternotomy. After opening the right atrium, a tumor was identified in the atrium, which was attached to the middle part of the septal cusp of the TV by a pedicle (Figure 1E, Supplementary material, *Video S3*). The tumor was completely resected. The postoperative course was uneventful. Microscopic analysis of the tumor revealed typical features and confirmed the diagnosis of papillary fibroelastoma.

The presence of an unexpected mass in the heart undoubtedly presents a diagnostic dilemma. In this case, we had no dilemma because the structure was presented as a regular round mass, attached by a pedicle. In contrast to the tumor, a thrombus is irregularly shaped and most commonly has no pedicle [1]. Vegetations characteristic of infective endocarditis are typically irregularly shaped. Primary cardiac tumors are rare, with an incidence rate of 0.0017%–0.019% in the autopsy series [2]. Papillary fibroelastomas account for less than 10% of primary cardiac tumors [3, 4]. They are mostly asymptomatic and incidentally diagnosed by high-resolution imaging methods. Even if benign, a tumor can cause embolic complications, acute myocardial infarction, syncope, ventricular arrhythmias, and sudden cardiac death [3]. They occur most frequently on valves, mainly the aortic and mitral valves, and much less commonly on the pulmonary and TV valves. Moreover, having in mind that papillary fibroelastomas are typically found on the downstream side of the valves [5], our case was rather unique as the tumor was located on the upstream side of the TV.

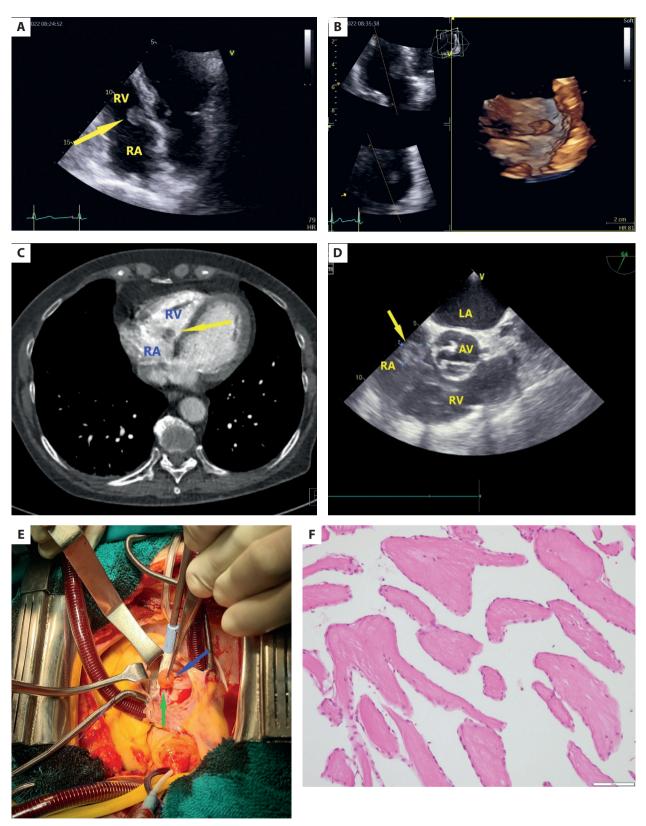


Figure 1. A. Two-dimensional transthoracic echocardiography showing a regular, round tumor (yellow arrow) that appeared to be attached to the interventricular septum by a small pedicle, just below the tricuspid valve. **B.** Three-dimensional reconstruction of the tumor by transthoracic echocardiography. **C.** Computed tomography of the heart showed an oval hypodense change (yellow arrow) that appeared to be located subvalvularly and seemed to be attached to the interventricular septum by a small pedicle. **D.** Preoperative transesophageal echocardiographic image showing a tumor (yellow arrow) in the right atrium. **E.** The surgical view after opening the right atrium showed a tumor (blue arrow) in the right atrium attached to the upstream side of the septal cusp of the tricuspid valve by a small pedicle (green arrow). **F.** Hematoxylin-eosin staining (H&E, 20×) showed the branching avascular papilla, composed of the hyalinized collagen matrix, which were covered by the endothelium

Abbreviations: AV, aortic valve; LA, left atrium; RA, right atrium; RV, right ventricle

Supplementary material

Supplementary material is available at https://journals. viamedica.pl/kardiologia_polska.

Article information

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