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SUPERIOR VENA CAVA SYNDROME ASSOCIATED WITH METASTATIC ADENOCARCINOMA TO THE HEART: THE DIAGNOSTIC UTILITY OF CARDIAC RESONANCE IMAGING

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Case Report

An 82-year-old female with no significant medical history was transferred to our hospital for evaluation of an anterior mediastinal mass identified by chest computed tomography (CT) (Figure 1). This mass seemed to extend from the right atrium to the right ventricle. Also, it was inseparable from the anterior border of the anterior wall of the superior vena cava. She had symptoms consistent with superior vena cava syndrome, with recent onset of shortness of breath and facial and bilateral upper-extremity edema.

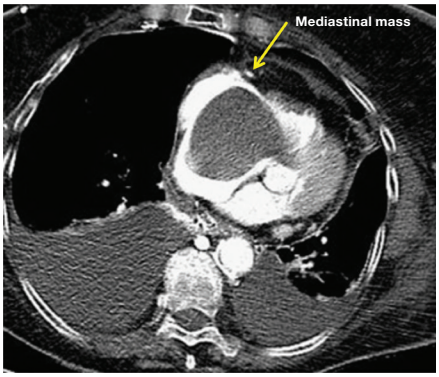

Figure 1. Computed tomography (CT) of chest showing a multilobulated hypoenhancing mass in the anterior mediastinum measuring 6.5 x 5.3 x 3.8 cm. This mass was inseparable from the anterior wall of the superior vena cava.

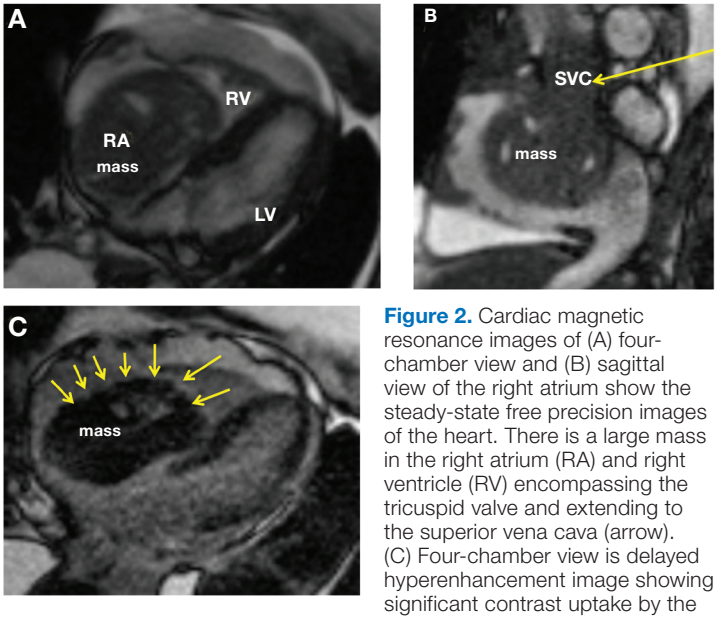

Figure 2. Cardiac magnetic resonance images of (A) four-chamber view and (B) sagittal view of the right atrium show the steady-state free precision images of the heart. There is a large mass in the right atrium (RA) and right ventricle (RV) encompassing the tricuspid valve and extending to the superior vena cava (arrow). (C) Four-chamber view is delayed hyperenhancement image showing significant contrast uptake by the mass.

At this stage of the workup, it was unclear whether this was a tumor or a thrombus, therefore a cardiac magnetic resonance imaging (CMR) was performed to assess tissue characterization of the mass and to delineate its intravascular extension. The CMR showed a large, well-circumscribed, heterogeneous mass in the right atrium (RA) extending into the right ventricle and also extending to and completely obliterating the superior vena cava (Figure 2; *to access a video of supplemental CMR cine images, visit www.debakeyheartcenter.com/journal/video*). This was suspicious for a primary or metastatic tumor of the heart. Transesophageal echocardiography (TEE) was performed using a three-dimensional (3D) matrix probe during percutaneous biopsy of the mass. The use of a 3D imaging probe allowed for simultaneous biplane imaging to assist with identification of the catheter-based bioptome and positioning within the mass (Figure 3).

A tissue biopsy of the RA mass showed sheets of markedly atypical cells with large vesicular nuclei, prominent nucleoli, and brisk mitotic activity admixed with lymphoplasmacytic infiltrate. Gland formation and papillary configuration were also noted. These features are consistent with moderately differentiated adenocarcinoma (Figure 4). Cytokeratin 20, TTF-1, napsin A, CD31,

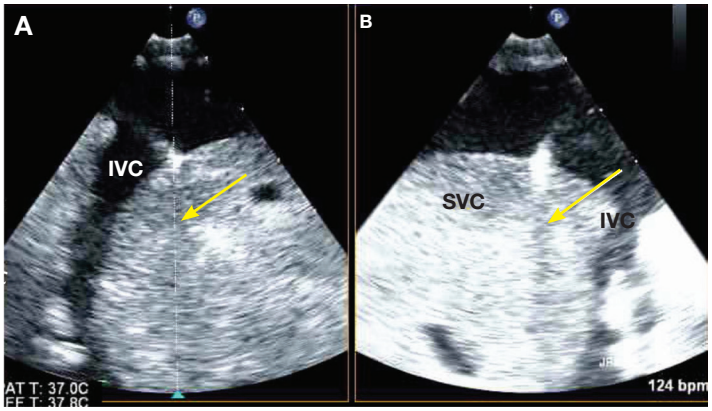

Figure 3. A) Transesophageal-guided biopsy of the mass in right atrium. (B) X-plane image of the right atrium mass during the biopsy by biotome (white arrow).

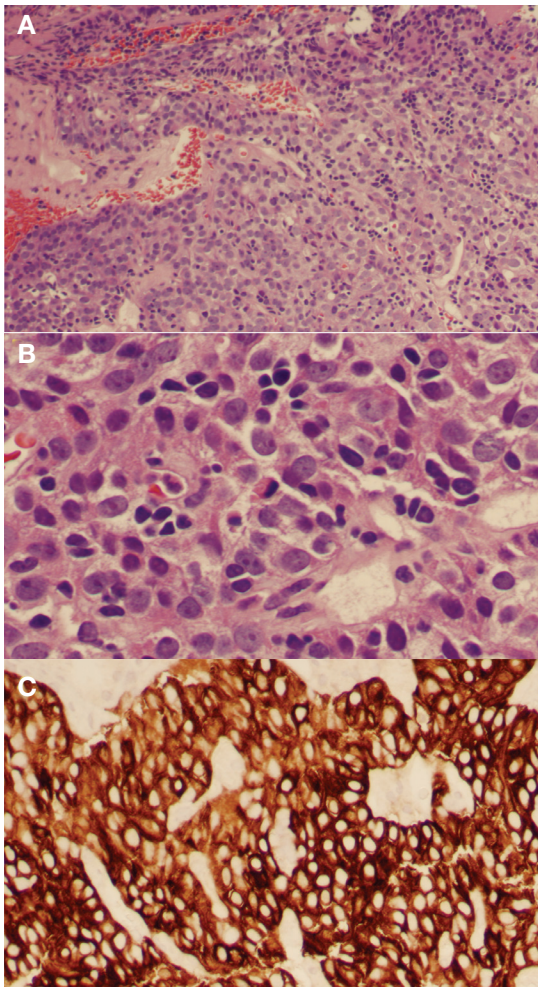

Figure 4. (A) Histologic section shows infiltrating nests of atypical epithelioid cells admixed with lymphocytic infiltrate (H and E stain, x40). (B) High-power magnification shows atypical cells with large vesicular nuclei and prominent nucleoli. Gland formation is focally seen (center) (H and E stain, x400). (C) Pan CK: Immunohistochemical stain for pan-cytokeratin (OSCAR) is diffusely and strongly positive for tumor cells.

and mammaglobin stains were negative, making the lung an unlikely source for the tumor. Staining data was inconclusive, and the primary source of the tumor could not be determined.

A CT of the abdomen and pelvis did not show any suspicious origin of this tumor. A dedicated PET scan could not be performed as the patient opted for hospice.

Cardiac tumors are a rare occurrence, and three quarters are benign. Among malignant tumors, metastatic tumors are more common than primary cardiac tumors.[1,2] The incidence of metastatic tumor by one report is up to 10%.[3] The ones that do metastasize to the heart are melanomas, leukemias/lymphomas, or solid tumors from the breast, lung, or esophagus.[1] In the absence of an obvious primary tumor site or history of previous malignancy, diagnosis can become challenging. We present a rather rare case of adenocarcinoma of unknown primary origin to the right atrium and right ventricle presenting as superior vena cava syndrome. This case highlights how a multimodality imaging approach can be used to provide broad tissue characterization of the mass and to facilitate percutaneous tissue biopsy for a definitive diagnosis. The patient elected not to undergo any further treatment.

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Keywords: superior vena cava syndrome, metastatic adenocarcinoma

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