

Cardiac Myxoma

The Great Mimicker



Abdallah El Sabbagh, MD,^a Mohammed A. Al-Hijji, MD,^a Jeremy J. Thaden, MD,^a Sorin V. Pislaru, MD, PHD,^a Cristina Pislaru, MD,^a Patricia A. Pellikka, MD,^a Adelaide M. Arruda-Olson, MD,^a Martha Grogan, MD,^a Kevin L. Greason, MD,^b Joseph J. Maleszewski, MD,^{a,c} Kyle W. Klarich, MD,^a Vuyisile T. Nkomo, MD, MPH^a

CARDIAC MYXOMA IS THE MOST COMMON PRIMARY CARDIAC NEOPLASM IN ADULTS. THEY MOST commonly arise within the left atrium, but may arise from other cardiac chambers, rarely from the valves. Histologically, cardiac myxomas consist of lepidic ("myxoma") cells within a myxoid stroma. They can be of variable size, shape, and mobility. Some have a smooth surface, and others are more villiform. The latter tends to associate more with thromboembolic phenomenon, either from friable tumor or adherent surface thrombus forming on the frond-like areas. Intratumoral hemorrhage, owing to the vascular nature of the lesions, and calcification are common. Clinical manifestations of cardiac myxoma are protean and generally nonspecific, often resulting in delayed treatment of prompt surgical resection. Recognition of cardiac myxoma, and discrimination from other cardiac masses, is necessary for accurate treatment and follow-up management. Our aim is to provide an imaging-based description of the varied presentations of cardiac myxoma (Figures 1 to 6).

The clinical presentations of cardiac myxoma are largely determined by the size, architecture, and location of the tumor. Although cardiac myxomas are histologically benign, the manifestations (e.g., embolism) can be serious. Multimodality imaging is pivotal in diagnosing atrial myxomas. Nevertheless, gross and histopathologic evaluation are necessary to confirm the diagnosis.



FIGURE 1 Cardiac Myxoma Mimicking Mitral Valve Disease

A 65-year-old woman presented with progressive exertional dyspnea. Transthoracic and transesophageal echocardiography revealed a large left atrial (LA) mass (A) attached to the atrial septum and (B) prolapsing across the mitral valve in diastole causing mitral valve obstruction. (C) Pre-operative coronary angiogram revealed a tumor blush (arrow). The mass was surgically resected; gross and histologic examination confirmed cardiac myxoma. A 59-year-old woman presented with (D) flash pulmonary edema from (E) an LA mass associated with severe mitral regurgitation. The mass was surgically resected and the valve repaired. (F) Gross and histopathologic evaluation of the mass confirmed cardiac myxoma. LV = left ventricle; RA = right atrium; RV = right ventricle.

From the ^aDivision of Cardiovascular Diseases and Internal Medicine, Mayo Clinic, Rochester, Minnesota; ^bDivision of Cardiovascular Surgery, Mayo Clinic, Rochester, Minnesota; and the ^cDivision of Anatomic Pathology, Mayo Clinic, Rochester, Minnesota. The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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FIGURE 2 Embolic Myxoma Masquerading as Central Nervous System Vasculitis

A 41-year-old woman with a 2-year history of central nervous system vasculitis presented with acute severe headache and left hemiparesis. (A) Head magnetic resonance imaging showed right frontal lobe intraparenchymal hemorrhage (arrow) surrounding a mass (asterisk). (B)The hematoma was evacuated and histologic evaluation of the biopsied mass revealed myxoma. (C) Transesophageal echocardiography showed a small sessile LA mass (arrow) attached to the atrial septum. The mass was surgically resected and (D) confirmed to be a myxoma. (E, F) She later presented with bilateral hip pain from multiple metastatic biopsy proven myxoma lesions (arrows) that were treated with chemotherapy and radiation. AO = aorta; other abbreviations as in Figure 1.



FIGURE 3 Cardiac Myxoma, Hyperpigmentation, and Endocrinopathy

An 18-year-old woman presented with moon facies and (A) facial lentigines on physical examination. Cortisol level was elevated. (B) Computed tomography of the adrenal glands revealed bilateral micronodular adrenal hyperplasia (arrow), and she underwent adrenalectomy. She later developed amenorrhea with elevated prolactin. (C) Brain magnetic resonance imaging showed pituitary adenoma (arrow). A multiple neoplasia syndrome was suspected and a right atrial mass (arrow) was found on (D) transthoracic and transesophageal echocardiographies, (E) resected, and confirmed to be a cardiac myxoma. (F) Myxoma recurred 3 years later on the RV free wall but remained stable on annual cardiac magnetic resonance surveillance. Abbreviations as in Figure 1.

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