International Journal of Medical Science and Clinical Inventions 4(7): 3116-3118, 2017
DOI:10.18535/ijmsci/v4i7.14 **ICV 2015: 52.82**
e-ISSN:2348-991X, p-ISSN: 2454-9576

Case report

Multiple Melanomas of the Right Atrium as a First Sign of a Metastatic Disease

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Abstract: Out of all malignant tumors, metastatic melanoma has the highest propensity of heart metastases. Metastatic heart tumor is often a sign of disseminated malignant disease and poor prognosis. In our 39-year-old female patient, the symptoms of heart failure caused by metastatic melanoma of the right atrium were the first sign of advanced metastatic disease. The tumor masses were completely resected along with the affected wall of the right atrium, which was reconstructed with an autologous pericardial patch. The patient underwent further oncological treatment, and was asymptomatic for four years after surgery. Considering new treatment options in oncology a complete resection of metastatic heart melanoma offers a chance to improve quality of life and prognosis of these patients.

Key Words: Cardiac, Metastases, Melanoma, Surgery

INTRODUCTION

Metastatic heart tumors occur 20-40 times more frequently than primary heart tumors, whose incidence is about 0.1% [1]. Beside melanoma, which is the most common metastatic heart tumor, heart metastases are often found in mesothelioma, lung and breast carcinoma [2]. In 1/3 of the cases, malignant melanoma advances to metastatic disease and in about 50% of these patients there is a heart involvement [1]. The presence of metastatic heart tumor is often a sign of advanced malignant disease and poor prognosis.

We will demonstrate the case of 39-year-old female patient in whom metastatic melanoma of the right atrium was the first sign of disseminated malignant disease, 15 years after the resection of primary cutaneous melanoma.

Case report

In July 2011, 39-year-old female patient was admitted to emergency department due to progressive dyspnea and exercise intolerance. These symptoms lasted for 3 weeks and progressed to NYHA III grade of heart failure. The patient had no chronic illnesses except euthyroid goiter. She had a history of the surgical excision of cutaneous melanoma in the left shoulder region 15 years ago. On the physical examination a systolic-diastolic murmur was heard over precordium. Laboratory tests showed sideropenic anemia, elevated C-reactive protein at 11 mg/L, and lactate dehydrogenase (LDH) at 550 IU. Troponin I levels were in the normal range at 0.02 µg/l.

Transthoracic echocardiography showed two tumor masses in right atrium. The larger tumor (52x34 mm) was on the atrial wall above the annulus of tricuspid valve with a diastolic prolapse through the orifice of tricuspid valve. Smaller tumor (30x17mm) was sessile on the posterolateral right atrial wall (Figure 1). The tumor masses functionally narrowed tricuspid valve orifice to 1.41 cm^2, with a mean transtricuspid gradient of 11.7 mmHg.

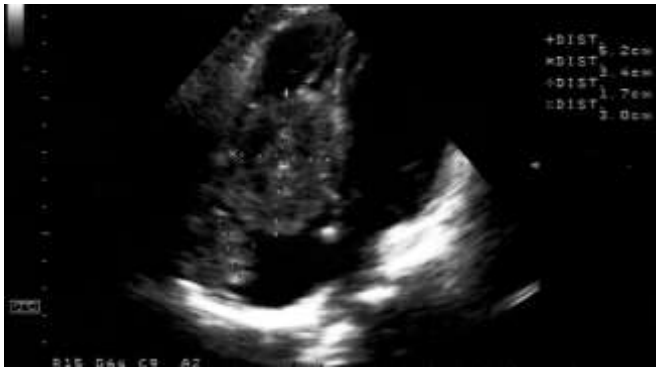

Figure 1. – Echocardiography showing tumor mass in the right atrium

According to echocardiography we suspected of a right atrial myxoma and due to acute heart failure cardiac surgery was indicated.

Surgical approach was through the median sternotomy, with bicaval venous cannulation. Minimal manipulations were made to avoid tumor embolization. A transverse incision of the right atrial wall above the tumor site was made. Three tumor masses with hemorrhagic zones were found, the largest around 6 cm in diameter on a wide base that infiltrated right atrial wall next to right ventricle along the posterior leaflet of tricuspid valve. The other two tumors were smaller, and with a wide base that infiltrated the lateral side of right atrial wall (Figure 2).

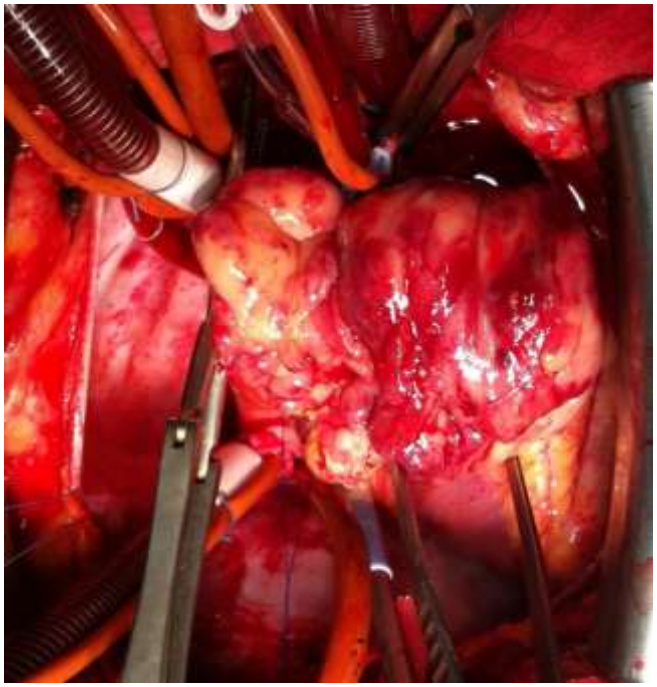

Figure 2. – Intraoperative view of a large tumor masses of the right atrium with focal necroses. The right atrial wall above the tumor was excised.

Complete resection of the tumor masses was made along with their base in the wall of the right atrium. Autologous pericardial patch was then used for reconstruction of the right atrial wall.

Patohistological analysis showed tumor tissue composed of clusters of atypical epithelial cells with numerous mitoses, focal necrosis or inflammatory mononuclear infiltrate. Immunohistochemistry was pancitokeratin negative, vimentin positive, LCA (human leukocyte common antigen) focal positive on inflammatory infiltrate, CD3 (focal positive), CD20 focal positive, HMB 45 (anti-gp100) positive. Histological analysis and immunohistochemistry cofirmed metastatic melanoma of the heart.

Postoperative course of our patient was uneventful, with no further cardiac symptoms. One month after surgery a PET-CT was made, which showed no new cardiac metastases, but an increased glucose metabolism that was present in the left gluteal region, liver segments IV and VII, omental bursa next to pancreatic tail, and enlarged lymph nodes near the medial side of the left adrenal gland, lower lobe of left kidney, and in the left pararenal space. The patient was referred to further oncology treatment.

Two months after the resection of cardiac metastases, the patient was again admitted to hospital, due to ileus. Laparotomy showed invagination of the terminal ileum into caecum, thus right hemicolectomy was made. Pathohistological analysis of invaginated bowel showed metastatic melanoma. The patient underwent further chemotherapy and immunotherapy. She received two cycles of dacarbazine, and then was involved in the clinical study of the vemurafenib treatment.

Four years after the resection of multiple cardiac melanomas the patient was symptom free. PET-CT of thorax, abdomen and pelvis showed no new metastases, and earlier described metastases were in regression. There were no pathologically enlarged lymph nodes. Cardiac MR showed normal position of heart chambers, without dilation, and normal contractility with a physiological myocardial signal in T1 and T2 phase, normal morphology of valvular apparatus and no signs of recurrent malignant disease.

In the fifth postoperative year the patient died after her neurological condition rapidly worsened and was diagnosed with multiple brain metastases from malignant melanoma.

Discussion

Out of all malignant tumors metastatic melanoma has the highest propensity of heart metastases [3]. Metastatic melanoma is seldom diagnosed ante-mortem in 2% of the patients [4], and just 20% of these patients have cardiac symptomatology [1, 3]. In contrast, autopsy shows cardiac metastases in 50% of patients with metastatic melanoma.

Cardiac symptomatology is very rarely the first manifestation of metastatic melanoma, which was the case in our patient [5]. More often symptoms of cardiac metastases overlap with the other symptoms of advanced metastatic melanoma. Malignant melanoma of the heart most frequently involves the pericardium (58%), myocardium (30%) and endocardium (12%) [6]. Intracavitary metastases of melanoma are most common in the right atrium [1]. The symptoms of metastatic heart melanoma are nonspecific and differ due to tumor location. Hence cardiac melanoma can give a wide range of symptoms such as pericarditis, pericardial effusion, tamponade, congestive heart failure, conduction disturbances or peripheral embolization.

Echocardiography is the most frequently used diagnostic tool in the verification of a heart tumor. Differential diagnosis of intracavitary mass can be thrombus, vegetation or foreign body. Further diagnostic evaluation includes CT, PET-CT and MR, which can show tumor lesions and their metabolic activity, spread of metastatic disease and tumor staging.

The possibility of surgical resection of metastatic heart tumor depends of its anatomic localization. If no complete resection is possible, partial tumor resection prevents or relieves cardiac symptoms. Intracavitary metastases, as in our case, should be completely resected. Thus surgery should be the first line of treatment, especially if metastatic heart tumor disturbs normal cardiac function [6].

Most of the literature mentions poor prognosis of patients with metastatic melanoma, with a median survival of 6.4 months [7]. In our patient complete surgical resection of cardiac melanoma was made. The patient began to recieve standard oncological treatment for metastatic melanoma and then underwent the vemurafenib treatment clinical study. Vemurafenib is selective inhibitor of BRAF serin-threonin kinase, which is active in about 50% of patients with melanoma. The vemurafenib treatment showed longer disease

free intervals without progression of metastatic disease compared to patients on dacarbazine treatment [8].

Four years after the diagnosis of metastatic cardiac melanoma and its complete surgical resection, our patient was symptom free and without new metastases, however in the fifth year of follow up she had multiple brain metastases with a fatal outcome.

Conclusion

Considering new treatment options, the early diagnosis of metastatic heart melanoma has important prognostic and therapeutic implications. In every patient with cardiac symptoms and history of malignant melanoma, we should suspect the possibility of metastatic cardiac melanoma.

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