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Three-dimensional transesophageal echocardiography guided endomyocardial biopsy in diagnosis of cardiac tumor

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Primary malignant cardiac tumors are extremely uncommon (< 0.3% of cardiac tumors in postmortem studies) and are associated with poor prognosis [1, 2]. Therefore, to increase the survival rate, an early and effective diagnostic process is necessary. Although noninvasive imaging modalities are useful, a definite diagnosis in most cases requires histologic examination, which remains a gold standard. Reported herein, is a patient with new-onset heart failure (HF), in which three-dimensional (3D) transesophageal echocardiography (TEE)-guided endomyocardial biopsy (EMB) confirmed the diagnosis of cardiac angiosarcoma.

The 73-year-old female with no relevant past medical history was admitted to the hospital due to signs of HF de novo. On admission, she presented shortness of breath upon exertion (New York Heart Association class II); heart rate, 90 beats/min; blood pressure, 95/78 mmHg; systolic-diastolic murmur on cardiac auscultation; general peripheral edema; signs of bilateral pleural effusion and liver enlargement. Laboratory studies showed elevated cancer biomarker: Ca-125, 290 U/mL (N: < 35 U/mL).

Transthoracic echocardiography (TTE) and TEE revealed a large mass with heterogeneous echogenicity almost completely filling the cavity of the right atrium (RA), infiltrating its wall, the visceral pericardium of the right ventricle and the apex of the heart causing severe obstruction of the inflow from both venae cavae with a mean gradient difference of 7 mmHg (Fig. 1).

Chest, abdomen, and pelvis contrast-enhanced computed tomography confirmed a large polycyclic tumor (88 × 67 × 74 mm) with heterogeneous densities in the RA and its surroundings. Cardiac magnetic resonance (CMR) was not performed on this patient because of kidney insufficiency.

As the primary differential diagnosis suggested a primary or secondary malignant cardiac tumor, a TEE-guided EMB from the access point of the right internal jugular vein was performed. Biopsy forceps (Cordis Corp) were inserted into a 7F (Cordis Corp) long sheath and advanced into the RA. The TEE allowed detailed visualization of the tumor, bioptome position on the tumor surface, and exact selection of the sample site. Periprocedural imaging was further enhanced with the use of 3D imaging (Fig. 1). Ten tissue samples were gathered from the tumor surface without periprocedural complications. Histochemistry, immunohistochemistry, and electron microscopy study of the EMB specimens revealed malignant vascular tumor features (Fig. 2).

The diagnostic and therapeutic approach of cardiac tumors is very demanding because of the various clinical presentation and intracardiac localization [3]. An early diagnosis of angiosarcoma is crucial for managing therapeutic options and the patient's prognosis.

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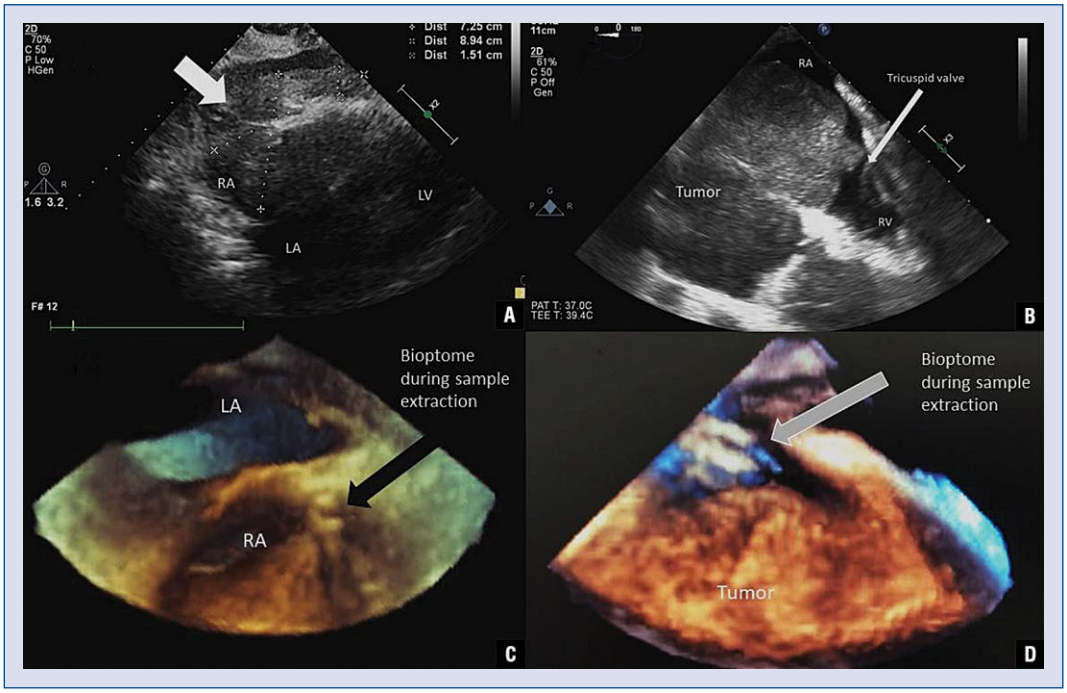

Figure 1. Imaging of the tumor; **A.** Large tumor with heterogeneous echogenicity (shown with a white arrow) infiltrating the wall of the right atrium (RA) and filling its cavity almost completely, visible from the substernal view in transthoracic echocardiography; **B.** Visualization of relation between tumor and tricuspid valve in transesophageal echocardiography (TEE); **C, D.** Three-dimensional TEE imaging showing exact localization of bioptome during extraction of tumor tissue samples; LA — left atrium; LV — left ventricle; RV — right ventricle.

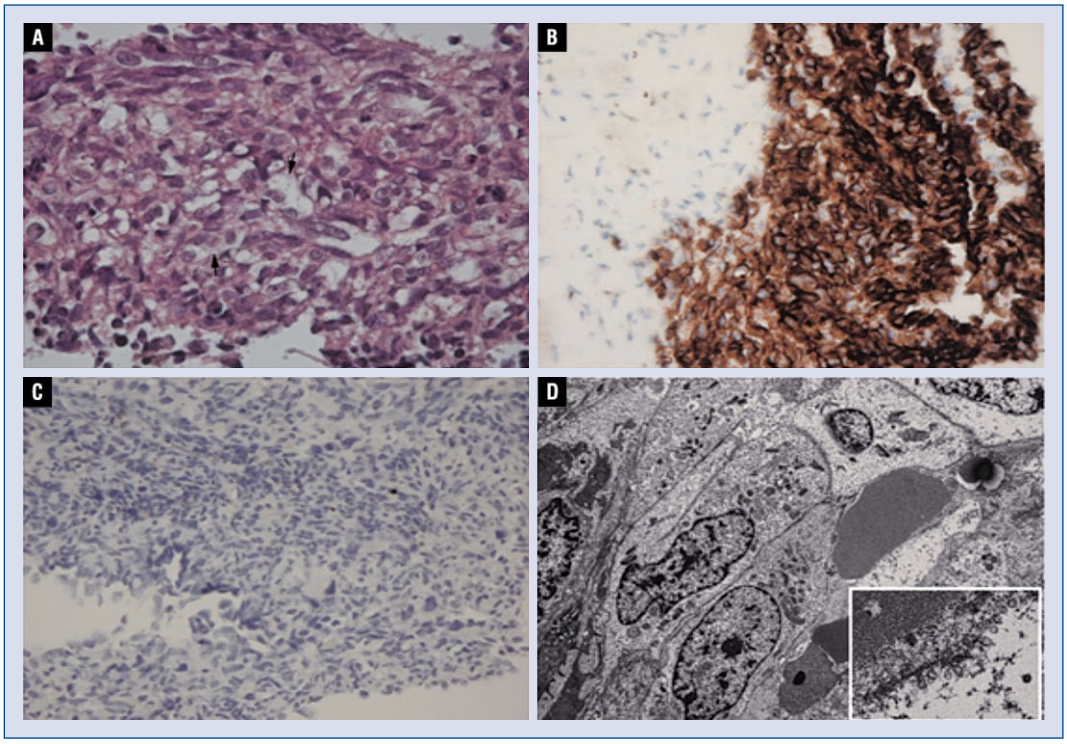

Figure 2. Primary cardiac angiosarcoma; **A.** Anaplastic cells with poorly formed vascular channels (arrows) (hematoxylin and eosin); **B.** Strong immunohistochemical staining for CD31 marker (brown color); **C.** Negative immunohistochemical staining for cytokeratin filaments AE1/AE3 (brown color); **D.** Electron micrographs showing immature endothelial cells.

Comprehensive clinical and multimodality imaging evaluation of cardiac tumors, including echocardiography, contrast enhanced computed tomography and CMR, is fundamental to obtain a proper initial differential diagnosis [4]. Angiosarcomas are mostly immobile and broad-based with endocardial to myocardial growth, however not all tumors infiltrate surrounding tissue [1]. CMR findings in angiosarcomas include heterogeneous T1- and T2-weighted signal intensity and a heterogeneous contrast enhancement pattern [1]. A position emission tomography scan with the use of 18F-2-fluoro-2-deoxy-D-glucose (FDG) can reveal areas of high FDG uptake within the mass and evidence of metastatic disease [5, 6].

Histopathological confirmation is needed for chemotherapy initiation and assessment of prognosis. In some cases, samples are obtained during cardiac surgery that is performed to remove the tumor but in patients disqualified from surgical treatment EMB is the only way to obtain tissue samples and a definite diagnosis. Despite a very low complication rate (< 1%), it is not commonly performed [7, 8]. EMB using broad histologic and immunohistochemical methods allows for the definition of the type of tumor, management of the treatment methods, and better risk stratification. Echocardiography-, electroanatomic mapping- or in the future, CMR-guided EMB increases the accuracy and safety of the procedure [9]. In the present case, TEE guidance allowed direct visualization of the tumor and bioptome position on its surface. The procedure proved that biopsy forceps, when guided by TEE, are feasible for the diagnosis of intracardiac tumors. The use of 3D TEE indicated the place of sample collection and minimized the risk of complications. 3D visualization ensures permanent visualization of the biopsied tissue in a pumping heart. This was crucial since the RA cavity was to a great extent occupied with the tumor and the targeted mass was in close proximity to the atrial septum and tricuspid valve leaflets. To increase the diagnostic accuracy and sampling error, it is necessary to gather at least five tissue samples (recommended 5–10), each 1–2 mm in size. The main limitation of EMB is a need for experienced physicians and histopathologists; thus, patients should be referred to tertiary medical centers.

In terms of treatment, angiosarcomas, when localized without infiltration of adjacent structures, may undergo surgery with or without upfront chemoradiotherapy. The majority of angiosarcomas have a poor overall prognosis related to EMB--targeted therapeutic management and complete surgical resection of the tumor.

To conclude, this case report showed that EMB guided by TEE might increase the accuracy and safety of the procedure and enable a definite diagnosis of the type of intracardiac tumor, facilitating further management.

Ethical statement

The authors are accountable for all aspects of the work and ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

Conflict of interest: None declared

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