CASE REPORT

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Giant Right Atrial Myxoma with Symptoms of Right Heart Failure

Nabil Naser[1], Nura Hadziomerovic[1], Djenan Bahram[2], Mirsad Kacila[2], Sanko Pandur[2]

[1]Polyclinic "Dr. Nabil", Sarajevo, Bosnia and Herzegovina
[2]Center for Heart Disease Sarajevo, Sarajevo, Bosnia and Herzegovina

Corresponding author: Prof. Nabil Naser, MD, PhD. Polyclinic "Dr. Nabil", Sarajevo, Bosnia and Herzegovina. E-mail: nabil@bih.net.ba. ORCID ID: http://www.orcid.org/0000-0002-278-8574.

ABSTRACT

Background: Cardiac myxoma is the most common benign tumor of the heart. It presents with a variety of clinical signs and symptomatology making diagnosis frequently quite a challenge. Objective: The aim of this article is to present a case report of giant right atrial myxoma with symptoms of right heart failure in adult patient. Case report: We present a case of large right atrial myxoma which is an uncommon location for this type of heart neoplasms, discovered incidentally in a female patient 77-year-old who came to our polyclinic for cardiological exam with hypertension last 11 years and obesity. Results and Discussion: Various clinical signs and symptoms produced by cardiac myxomas have been reported in the literature. Depending on location and morphology, cardiac tumors can produce four types of clinical manifestations: systemic-constitutional, embolic, cardiac, and secondary metastatic manifestation. Echocardiography as non-invasive imaging method and Transesophageal echocardiography has superior role for precise evaluation of cardiac tumors. Transesophageal echocardiography has superior role for accurate diagnostic evaluation of cardiac mass. Surgical excision of cardiac myxoma carries a low-operative risk and gives excellent short- and long-term results. Conclusion: Myxoma is the most prevalent primary heart tumor. It is rare to find a myxoma in the right atrium, occurring only in 15-20% of myxoma cases. Clinical manifestations of myxomas consist in a triad: constitutional symptoms, embolization and intracardiac obstruction. Transesophageal echocardiography has superior role for precise evaluation of cardiac tumors. Currently, there is no effective medical treatment, and surgical excision of the tumor is necessary.

Keywords: Cardiac mass, Right atrium, Cardiac myxoma, Transesophageal echocardiography, Right heart failure.

1. BACKGROUND

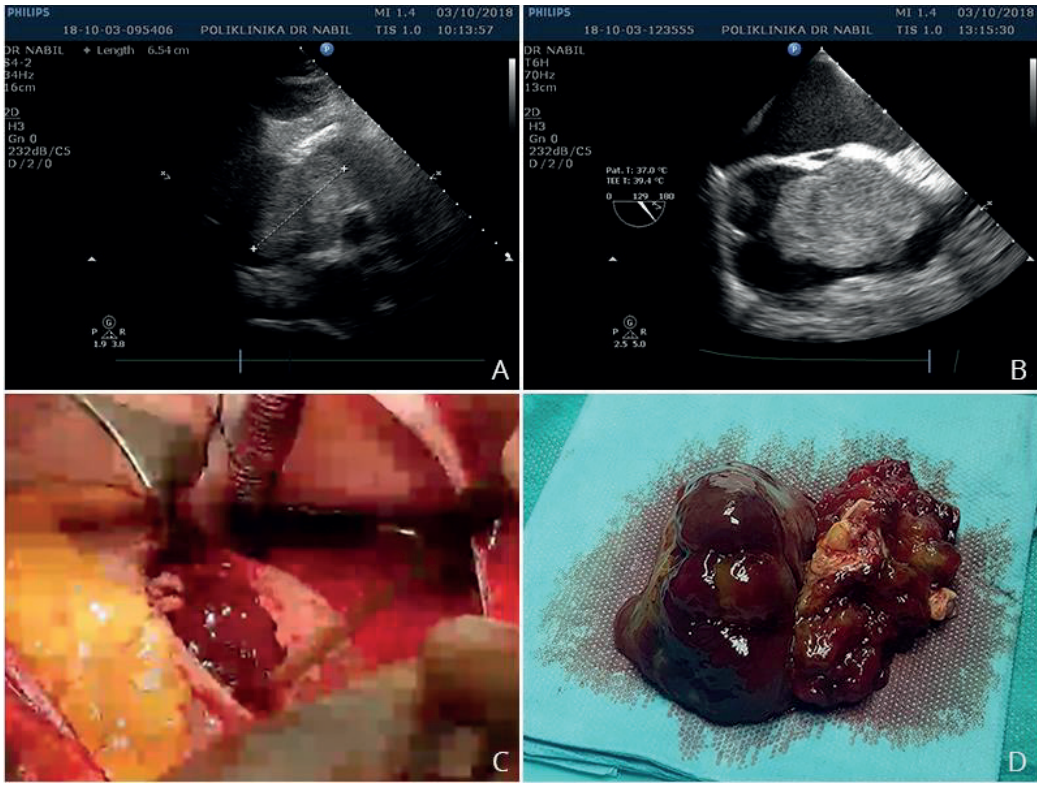
Primary cardiac tumors are rare with an incidence of 0.0001–0.030% in autopsy series, one-fourth of which are presumed to be malignant. Secondary malignancies (metastatic) are the most frequent, with a 20–40 times higher incidence than primary tumors. Intracardiac myxoma is most often a single tumor, arising from the fossa ovalis of the interatrial septum. Myxoma is primary, benign cardiac tumor. About 75% of myxomas occur in the left atrial cavity, but they can also appear in the right atrium (18%) as unusual location. Approximately 75% of primary cardiac tumors are benign pathologically, but those tumors can have "malignant" hemodynamic consequences due to the obstruction of normal blood flow. Accurate diagnostic evaluation of cardiac tumours remains a clinical challenge. Clinical manifestations of myxomas consist in a triad: constitutional symptoms, embolization and intracardiac obstruction. Cardiac surgery without pre-operative precise diagnosis could have disastrous consequences for the patient.

2. OBJECTIVE

We present a case of large right atrial myxoma which is an uncommon location for this type of heart neoplasms, discovered incidentally in a female patient 77-year-old who came to our polyclinic for cardiological exam with hypertension last 11 years and obesity.

3. CASE REPORT

A female patient, 77-year-old, came to our polyclinic for cardiological exam with hypertension last 11 years and obesity. She complained of exertional dyspnea with progressive episodes, palpitation, vertigo, occasional episodes of cardiac arrhythmias, bilateral pitting ankle edema. There were no signs of pulmonary edema. There were signs of right heart failure in the form of



Panel A. Echocardiogram subcostal view showing a huge echogenic mass which almost occupied the right atrium. Panel B. Transesophageal echocardiogram showing a large echo dense mass arising from the interatrial septum in the right atrium which almost completely occupied by the tumor. Panel C. Intraoperative view of tha right atrial giant myxoma. Panel D. Macroscopic aspect of the resected tumor mass which appear multilobulated, well-defined, smooth mass suggestive of cardiac primary myxoma.

jugular venous distension, hepatomegaly of 2 cm with bilateral pitting ankle edema. During physical exam the blood pressure was 150/90 mmHg, pulse 114 beats/min, oxygen saturation 95% on room air, respiratory rate 20 breaths/min. The cardiovascular examinations revealed regular but tachycardic heartbeat with a diastolic murmur of grade 2/6 and an early diastolic tumor "plop" at the left parasternal edge, mild jugular venous congestion. Electrocardiography showed sinus rhythm but was nonspecific. A standard transthoracic 2D echocardiographic window due to obesity had poor quality, but subcostal echo exam showed a large right atrial mass. The examination of other systems was normal. Complete blood count and basic biochemical parameters were normal. Sedimentation rate was 14 mm/h, CRP-hs was 14,9 mg/l. After several hours transesophageal (TEE) exam was performed and confirmed the existence of multilobulated, well-defined, smooth, mobile echogenic mass suggestive of myxoma with mean maximal diameter 6,83 x 1,94 x 5,06 cm, which was attached to the fossa ovalis of interatrial septum. The atrial mass was found to prolapse through the tricuspid valve into the right ventricle, causing obstruction during diastole with protrusion in the right ventricle. The patient underwent open-heart operation and en-bloc excision of the mass with interatrial septum, closing the septum directly with polypropylene and with subsequent significant improvement in symptoms (Panels A, B, C, D). Resected tumor (7.0 × 2,1 × 5.1 cm) having a nodular hemorrhagic surface and mucoid glistening variegated cut surface with focal hemorrhage. Microscopic examination (H&E stain) findings were typical for cardiac myxoma (stellated, fusiform, and polygonal cells. Myxoma cells are arranged singly, forming rings or cords around the capillaries).

4. RESULTS AND DISCUSSION

Primary heart neoplasms are rare occurring with an estimated incidence of 0.0017-0.19%. Primary tumors of the heart are rare. Based upon the data of 22 large autopsy series, the frequency of primary cardiac tumors is approximately 0.02%–corresponding to 200 tumors in 1 million autopsies (1, 2). Myxoma is the most prevalent primary heart tumor. It is rare to find a myxoma in the right atrium, occurring only in 15-20% of myxoma cases. In a recent publication reporting 19 years of experience with surgical treatment of primary intracardiac myxoma, seven (17%) cases out of 41 originated from the RA (3-6).

Various clinical signs and symptoms produced by cardiac myxomas have been reported in the literature. Depending on location and morphology, cardiac tumors can produce four types of clinical manifestations: systemic-constitutional, embolic, cardiac, and secondary metastatic manifestation (7-9). We report a rare case of a massive right atrial myxoma causing tricuspid valve obstruction during with right heart failure signs. This case illustrates the influence of myxoma's size, position, and mobility as well as patients complain and signs. Death is typically caused by coronary or systemic embolization

or by obstruction of blood flow at the mitral or tricuspid valve. Morbidity is related to symptoms produced by tumor embolism, heart failure, mechanical valvular obstruction, and various constitutional symptoms. Taken together, giant right atrial myxomas are very rare, the tumor was attached to fossa ovalis of the interatrial septum as atypical locations, and in such cases underlying malignancy should be considered (7-12).

5. CONCLUSION

Myxoma is the most prevalent primary heart tumor. It is rare to find a myxoma in the right atrium, occurring only in 15-20% of myxoma cases. Clinical manifestations of myxomas consist in a triad: constitutional symptoms, embolization and intracardiac obstruction. Transesophageal echocardiography has superior role for precise evaluation of cardiac tumors. Myxomas have the potential to cause serious complications, including coronary or systemic embolization events and partial or complete obstruction of intracardiac blood flow. This case illustrates the influence of myxoma's size, position, and mobility as well as patients complain and signs. Currently, there is no effective medical treatment, and surgical excision of the tumor is necessary. The surgical excision of cardiac myxoma carries a low-operative risk and gives excellent short- and long-term results.

Panel A. Echocardiogram subcostal view showing a huge echogenic mass which almost occupied the right atrium. Panel B. Transesophageal echocardiogram showing a large echo dense mass arising from the inter-atrial septum in the right atrium which almost completely occupied by the tumor. Panel C. Intraoperative view of that right atrial giant myxoma. Panel D. Macroscopic aspect of the resected tumor mass which appear multilobulated, well-defined, smooth mass suggestive of cardiac primary myxoma.

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REFERENCES

1. McAllister HA, Jr, Hall RJ, Cooley DA. Tumors of the heart and pericardium. Curr Probl Cardiol. 1999; 24: 57-116. doi: 10.1016/S0146-2806(99)80001-2.
2. Habertheuer A, Laufer G, Wiedemann D, Andreas M, Ehrlich M, Rath C, Kocher A. Primary cardiac tumors on the verge of oblivion: a European experience over 15 years. J Cardiothorac Surg. 2015 Apr 18; 10: 56. doi: 10.1186/s13019-015-0255-4.
3. Mitchell C. et al. Guidelines for Performing a Comprehensive Transthoracic Echocardiographic Examination in Adults: Recommendations from the American Society of Echocardiography. J Am Soc Echocardiogr. 2019; 32: 1: 1-6.
4. Buksa M, Gerc V, Dilic M, Loza V, Naser N, Skolovic S, Hodzic E, Brdjanovic S, Kulic M. Clinical Echocardiographic and Echophonocardiographic Characteristics of the Atrial Myxomas in 22 years period. Med Arh. 2009; 63(6): 320-322.
5. Lancellotti P, Zamorano JL, Habib G, Badano L. The EACVI Textbook of Echocardiography. 2nd edition. Oxford, Oxford University Press; 2016.
6. Bukša M, Naser N, Sokolović S, Kulić M, , Brdžanović S. Atrial Myxomas in Seventeen years Period at Clinic of Cardiology in Sarajevo. 5th Congress of the Croatian Cardiac Society with International Participation, Liječnički vjesnik. 2004;126 (suppl. 1): 102.
7. Hanah RT. et al. Guidelines for Performing a Comprehensive Transesophageal Echocardiographic Examination: Recommendations from the American Society of Echocardiography and the Society of Cardiovascular Anesthesiologists. J Am Soc Echocardiogr. 2013; 26: 921-964.
8. Rudski GL. et al. Guidelines for the Echocardiographic Assessment of the Right Heart in Adults: A Report from the American Society of Echocardiography Endorsed by the European Association of Echocardiography, a registered branch of the European Society of Cardiology, and the Canadian Society of Echocardiography. J Am Soc Echocardiogr. 2010; 23: 685-713.
9. Pepi M, Evangelista A, Nihoyannopoulos P. et al. on behalf of the European Association of Echocardiography Recommendations for echocardiography use in the diagnosis and management of cardiac sources of embolism. 2010; 11: 461-476.
10. Samanidis G, Perreas K, Kalogris P, Dimitriou S, Balanika M, Amanatidis G, Khoury M, Michalis A. Surgical Treatment of Primary Intracardiac Myxoma: 19 Years of Experience. Interactive CardioVascular and Thoracic Surgery. 2011; 13: 597-600.
11. Ojji DB, Ajiduku SS, Omonua OO, Abdulkareem LL, Parsonage W. A Probable Right Atrial Myxoma Prolapsing through the Tricuspid Valve into the Right Ventricle: A Case Report. Cases Journal. 2008; 1: 386.
12. Garatti A, Nano G, Canziani A, et al. Surgical excision of cardiac myxomas: twenty years' experience at a single institution. Ann Thorac Surg. 2012; 93: 825-831.