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Clinical Case Report

Primary cardiac epithelioid angiosarcoma with frond-like features: a rare and ominous radiological mimicker of benign cardiac tumors

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ABSTRACT

Most primary cardiac tumors are benign neoplasms, which generally can be differentiated from malignant neoplasms via certain radiological features. We present briefly a case of a 26-year-old man undergoing resection of a right atrial mass that based on preceding radiologic findings represent a myxoma. After pathologic examination, the lesion was determined to be an epithelioid angiosarcoma with unique frond-like architecture and multiple pedicular attachments to the atrial wall.

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1. Introduction

Angiosarcoma is the most common primary malignant differentiated neoplasm to arise from the heart [1,2]. Radiological modalities — commonly echocardiogram, but also including magnetic resonance imaging (MRI) and computed tomography (CT) — provide recognition of features that favor the presence of a benign versus malignant neoplasm [1,3].

2. Case report

A 26-year-old Caucasian man with no significant preceding medical history originally presented to an outside institution 1 month prior with a pericardial effusion. Echocardiogram performed on a follow-up clinic visit showed a right atrial heteroechoic mass attached by a stalk that prolapsed across the tricuspid valve into the right ventricle in addition to a persistent pericardial effusion: the patient was immediately transferred to our institution. Cardiac MRI noted the pericardial effusion and the mobile mass attached to the posterior inferior right atrium prolapsing through the tricuspid valve during diastole (Fig. 1A–C). Intraoperative transesophageal echocardiogram (TEE) additionally noted that mass was irregular in shape and echogenic in texture, with multiple pedunculated mobile elements (Fig. 1D).

In the operating room, a hemorrhagic pericardial effusion was first drained (cytopathologic examination showed no evidence of malignancy). The right atrium was opened, and the atrial wall was resected entirely where the mass was attached and apparently infiltrated. The surgery was completed without complication.

Pathologic gross examination noted a 10.5×6.0×4.0-cm friable, frond-like mass, almost papillary in appearance, attached to the atrial wall via narrowed pedicles (Fig. 1 E–F). Upon serial sectioning, the mass's frond-like projections were pale tan, with the lesion toward and involving the atrial wall exhibiting a hemorrhagic appearance with foci of necrosis. Detailed histopathologic features are described in the caption of Fig. 1 (Fig. 1G–M). Immunoperoxidase staining revealed that the neoplastic cells expressed ERG, CD31, and CD34 and were negative for D2-40, HHV8, and cytokeratin AE1/AE3 (Fig. 1N–Q). Unfortunately, there was evidence of a focally positive surgical resection margin (Fig. 1F grossly and Fig. 1G–H microscopically). The morphologic and immunohistochemical features supported a diagnosis of an epithelioid angiosarcoma.

Postoperative course was unremarkable, and the patient would be discharged 4 days after surgery. Staging positron emission tomography (PET–CT) was performed 2 weeks postsurgery and was notable for mildly hypermetabolic subpleural nodular opacities bilaterally at the lung bases. The patient is at the time of submission of this manuscript 2months postsurgery and clinically stable; however, he has yet to

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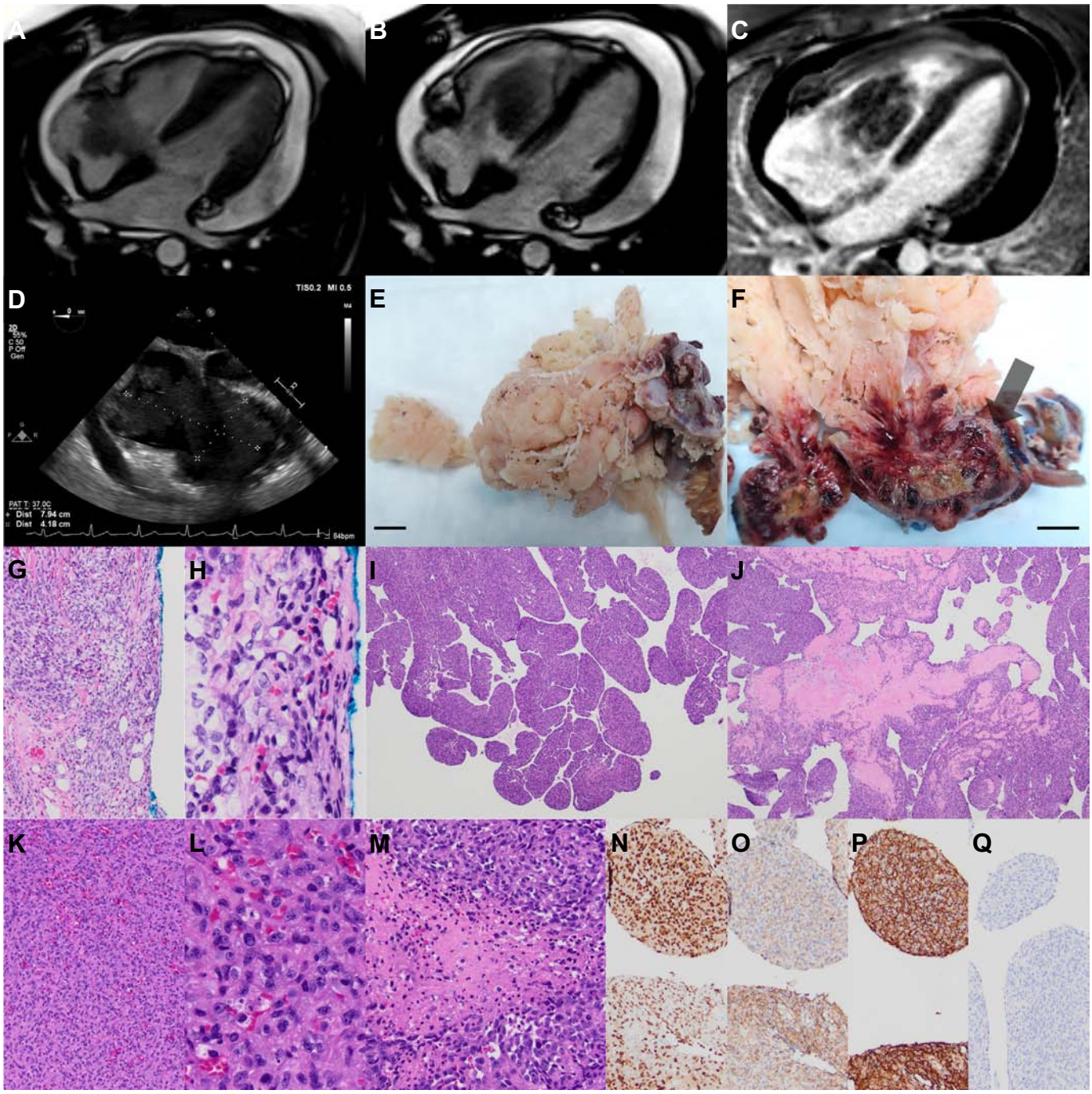

Fig. 1. Cardiac MRI revealed the frond-like mass attached to the posterior wall of the right atrium that exhibited mobility from the right atrium (A, systole) into the right ventricle (B, diastole) (A and B are balanced steady-state free precession images). A delayed contrast-enhanced four-chamber image shows the heterogeneous enhancement associated with the mass (C). Intraoperative TEE similarly revealed the mobile, irregularly shaped mass entering into the right ventricle during diastole (D). Radiologic studies also noted a pericardial effusion (A–D). The gross appearance of the resected mass was congruent with its frond-like radiologic character, with most of the mass upon sectioning appearing pale tan (E), and the lesion infiltrated the atrial wall and exhibited a hemorrhagic appearance with foci of necrosis (F) (bar scale for E and F equals 1 cm). The neoplasm was focally involving the blue-inked surgical resection margin grossly (F, indicated with arrow) and microscopically (G and H). Note the distinct pedicles by which the neoplasm attaches to the atrial wall (F). The pale-tan fronds corresponded to branches of the neoplasm that were cellular (I) or with a collagenous core (J). Sections from the pedicles and atrial wall reveal a dense, infiltrative neoplasm characterized by epithelioid cellular morphology and high mitotic activity, and with slit-like spaces containing erythrocytes (K–L). Focal tumor necrosis was also evident (M). Immunohistochemistry showed that the neoplastic cells expressed ERG (N), CD31 (O), and CD34 (P) and were negative for cytokeratin AE1/AE3 (Q). Original magnification for I and J at ×40; for G, K, and N–Q at ×100; for M at ×200; and for H and L at ×400.

begin planned radiation therapy and weekly paclitaxel secondary to a lack of medical insurance coverage.

3. Discussion

Primary cardiac angiosarcoma can be typically differentiated from benign neoplasms via radiological modalities [1]. Angiosarcomas are usually found in the right atrium, infiltrative and/or protruding into the chamber, and broad based and not pedunculated [1,4]. Benign neoplasms (i.e., myxoma or papillary fibroelastoma) are mobile and have a pedicle/stalk-like attachment, features shared with the presented case. Cardiac myxomas are more likely to develop in the left atrium, while cardiac angiosarcomas develop more commonly in the right atrium [3]. Pericardial effusions are present in most cases in which TEE is

performed and generally without cytopathologic evidence of being positive for neoplastic cells [1]. The high prevalence of concomitant pericardial effusions in the setting of a cardiac angiosarcoma can in part obfuscate the sensitivity of detection and characterization of the cardiac neoplasm by echocardiography [1]. Histologically, the cytologically atypical cells forming vascular spaces with a vascular immunophenotype of an angiosarcoma are not shared by benign primary neoplasms of the heart, including myxomas, hemangiomas, etc.

The architecture of the angiosarcoma in this case belied a benign radiologic appearance, and its growth pattern was likely secondary to rheological forces within the atrial chamber. Angiosarcomas of the heart and other anatomic sites are usually infiltrative, and only rarely has such a growth pattern been previously described [5]. This case highlights a rare although potential diagnosis in the differential of right atrial tumors that would generally be favored to represent a benign neoplasm based on radiologic features.

Conflicts of interest

None.

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