Case Report

Isolated Right Atrial Metastasis of Malignant Melanoma Mimicking a Myxoma

Loizos Kontozis[1], Marinos Soteriou[1], Demetris Papamichael[2], Constantinos Economides[3], Evis Bagdades[3], Christos Christou[1], Christina Oxynou[3]

[1]*American Heart Institute,* [2]*Bank of Cyprus Oncology Center,* [3]*Apollonion Hospital, Nicosia, Cyprus*

Key words: **Cardiac mass, cancer, echocardiography.**

Malignant melanoma is the malignancy with the highest propensity for cardiac metastasis. Metastasis to the heart usually occurs in the setting of disseminated disease and is therefore commonly associated with a poor prognosis. We report the case of a 51-year-old woman with a previous history of cutaneous malignant melanoma who presented with a symptomatic, isolated right atrial metastasis attached via a narrow stalk to the interatrial septum, thus resembling a myxoma. The lesion was completely resected, rendering the patient symptom and, potentially, disease-free. The case illustrates the importance of cardiac evaluation in the management of patients with melanoma.

Manuscript received: March 3, 2010;
Accepted: July 28, 2010.

Address: Loizos Kontozis

3, Protagorou St. 2008 Nicosia, Cyprus
e-mail: ltkon@hotmail.com

Malignant melanoma has the highest incidence of cardiac metastasis of all malignant tumors.[1,2] Cardiac lesions usually occur in the context of disseminated disease and only rarely are they encountered in isolation.[1] We report a case of an isolated cardiac metastasis mimicking a right atrial myxoma, in a woman with a previous history of malignant melanoma of the breast.

Case presentation

A 51-year-old Caucasian woman presented with a 3-month history of fatigue, malaise and headaches. During the same period she developed progressively deteriorating exertional dyspnea. There was no orthopnea but she complained of a pounding sensation in the neck when lying down. She denied fever, night sweats, weight loss or syncope.

Malignant melanoma of the left breast had been diagnosed 13 years earlier and was completely resected. At that time she was staged, and the lesion was Clarke's level IV, Breslow's thickness 2.82 mm. She remained disease-free for 9 years, when a multicentric recurrence in the left breast was discovered. The patient underwent extensive thoracic wall surgery with left mastectomy and axillary dissection. A left supraclavicular fossa node was involved by melanoma. In view of these findings, she received 'adjuvant' high-dose interferon therapy for 1 year. She had been disease free thereafter.

Clinical examination showed a blood pressure of 104/86 mmHg, a regular tachycardia of 104 beats per minute, an elevated jugular venous pressure of 10 cm with prominent a-waves and a fourth heart sound. The liver was palpable 3 cm below the costal margin. The electrocardiogram showed evidence of right atrial enlargement and was otherwise unremarkable. Chest radiography was normal. A transthoracic echocardiogram revealed a normal left side and a highly mobile oval tumor measuring 5.3 × 4.1 cm, almost completely filling the right atrium and prolapsing into the right ventricle in diastole (Figure 1). The inferior *vena cava* and he-

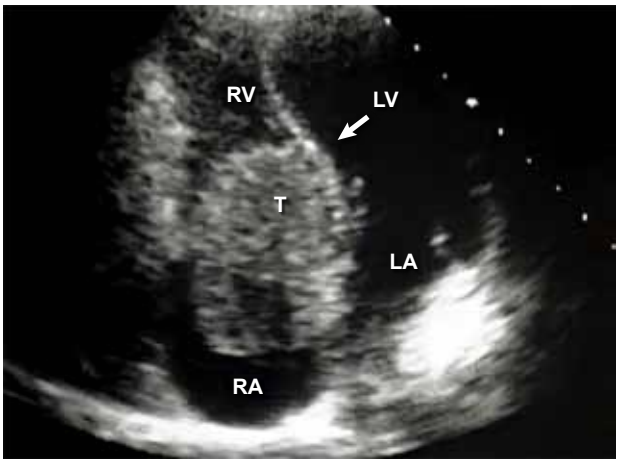

Figure 1. Transthoracic apical 4-chamber view of the heart in diastole showing a large right atrial tumor (T) prolapsing through the tricuspid valve and displacing the interventricular septum to the left (arrow). LA – left atrium; LV – left ventricle; RA – right atrium; RV – right ventricle.

patic veins were distended but clear. No pericardial effusion was noted. Transesophageal echocardiography confirmed the above findings and demonstrated, furthermore, that the mass was lobulated, of homogenous echogenicity and attached by means of a narrow stalk to the lower margin of the *fossa ovalis* (Figure 2). The rest of the heart and the great vessels were normal. On laboratory testing there was a mild elevation of the liver enzymes up to twice the upper limit of normal. The rest of the results, including hematology, inflammation and autoimmune markers were unremarkable. A spiral computed tomographic scan of the chest and abdomen showed no additional pathology. A radioisotope bone scan was normal.

The patient underwent a median sternotomy and the right atrium was opened. A grayish-black mass measuring 5.5 × 5 × 3 cm was identified, attached with a stalk to the interatrial septum in the area of the triangle of Koch just above the coronary sinus and partially obstructing the inferior *vena cava* (Figure 3). The interatrial septum was opened at the *fossa ovalis* and the septum containing the stalk was resected with a 1 cm margin. The resulting defect was repaired with an autologous pericardial patch. No other lesions were present in the right atrium or ventricle. Histopathological examination showed epithelioid cell proliferation with a generally nodular pattern of growth, areas of ischemic necrosis, moderate cytologic pleomorphism and marked melanin-type pigmentation, consistent with malignant melanoma (Figure 4). There was neoplastic infiltration of the stalk and of myocardial elements in the interatrial septum in the area where the stalk was attached.

The patient had an uncomplicated postoperative course and was discharged home 5 days after surgery. On follow up at 5 months she was well, without evidence of tumor recurrence on the transthoracic echocardiogram.

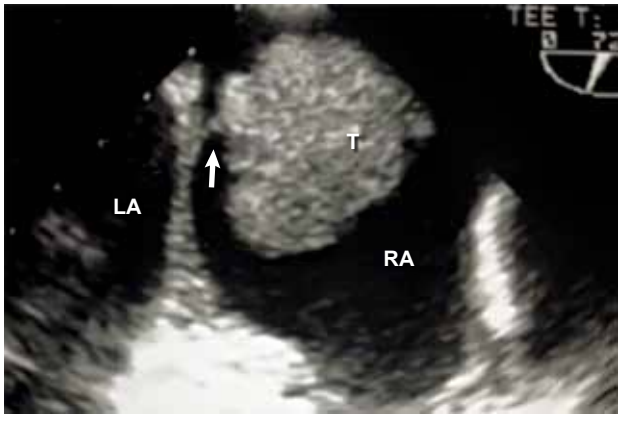

Figure 2. Transesophageal modified biatrial view demonstrating attachment of the lobulated tumor (T) to the interatrial septum via a narrow stalk (arrow). LA – left atrium; RA – right atrium.

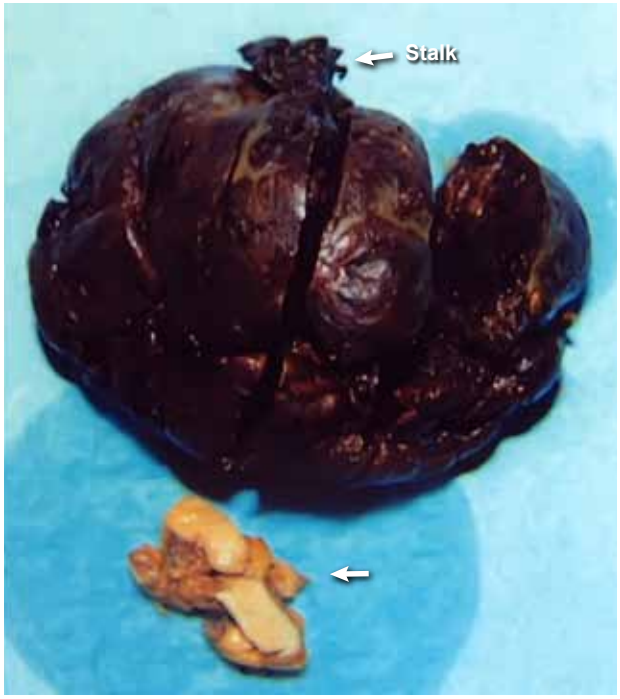

Figure 3. Pedunculated grayish-black tumorous mass and excised segment of atrial septum to which this was attached (white arrow).

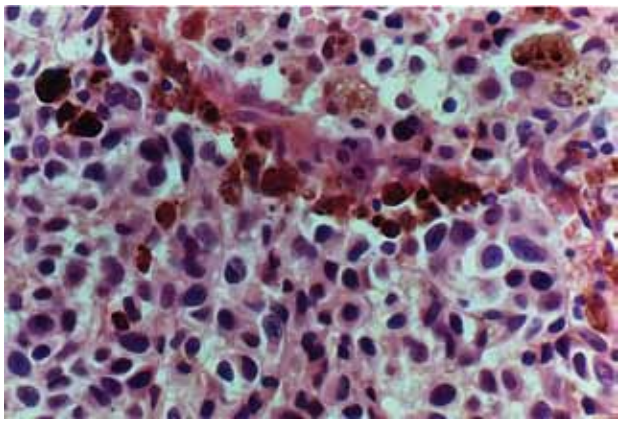

Figure 4. Histological section showing infiltration by atypical epithelioid melanocytes with fine melanin pigmentation (hematoxylin-eosin).

Discussion

Primary tumors of the heart are quite rare, with a reported incidence of <0.1% of all autopsies.[3,4] Of these, myxoma is by far the commonest.[3] Metastatic cardiac lesions are by comparison at least 20 times as frequent, encountered in approximately 11% of the autopsies of patients with malignancy.[2,4,5] The incidence of secondary cardiac tumors appears to have increased in recent years, possibly in part due to improved treatment modalities resulting in increased survival times.[2,6] Ante mortem detection rates have also improved with the widespread use of 2-dimensional, especially transesophageal echocardiography and the increasing use of magnetic resonance imaging and ultrafast computed tomography.

Among malignant tumors, melanoma has the highest propensity for cardiac metastases, detected in 64% of the cases in the largest published autopsy series of metastatic melanoma.[1] Metastases may appear decades after initial diagnosis of the primary tumor.[7-9] They most often affect the myocardium, suggesting a predominantly hematogenous mode of spread.[1,6] Pericardium and endocardium are also frequently involved, with a significant proportion of patients having deposits in all 3 layers. Endocardial lesions are intracavitary, a fact that facilitates their detection and makes them amenable to surgical intervention. Any, and often all of the 4 chambers may be involved, although the right side of the heart is more often affected than the left, with the right atrium being the most frequent site of metastasis.[1] Secondary lesions of the right atrium typically arise from the atrial free wall and are sessile.[1,9-13] This contrasts with myxomas, which are usually attached to the interatrial septum at the border of the *fossa ovalis* and are pedunculated and highly mobile.[14] Despite their high prevalence, melanoma metastases to the heart are clinically silent in the majority of the cases, with only 20% of the patients developing symptoms or signs of cardiac dysfunction.[1,6] When present, cardiovascular manifestations are rarely seen in isolation or as a presenting symptom. This is a consequence of cardiac involvement generally occurring late in the course of the disease, by which time the clinical picture is determined by the widespread dissemination of the tumor to several organs. For the same reason, cardiac lesions are usually multiple at diagnosis. Thus, cardiac involvement, as a rule, implies an adverse prognosis and precludes surgical intervention. However, isolated lesions do occasionally occur and there have been several reports of successful excision, usually partial, of such masses, providing significant palliation and life prolongation to selected patients.[7-13,15,16]

It is evident from the above that in our patient many of the typical features of metastatic malignant melanoma were absent. This is to our knowledge the first reported case of a symptomatic, solitary, right atrial pedunculated lesion attached to the interatrial septum in the area of the *fossa ovalis*, in the absence of any extracardiac metastases. As a consequence, complete and possibly curative resection could be undertaken. Thus, the tumor resembled both clinically and echocardiographically more a myxoma than a malignant melanoma. That it ultimately proved to be a melanoma lends further weight to the adage that "any symptom in a patient with a history of melanoma must be considered to be secondary to metastatic disease until proven otherwise".[17] It is worth noticing that the patient did not undergo a cardiology assessment when the melanoma recurred 4 years prior to presentation. Therefore, we can only speculate as to whether the cardiac metastasis was present at the time.

This case underlines the importance of maintaining a high index of suspicion and actively searching for cardiac involvement in patients with malignant melanoma, both at initial diagnosis and at follow up. Despite the grave prognosis cardiac metastases commonly signify, there are patients with isolated lesions who can benefit from surgery. With improved therapeutic and diagnostic modalities the number of these patients is likely to increase.

References

1. Glancy DL, Roberts WC. The heart in malignant melanoma. A study of 70 autopsy cases. Am J Cardiol. 1968; 21: 555-571.
2. Klatt EC, Heitz DR. Cardiac metastases. Cancer. 1990; 65: 1456-1459.
3. Reynen K. Frequency of primary tumors of the heart. Am J Cardiol. 1966; 77: 107.
4. Lam KY, Dickens P, Chan AC. Tumors of the heart. A 20-year experience with a review of 12,485 consecutive autopsies. Arch Pathol Lab Med. 1993; 117: 1027-1031.
5. Abraham KP, Reddy V, Gattuso P. Neoplasms metastatic to the heart: review of 3314 consecutive autopsies. Am J Cardiovasc Pathol. 1990; 3: 195-198.
6. Reynen K, Köckeritz U, Strasser RH. Metastases to the heart. Ann Oncol. 2004; 15: 375-381.
7. Mindell SM, Chernick AW, Sugarman MH, Zirkin RM, Bloom RE. Right ventricular metastatic melanoma 27 years after resection of the primary tumor. Cancer. 1989; 63: 1237-1239.
8. Gibbs P, Cebon JS, Calafiore P, Robinson WA. Cardiac metastases from malignant melanoma. Cancer. 1999; 85: 78-84.
9. Chen RH, Gaos CM, Frazier OH. Complete resection of a right atrial intracavitary metastatic melanoma. Ann Thorac Surg. 1996; 61: 1255-1257.
10. Mousseaux E, Meunier P, Azancott S, Dubayle P, Gaux JC. Cardiac metastatic melanoma investigated by magnetic resonance imaging. Magn Reson Imaging. 1998; 16: 91-95.
11. Petropoulakis PN, Steriotis JD, Melanidis JG, Asimakopoulos PJ. Metastatic malignant melanoma as an intracavitary obstructive mass in the right heart. Eur J Cardiothorac Surg. 1998; 14: 538-540.
12. Prabhakar G, Vasilakis A, Hill RC, et al. Right atrial metastatic melanoma in a patient with transient ischemic attacks. Ann Thorac Surg. 1998; 65: 844-846.
13. Bossert T, van Son JA, Autschbach R, Mohr FW. Resection of a right atrial metastatic melanoma with unknown origin of primary tumor. Eur J Cardiothorac Surg. 1999; 15: 373-375.
14. Reynen K. Cardiac myxomas. N Engl J Med. 1995; 333: 1610-1617.
15. Canver CC, Lajos TZ, Bernstein Z, DuBois DP, Mentzer RM Jr. Intracavitary melanoma of the left atrium. Ann Thorac Surg .1990; 49: 312-313.
16. Pinho T, Rodrigues-Pereira P, Araújo V, et al. Cardiac metastasis of melanoma as first manifestation of disease. Rv Port Cardiol. 2009; 28: 633-639.
17. Morton DL, Essner R, Kirkwood JM, Wollman RC. Malignant melanoma. In: Bast RC Jr, Kufe DW, Pollock RE, et al, editors. Cancer medicine. 5th ed. Ontario: BC Decker, 2000. p. 1854.