Right ventricular undifferentiated pleomorphic sarcoma: A case report

Sadra Yadsar, Mahnaz Abavisani, Rasool Lakziyan, Zohreh Sarchahi
Departments of Cardiologist and Nursing, Neyshabur University of Medical Science, Neyshabur; Department of Critical Care Nursing, Rajaie Cardiovascular, Medical and Research Center, Tehran, Iran

ABSTRACT
In this paper, a case of undifferentiated pleomorphic sarcoma in a patient with right-sided heart failure has been explored. A 61-year-old woman complaining from cough and dyspnea for a week following indistinctive surgery on right buttock area with reported pathology of malignant undifferentiated tumor presented to the hospital. Laboratory results indicated negative TPi enzyme and D.dimer of 4127.81 mg/L. In transthoracic echocardiography, a mass filling the entire right ventricular space was observed. A resection surgery was performed and the pathology was reported as primary cardiac sarcoma.

Keywords: Cardiac tumor, right ventricle, sarcoma

Address for correspondence: Dr. Zohreh Sarchahi, Department of Nursing, Neyshabur University of Medical Sciences, Neyshabur, Iran.
E-mail: sarchahiz1@nums.ac.ir
Submitted: 22-Mar-2019 Revised: 19-May-2019 Accepted: 06-Jun-2019 Published: 22-Jan-2021

INTRODUCTION
Primary cardiac tumors are rare with an incidence rate of 0.001–0.03 reported in autopsy studies. Most cases of sarcomas occur between the third and fifth decades of life with a male preponderance (male-to-female ratio 2:1). Sarcomas usually involve right-sided heart, and in 80% of cases, the right atrium is involved. These tumors have poor prognosis with rapid progression, which lead to death due to hemodynamic disturbances, localized invasion, or distant metastasis during few weeks to months after onset of the disease. In this paper, we explored a case of undifferentiated pleomorphic sarcoma in a patient with right-sided heart failure.

CASE REPORT
A 61-year-old woman presented to the hospital with 1-week history of activity-related dyspnea and coughing after indistinctive mass resection surgery on right buttock area. The pathology of removed mass was reported as being a malignant undifferentiated tumor.

The patient was on old conscious woman who was responsive to the questions and was not toxic and ill. Vital signs were as follows – BP: 120/70 mmHg, HR: 82, RR: 18 breaths/min, and oral body temperature 36.0°C and O₂ sat 97%. In examination, conjunctiva was pale and S1 and S2 were heard without murmur; friction rub was not heard. Respiratory sounds were normal. Edema, lymphadenopathy, Kussmaul's sign, intermittent pulse, and Crackles were not observed.

Initial laboratory results were as follows: negative TPi enzyme, CKMB: 55 u/L, D.dimer: 4127.81 mg/L, Hb: 9.4, PT: 14.2 s, PTT: 36 s, INR: 1.5, NT-proBNP:
7,569 ng/L, ESR: 30, and CRP: 41 mg/L. ABG revealed metabolic acidosis compensated with respiratory alkalosis. Electrocardiography and coronary angiography findings were normal.

Due to severe dyspnea and coughing, the patient underwent two-dimensional transthoracic echocardiography, which revealed adequate left ventricular functioning, enlarged right ventricle, mild mitral valve regurgitation (+1), mild tricuspid valve stenosis (+1) along with moderate tricuspid valve regurgitation (+2). Also, it revealed a large mass in the right ventricle. The patient was admitted with suspicion of thromboembolism and pulmonary computed tomography (CT) angiography was requested.

Pulmonary CT angiography showed a 4.3 × 3.6-cm large tumoral mass in the right ventricle with adhesion extending into the pulmonary artery [Figure 1]. Right and left pulmonary arteries were normal without thrombosis and mild pleural effusion was reported in the left side. The patient underwent emergency sternotomy and open heart surgery due to hypotension and low O₂ sat. A 4.7 × 4-cm cream-colored solid mass inside right ventricle and pulmonary artery truncus was removed and sent to pathology department. In pathology report, an infiltrative neoplastic tissue consisting of pleomorphic spleen cells with large vesicular and eosinophilic nucleus was observed. In immunohistochemistry, S100, Desmin, and CD31 were negative that confirmed diagnosis of undifferentiated pleomorphic sarcoma. Considering the history of mass resection surgery on right buttock with pathological report of malignant tumor, primary right ventricular sarcoma tumor was considered. The patient was transferred to oncology ward and underwent chemotherapy. After the procedure, she suffered from increased shortness of breath, supraventricular tachycardia, and atrial fibrillation and passed away 2 days later due to respiratory-cardiac arrest.

DISCUSSION

The overall prevalence of primary cardiac tumors is about 0.5/million each year. In various studies, the incidence of primary cardiac tumors from all heart surgeries has been reported to be lower than one percent. It is noteworthy that this study was carried out due to the rarity of undifferentiated pleomorphic sarcoma. Also, the case was a female in her sixth decade of life presenting with sarcoma, which is inconsistent with available data regarding sarcomas. Bouma et al. showed that clinical symptoms in malignant tumors are often nonspecific and dyspnea is the most common symptom. In >50% of cases, systemic metastasis is observed at diagnosis. From clinical perspective, these tumors are progressive and mortality occurs due to extensive metastasis. In this study, the initial sign was dyspnea and soft tissue metastasis was observed. Intracardiac thrombosis is considered as differential diagnosis of cardiac tumors. Intracardiac thrombosis is most frequently seen in patients with valve disease, myocardial infarction and cardiac dysfunction, atrial fibrillation, and prethrombotic syndromes. The distinction of tumor from thrombosis is of major importance. In this study, the patient presented with activity dyspnea and coughing after immobility and pulmonary thromboembolism was also considered as differential diagnosis, which fortunately was excluded using diagnostic methods. Transthoracic echocardiography is the major diagnostic and follow-up method in cardiac tumors, because it is a high precision, nonaggressive, and inexpensive method. It provides sufficient functional and anatomical information regarding intracardiac lesions but has limited diagnostic ability in differentiating benign and malignant tumors. Echocardiography is helpful in differentiation of three types of common intracardiac masses including tumor and thrombosis. In this study, transthoracic echocardiographic findings were sufficiently accurate regarding the presence of a mass, location, base, and mobility of the tumor. Additional data about size, base, and mobility of the tumor were gathered using pulmonary CT angiography. Coronary angiography is mainly carried out to exclude the possibility of a coronary vascular disease, which is usually performed on patients over 40 with probable coronary vascular involvement. In this study, coronary vascular angiography indicated normal findings.

![Figure 1: Pulmonary computed tomography angiography with a mass in the right ventricle and mild left-sided pleural effusion](image-url)
Bakaeen et al. reported that the in-hospital mortality or within 30 days of the operation was 22% for malignant tumors and 3% for benign tumors. Surgical morbidity was 67% for malignant tumors, which was more two times of that for benign disease. Risk factors for surgical mortality were reported to be multiple comorbidities, late diagnosis and treatment, coronary bypass co-surgery, cardiac valve disease, and malignant tumors. In this study, early diagnosis and treatment were not carried out due to the fact that the patient did not follow up the mass on the right buttock for a long time, and she presented with increased dyspnea, supraventricular arrhythmias, and atrial fibrillation and passed away 2 days after successful resection surgery due to respiratory-cardiac arrest.

CONCLUSION

Malignant tumors are considered as a challenge which, despite of surgical and complementary therapies, are associated with limited survival and poor prognosis due to distant metastasis and localized relapse. Ongoing research is carried out in hope of early diagnosis and proper treatment.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Mann D, Zipes D, Libby P, Bonow R. Braunwald’s heart disease: A textbook of cardiovascular medicine, single volume. USA: Saunders; 2012. p. 1-2040.
2. Kodali D, Seetharaman K. Primary cardiac angiosarcoma. Sarcoma 2006;2006:39130.
3. Kasper DL, Fauci AS, Hauser SL, Longo DL, Jameson JL, Loscalzo J. Harrison’s Principles of Internal Medicine. Vols. 1 and. 2. New York: McGraw Hill Professional; 2018.
4. MacGowan SW, Sidhu P, Aherne T, Luke D, Wood AE, Neligan MC, et al. Atrial myxoma: National incidence, diagnosis and surgical management. Ir J Med Sci 1993;162:223-6.
5. Sezai Y. Tumors of the heart. Incidence and clinical importance of cardiac tumors in Japan and operative technique for large left atrial tumors. Thorac Cardiovasc Surg 1990;38(Suppl 2):201-4.
6. Bouma W, Lexis CP, Willems TP, Suurmeijer AJ, van der Horst IC, Ebels T, et al. Successful surgical excision of primary right atrial angiosarcoma. J Cardiothorac Surg 2011;6:47.
7. Miller DV, Edwards WD. Cardiovascular tumor-like conditions. Semin Diagn Pathol 2008;25:54-64.
8. Matebele MP, Peters P, Mundy J, Shah P. Cardiac tumors in adults: Surgical management and follow-up of 19 patients in an Australian tertiary hospital. Interact Cardiovasc Thorac Surg 2010;10:892-5.
9. Perek B, Misterski M, Stefaniak S, Ligowski M, Puslecki M, Jemielity M. Early and long-term outcome of surgery for cardiac myxoma: Experience of a single cardiac surgical centre. Kardiol Pol 2011;69:558-64.
10. Bateman TM, Gray RJ, Raymond MJ, Chaux A, Czer LS, Matloff JM. Arrhythmias and conduction disturbances following cardiac operation for the removal of left atrial myxomas. J Thorac Cardiovasc Surg 1983;86:601-7.