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journal homepage: www.casereports.comA giant right atrial villous myxoma with simultaneous pulmonary embolism[☆]Cemalettin Aydın^{a,*}, Abdurrahman Taşal^b, Yasin Ay^a, Mehmet Akif Vatankulu^b, Bekir İnan^a, Ahmet Bacaksız^b^a Department of Cardiovascular Surgery, Bezmialem Vakıf University, 34093 Fatih, Istanbul, Turkey^b Department of Cardiology, Bezmialem Vakıf University, 34093 Fatih, Istanbul, Turkey

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ABSTRACT

INTRODUCTION: Primary cardiac tumors are rare and approximately three quarters of them are benign and up to half of the benign tumors are myxomas. Right atrial villous myxoma with pulmonary embolism is an unusual apparition.

PRESENTATION OF CASE: A 29 year-old male was admitted to our outpatient clinic with progressive exertional dyspnea, chest pain and intermittent feeling faint. A giant right atrial villous mobile mass was detected by means of transthoracic echocardiography. To exclude possible pulmonary embolism, chest computed tomography scan was performed and showed filling defects in the branch of the pulmonary artery. The mass was totally resected.

DISCUSSION: RA villous myxoma is a rare subtype in an unusual location with high potential of pulmonary embolism. Early surgery for villous myxoma has a great importance in order to reduce the risk of pulmonary embolism.

CONCLUSION: 3D TEE should be a sufficient technique for diagnosis and evaluation of shape, size and origin of the cardiac mass an adequate guide to surgical treatment.

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1. Introduction

Primary cardiac neoplasms are uncommon and occurring with an approximate incidence of 0.001–0.2%¹. Approximately 50% of benign cardiac tumors are myxomas and most of them are typically polypoid and located on the left atrium.^{1,2} The right atrium is an unusual localization, observed only in 15–20% of myxoma cases.² The villous right atrial myxoma is a very rare subtype in particular, and shows high tendency for pulmonary embolism.³ Myxomas may remain asymptomatic or may appear with constitutional, obstructive or embolic symptoms. Transthoracic echocardiography (TTE), transesophageal echocardiography (TEE), computed tomography scan and cardiac magnetic resonance imaging are the techniques being used for diagnosis. The treatment for cardiac myxoma is urgent surgical resection. We report a giant right atrial villous myxoma with simultaneous pulmonary embolism in a 29 year-old male (Figs. 1–4).

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2. Case report

A 29 year-old male admitted to our outpatient clinic with progressive exertional dyspnea, chest pain and intermittent feeling faint complaints. The examinations of the respiratory and other systems were normal. Routine blood tests and chest X-ray was not diagnostic. The electrocardiogram showed sinus rhythm with negative T wave on V1–V2 precordial derivation. TTE showed a villous mobile mass with a diastolic movement through the tricuspid valve toward the right ventricle. The patient underwent TEE evaluation, which confirmed the findings of the TTE and added anatomical detailing and its insertion in the interatrial septum. During the TEE examination, 3D images were recorded for reconstruction. After reconstruction, 3D TEE showed the real shape, size and origin of the mass. For a possible pulmonary embolism, chest computed tomography (CT) scan was performed and showed filling defects in the branches of pulmonary artery. The patient underwent surgery of the right atrial mass, which was a villous of elastic consistency and violet to grayish-yellow in color. The histopathological examination showed it to be a myxoma. The patient was discharged 4 days after surgery, and doing very well after 6 months.

3. Discussion

We present a rare clinical case of a giant villous RA myxoma with simultaneous pulmonary embolism which is an uncommon clinical apparition. A low incidence of RA myxoma has been

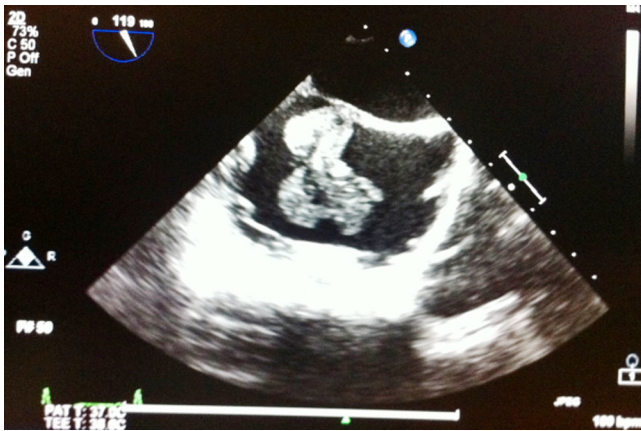


Fig. 1. Right atrial mass on 2D-transesophageal echocardiography. A bicaval view showing a giant villous mass in the right atrium.

reported in several series of autopsy cases. The Mayo clinic series (1954–1979) included 23,673 patients and found 41 patients with primary heart tumors (0.17%).⁴ A myxoma was found in 28 patients and its location was the left atrium in 17 cases and the right atrium in 4 cases (14%). Recently, in a surgical case series, Yu et al. reviewed 33,108 patients submitted to cardiac surgery and found 234 cases with a confirmed diagnosis of primary heart tumor (0.71%).⁵ Among 184 patients with myxoma the RA localization was observed in only 12 (6.5%) of those cases.

According to size, mobility, and location of the tumor as well as physical activity and body position, RA myxomas may remain asymptomatic or may appear with constitutional, obstructive or embolic symptoms. Pulmonary embolism of tumor fragments or thrombus from the tumor surface may also occur, resulting in dyspnea, chest pain, haemoptysis, syncope, pulmonary hypertension and right sided heart failure or death. In this report, the patient complained of exertional dyspnea, atypical chest pain and intermittent feeling faint. TTE showed a villous mobile mass with a diastolic movement through the tricuspid valve toward the right ventricle. The patient underwent 3D TEE evaluation, which confirmed the findings of the TTE and added anatomical detailing and its insertion in the interatrial septum. For a possible pulmonary embolism, chest CT scan was performed that showed filling defects in the branches of pulmonary artery.

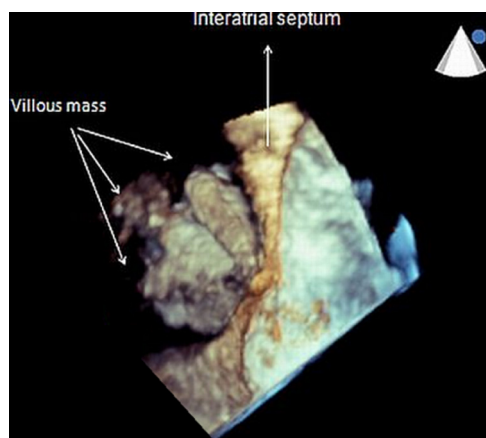


Fig. 2. Right atrial mass on 3D-transesophageal echocardiography. 3D full volume image reconstruction better defined the mass real shape, size and its attachment in the interatrial septum.

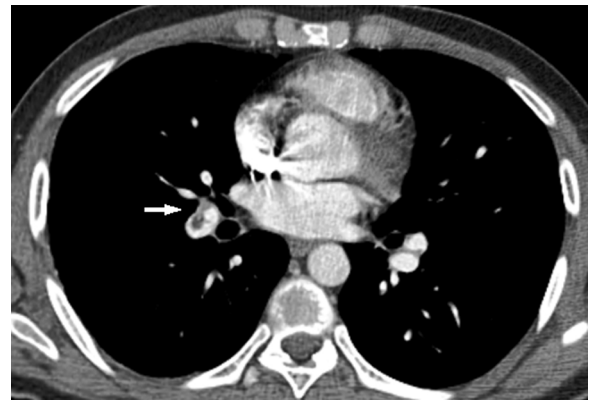


Fig. 3. An image of a chest computed tomography scan with intravenous contrast shows filling defects in the branch of pulmonary artery.

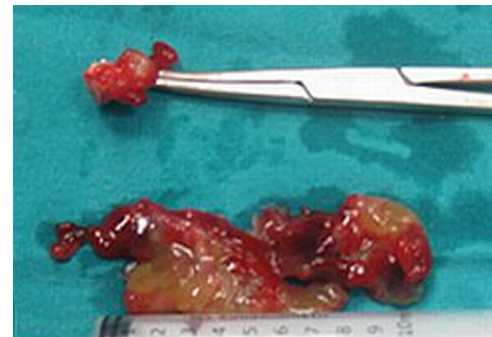


Fig. 4. Photograph of the right atrial myxoma resection.

Pulmonary embolism is a rare but important complication of RA myxoma. In contrast to polypoid myxomas which are usually compact and show little tendency toward spontaneous fragmentation, the less common villous myxomas have a surface that consists of multiple fine or very fine villous extensions. These extensions are gelatinous and fragile and tend to break off or into pieces.³ Once a cardiac myxoma is diagnosed, surgical excision should be performed without delay to minimize risk of embolic events.⁶

Although TTE and TEE provided a good visualization of the mass with a sensitivity of 95% and 100% respectively, 3D TEE may be helpful in surgery planning, allowing a better definition of the mass real shape, position and attachment in the interatrial septum. In this case, a RA mobile mass originating from the interatrial septum was detected by TTE. Mass size, boundaries and its relationship with the septum was assessed by 3D TEE in detail. The patient was operated and the mass was totally resected as a result of a successful operation. The histopathological examination showed it to be a myxoma.

4. Conclusion

RA villous myxoma is a rare subtype in an unusual location with high potential of pulmonary embolism. Early surgery for villous myxoma has a great importance in order to reduce the risk of pulmonary embolism. 3D TEE, should be a sufficient technique for diagnosis and evaluation of shape, size and origin of the cardiac masses and an adequate guide to surgical treatment.

Conflict of interest statement

None

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None

Ethical approval

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contributions

Cemalettin Aydın, study design, surgeon in the operation. Abdurrahman Taşal, writing, data collection. Yasin Ay, data collection, surgeon in the operation. Mehmet Akif Vatankulu, writing,

data analysis. Bekir İnan, review manuscript, surgeon in the operation. Ahmet Bacaksız, review manuscript, data collection.

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