

Atypical location of extracardiac myxoma: a case report

Sultan Pehlivan¹, Ramazan Akçan², Mahmut Şerif Yıldırım², Asude Gökmen¹, Mehtap Yöndem¹

ABSTRACT

Cardiac myxomas are mostly arising from interatrial septum, particularly from fossa ovalis, back and front atrial wall and appendix. However, 10% of myxomas arise from atypical locations. This report presents a rare case of atypical location of extracardiac myxoma in a case of sudden death.

A 43-year old female suddenly dies at home. At autopsy, three masses attached each other were observed in pericardial fat tissue localized between right atrium, vena cava superior wall, and right ventricle. Three masses, as a lobulated single mass, were 7x2x1 cm, 5x5x2 cm, 3x2x0.5 cm in size, respectively. Masses were observed to surround right coronary artery for a segment of 7 cm. Cardiac dissection revealed that mass infiltrates epicardial site of right atrium and ventricle wall but not infiltrated to myocardium and heart cavities. Histopathological and immunohistochemical findings were compatible with a rarely seen atypically localized extracardiac myxoma on right atrial epicardium.

Keywords: sudden death, myxoma, extracardiac myxoma, autopsy

INTRODUCTION

Myxoma is the most common primary tumor of the heart among adults. An overwhelming majority (75%) of primary cardiac myxomas is located in the left atrium, while only % 10-20 of myxomas take place in right atrium which is an atypical location for myxoma (1). Recently, a series presenting 13 years of experience regarding surgical treatment of primary cardiac myxoma revealed that right atrial myxoma was seen 17% of cases (2). Right atrial myxomas are structurally solid and tend to be broad-based, which are mostly arise from interatrial septum; however, there are reported cases with atypical locations such as junction of right atrium and superior vena cava (3). Here, we present an unusual case of atypical location and type of extracardiac myxoma on right atrial epicardium in a case of sudden death of adult female in order to attract attention to unusual locations and dimensions of cardiac myxomas.

CASE REPORT

A 43-year old female suddenly dies at home while resting. Death scene investigation was unremarkable. Deceased had no history for cardiac disease.

At autopsy, external examination of body was unremarkable. In internal examination, right and left lungs were 438 g and 720 g in weight, respectively. Slight pleural thickening and fibrosis was observed on the surfaces of both lungs. Lungs' sections revealed edema, increased consistency, and pneumonia related findings. Pericardium was intact and 400 cc of serous fluid of pericardial effusion was observed in the sac. Three masses attached each other, as a lobulated mass, were observed on pericardial fat tissue localized between right atrium, vena cava superior wall, and right ventricle. Masses were broad based, solid and 7x2x1cm, 5x5x2cm, 3x2x0.5cm in size, respectively. Masses were observed to surround right coronary artery for segment of 7 cm (Figure 1a and Figure 1b). Coronary arteries were intact, and myocardial sections were unremarkable. Cardiac dissection revealed that

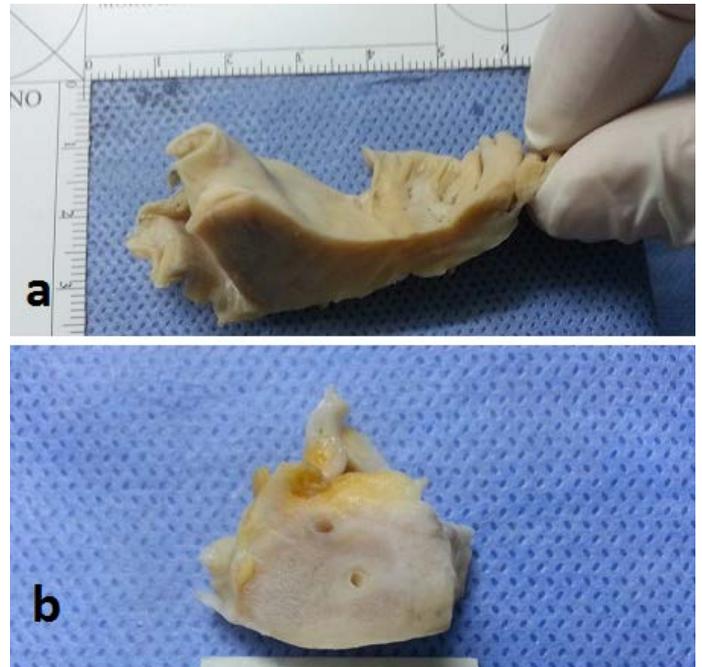


Figure 1: a) Solid tumoral mass arising from junction of right atrium and vena cava superior b) Tumoral mass surrounding coronary artery

described masses infiltrated extra-cardiac site of right atrium and ventricle but not infiltrated to myocardium.

Microscopic examination revealed that a myxomatous mass surrounding right coronary artery was infiltrated/destroyed pericardial fat (Figure 2a). This tumoral mass was characterized with collagenous connective tissue, dense amorphous extracellular matrix, abnormal vascular structures, and insignificant organization of plasmacytoid, spindle and polygonal shaped cells on a large myxoid stroma, in histopathological examination. Foci of smooth muscle cells, inflammation consisting of lymphocytes, plasma cells, and a small number of eosinophils were noticed in tumor mass (Figure

¹ Ankara Group Administration of the Council of Forensic Medicine, Ankara, Turkey.

² Hacettepe University Medical Faculty Department of Forensic Medicine Ankara, Turkey.

Correspondence: Ramazan Akcan

Hacettepe University Faculty of Medicine, Department of Forensic medicine, 06100.

Tel: +903123052004-108

E-mail: akcanmd@hotmail.com

2b and Figure 2c). However, there were no necrosis, mitosis, atypical changes and pleomorphism in tumoral mass. In immunohistochemical evaluation, calretinin (Figure 3a) and vimentin (Figure 3b) were positive in tumoral areas. Similarly PAS-AB (2.5) (Figure 3c) was positive in tumoral mucoid areas, CD34 (Figure 3d) was positive in rudimentary vascular structures, and S100 (Figure 3e) was positive in some of tumoral cells. However, HMB45F (Figure 3f) was negative in tumoral areas (IMPX 200). Findings were compatible with a rarely seen atypically localized extra-cardiac right atrial myxoma. Microscopic examination of lungs showed interstitial and alveolar polymorphonuclear leukocyte infiltration and alveolar macrophages, which were compatible with pneumonia. Detailed toxicological analyses were negative.

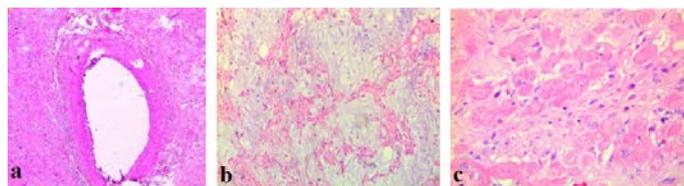


Figure 2: a) Tumoral mass surrounding coronary artery (H&E, X50) b) Endothelial cells, vessels, mononuclear cells, polygonal cells in mixoid stroma in tumoral mass section (H&E, X100) c) Smooth muscle cells and mononuclear cells in tumoral mass section (H&E, X400)

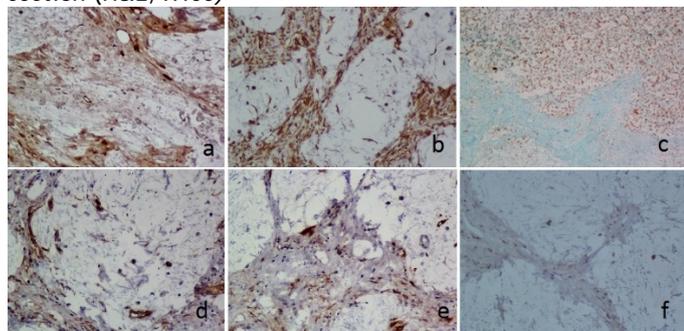


Figure 3: Immunohistochemical staining. Calretinin (a) and vimentin (b) staining in tumoral areas, PAS-AB (2.5) (c) staining in tumoral mucoid areas, CD34 (d) staining in rudimentary vascular structures, S-100 (e) and HMB-45 (f) staining in tumoral areas (IMPX 200)

DISCUSSION

Primary cardiac tumors, that are mostly myxomas, are rarely seen among adults. Myxomas are mostly located in left atrium, and the incidence of right atrial myxomas is quite low as previously reported in literature (1). A case report by Spirito et al. presented a case of unusual extra-cardiac right ventricular myxoma (4). Similarly, we present an unusual extra-cardiac epicardially located myxoma of right atrium, in this report. The literature states that approximately 70% of adult myxoma patients are women (5, 6), predominantly between 30 and 60 years of age. Additionally most of such cases are solitary and sporadic, while some are associated with certain syndromes (7, 8). In accordance with the literature, presented case was a

43 year-old schizophrenic woman without history of familial cardiac diseases.

Virtually all myxomas are polypoid or pedunculated, single, and round masses protruded into cardiac chambers (9). However, Spirito et al. reported an unusual case of extra-cardiac epicardial myxoma (4). Right atrial myxomas tend to be solid and broad based more than left atrial myxomas. Additionally, size of myxoma masses are reported to mostly varies from 1 to 15 cm in diameter (4). The consistency of these masses might vary from hard masses to soft, translucent or villous lesions showing gelatinous appearance with brownish color (10). On the contrary to common cases, three masses attached each other, as a lobulated single mass, were observed on pericardial fat tissue localized between right atrium, vena cava superior wall, and right ventricle, in the presented case. Masses were broad based (not pedunculated), solid and 7x2x1 cm, 5x5x2 cm, 3x2x0.5 cm in size, respectively. Masses were observed to surround right coronary artery for segment of 7 cm, in length. To the best of our knowledge, this report presents the largest unusual extra-cardiac right atrial myxoma case described in the literature.

Histological characteristics of myxomas include a loose myxoid stroma with minimal cellularity in which scattered spindle cells with an eosinophilic cytoplasm. However, mitosis, necrosis, and pleomorphism are not among common findings. The diagnosis of myxomas is difficult because of its atypical and quite variable signs and symptoms; furthermore, it sometimes might be diagnosed only in postmortem (11, 12).

Calretinin is being expressed in 75% of cardiac myxomas. In accordance, the presented case was positive for calretinin and vimentin staining. Furthermore, mucoid areas were positively stained with PAS-AB. Regarding differentiation for myxoid lipomas S-100 is being utilized; it shows variable staining patterns for tumoral cells in cardiac myxomas similar to our case. Additionally, CD34 staining can be positive with intratumoral rudimentary vascular structures (13). In regard to differential diagnosis from liposarcoma, while S100 is positive for both tumors, morphological characteristics and calretinin positivity for cardiac myxomas suggest that the tumor detected in our case was a cardiac myxoma (14). On the other hand HMB-45, which is utilized for differential diagnosis of angiomyolipoma, was negative in presented case.

Affected individuals might show an asymptomatic course until sudden death according to tumor's size, mobility and location, and activity of the patient (11, 14, 15). Similarly, microscopic features of our case were compatible with those reported in the literature. The presented case was also asymptomatic until death, and myxoma was diagnosed incidentally during autopsy.

The chance of diagnosis in living is reported to be approximately 95% with transthoracic echocardiography, of which sensitivity increases to 100% when followed by transesophageal echocardiography (2). However being a rare location of myxoma, right atrial and even extra-cardiac masses located on epicardium should be considered in the differential diagnosis in individuals with atypical signs and symptoms of hearth conditions of uncertain etiology.

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