Bilateral lung lesions: when the eyes deceive the brain!

Ann. Ital. Chir., 2012 83: 87-89

Alfonso Fiorelli*, Giovanni Vicidomini*, Alberto Palladino**, Carlo Curcio***, Umberto Caterino°, Giannantonio Nappi°°, Mario Santini*

**Thoracic Surgery Unit, Second University of Naples, Naples, Italy*
***Cardio-miology Unit, Second University of Naples, Naples, Italy*
****Thoracic Surgery Unit, Monaldi Hospital, Naples, Italy*
°*Pneumology Unit, Monaldi Hospital, Naples, Italy*
°°*Cardio-Thoracic Surgery Unit, Monaldi Hospital, Naples, Italy*

Bilateral lung lesions: when the eyes deceive the brain!

We report a clinic case of patient in whom angiosarcoma of the heart presents as bilateral pulmonary nodular infiltrates. The cardiac tumor was clinically silent, the electrocardiogram was normal and the cough was the only symptom. Chest CT scan (Fig. 1) showed bilateral diffuse nodular infiltrates ranging. Clinical clues, the results of laboratory tests and all of the cultures obtained excluded an infectious etiologies; the findings of CT-guided needle biopsy was inconclusive for a definitive diagnosis. Thus, the patient was scheduled for a thoracoscopic biopsy. Surprisingly, the pre-operatory echocardiogram showed a soft tissue mass fixed to the posterior wall of the right atrium. On retrospective reviewing of chest CT scan, a tumor was evident in the right atrium, but it was missed initially. In theory, the lung lesions attract the attention of the observer who had not taken into account anything else as to say: "the brain knows what the eyes want".
The diagnosis of pulmonary metastases was obtained by means of pleural biopsy during right thoracoscopy. Immunoistochemical staining revealed a CK(-), CK7(-), EMA(-), ESA(-), CEA(-), TTF1(-), Vimentina(+), CD31(+), CD117(+) lesion. Because at the time of diagnosis our patient already had lung metastases, he underwent chemotherapy

KEY WORDS: Angiosarcoma, Cardiac, Echocardiography, Lung metastases, Molecular biology.

Introduction

We report a clinic case of patient with bilateral lung lesions due to cardiac angiosarcoma initially missed on chest computed tomography (CT) evaluation and then incidentally discovered with preoperative transthoracic ecochardiogram (TTE). The diagnosis was obtained by means of thoracoscopic biopsy.

Clinical Summary

A 35 years old male was referred to a local hospital for evaluation of bilateral nodular infiltrates seen on chest-X-ray (CXR). He was good health until June 2010 when he had a complaint of cough. His past medical history was negative. Physical examination was unremarkable except for bilateral basal pulmonary crackles. Routine laboratory studies were normal. Chest CT scan (Fig. 1) showed bilateral diffuse nodular infiltrates ranging from

Pervenuto in Redazione Gennaio 2011. Accettato per la pubblicazione Marzo 2011.
Correspondence to: Prof. Mario Santini, Chirurgia Toracica - Seconda Università di Napoli, Piazza Miraglia 2, I-80138 Napies, Italy (e-mail: mario.santini@unina2.it)

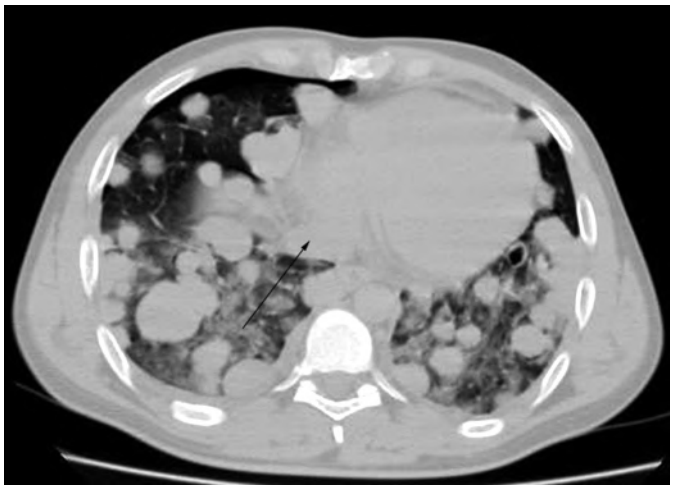

Fig. 1: Chest CT scan shows bilateral diffuse nodular infiltrates ranging from 5 mm to 4 cm with normal heart size.

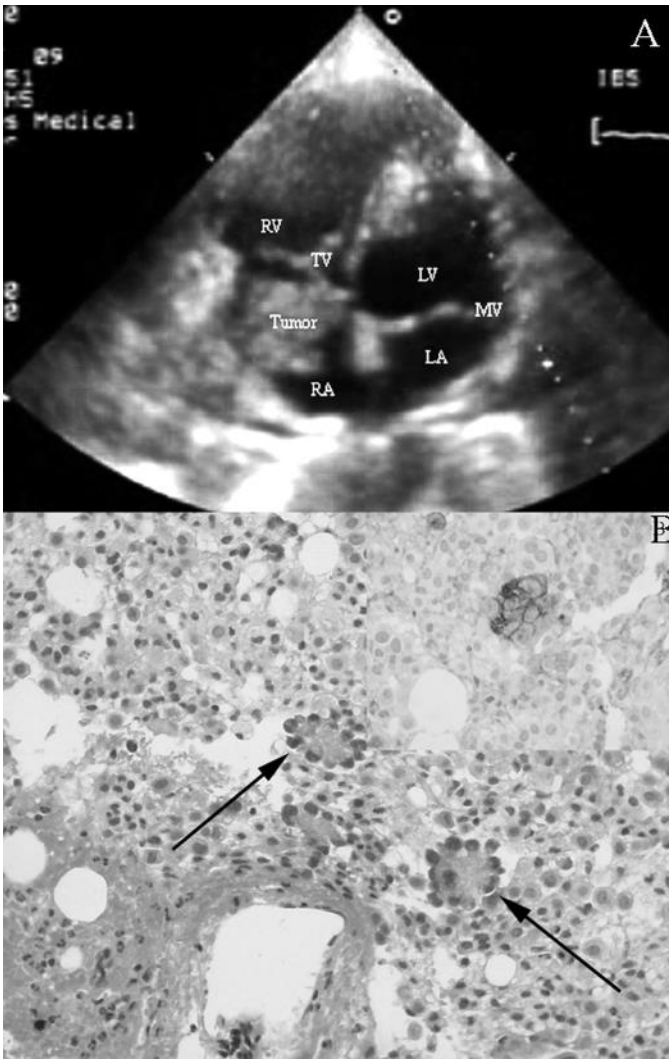

Fig. 2: Transthoracic apical four-chamber echocardiography shows a tumor in the right atrium (RA) without involvement of tricuspidal valve (TV) and of right ventricular (RV). Left atrium (LA), left ventricular (LV), and mitral valve (MV) have normal. There is not pericardium involvement (Part A). Hematoxylin-eosin stained (20x) reveales a papillary neoplasm composed of pleomorphic spindled cells arranged as interconnecting vascular spaces (black arrow). Insert: Immunohistochemical investigation tumor cells (20x) are positive for CD 31 (Part B).

5 mm to 4 cm. The heart was not enlarged and the electrocardiogram was normal. Systemic screening with CT and whole-body bone scan was performed, but no other lesions were found. Broncoscopic view revealed no abnormalities. No malignant cells were recovered by bronchoalveolar lavage and sputum; bacteriological and mycological investigations were negative. Other tests, including as antinuclear antibody, rheumatoid factor, antistreptolysin O, human immunodeficiency virus antibody test, thyroid-stimulating hormone, and purified protein derivate (PPD) skin test were negative. The findings of CT-guided needle biopsy was inconclusive for a definitive diagnosis. Thus, the patient was referred to our unit to attempt a biopsy via thoracoscopy. However a TTE performed for cardiac pre-operatory evaluation, showed a soft tissue mass fixed to the posterior wall of the right atrium and not mobile (Fig. 2). On thoracoscopic view, the lung was dark red and the pleura covered with multiple small tumors which were biopsied. The histopathological study diagnosed epithelioid angiosarcoma. Immunoistochemical staining (Fig. 3) revealed a CK (-), CK7(-), EMA(-), ESA(-), CEA(-), TTF1(-), Vimentina(+), CD31(+), CD117(+) lesion. The patient had an uneventful recovery and he is currently undergoing chemotherapy

Discussion

Cardiac angiosarcoma is a disease rarely diagnosed before death. Although it is the most common malignant primary cardiac tumor, its rare occurrence probably leads to a low index of suspicion. Due to its very high metastatic potential, treatment options are limited and mainly palliative with an average survival of nine months after the diagnosis [1].

The first interesting aspect of our case is that the primary tumor is clinically silent. Considering that it occupied almost the two-third of right atrium, we would expect the presence of symptoms due to pericardial disease or right-sided congestive heart failure with electrocardiogram changes [2]. However, the electrocardiogram is normal and the cough is the only symptom which let the patient to perform a CXR. In the light of the young age of the patient, the absence of previous malignant disease and the radiologic pictures, an infectious disease is supposed. However, clinical clues, the results of laboratory tests and all of the cultures obtained were negative, thus excluding an infectious etiologies. Then,

Wegener's granulomatosis is suspected, but this hypothesis is also ruled out because of negative immunological profile and of characteristics of lung lesions. Wegener's granulomatosis usually presents with cystic-cavitary lesions conversely to solid nodules presented in our case [4]. Because CT guided needle biopsy is inconclusive, the need to exclude a second primary tumor let us to perform a thoracoscopic biopsy. However, pre-operatory echocardiography document an unsuspected cardiac mass within right atrium. The diagnosis is more a favour for a cardiac tumor rather than thrombus and thus, no other exams as MRI or transesophageal echocardiography are required.

The second unusual aspect of the present case is that echocardiography surprisingly diagnoses a cardiac mass no detected by previous chest CT scan. Bic et al. [3] and Come et. [4] report a limited sensitivity of echocardiography in detecting cardiac tumor. In contrast, Lo et al. [5] find that CT may present lower sensitivity for endocardial lesion than echocardiography. However in the present case the potential discrepancy between CT and echocardiography is the result of an inaccurate evaluation. On retrospective reviewing of chest CT scan, a tumor is evident in the right atrium (black arrow in Figure 1), but it is missed initially. In theory, the lung lesions attract the attention of the observer who have not taken into account anything else as to say: *"the brain knows what the eyes want"*.

The treatment of cardiac angiosarcoma remains disappointing. Surgery may provide a longer survival in patients without metastases while heart transplantation is an attractive option but still limited [1]. At the time of diagnosis, our patient already has lung metastases and he is currently undergoing chemotherapy. Finally, cardiac angiosarcoma should be considered in the differential diagnosis of young patients presenting with lung lesions despite clinical symptoms are absent. In this case, echocardiography may help to show lesions less evident on CT scan in order to avoid a delay diagnosis and treatment.

Riassunto

Riportiamo il caso clinico di un paziente affetto da angiosarcoma cardiaco con metastasi multiple polmonari bilaterali. Al ricovero, il paziente lamentava tosse, sintomatologia per la quale aveva praticato la TAC del torace che mostrava lesioni multiple polmonari bilaterali. Gli esami laboratoristici e microbiologici non diagnosticavano alcun processo infettivo in atto; l'agoaspirato TAC guidato delle lesioni polmonari risultava negativo per la ricerca di cellule neoplastiche. Pertanto veniva programmata la biopsia toracoscopica delle lesioni polmonari alfine di ottenere la diagnosi. Prima di procedere all'intervento chirurgico, il paziente eseguiva l'ecocardiogramma che diagnosticava la presenza di una neoformazione sulla parete posteriore dell'atrio destro. Riesaminando nuovamente l'esame TAC, si concludeva che tale lesione, benché presente, non era stata precedentemente diagnosticata. Probabilmente, il drammatico quadro polmonare aveva catturato l'attenzione del refertatore che focalizzava la sua attenzione sulle lesioni polmonari mettendo in secondo piano ciò che era meno evidente, cioè l' aumento delle dimensioni del cuore, come dire: *"quando gli occhi ingannano il cervello!"*

Nel corso della toracoscopia eseguita a destra, si rinvenivano multiple neoformazioni localizzate sulla pleura parietale che venivano bioptizzate. L'esame immunoistochimico era il seguente: CK(-), CK7(-), EMA(-), ESA(-), CEA(-), TTF1(-), Vimentina(+), CD31(+), CD117(+) con diagnosi di metastasi da angiosarcoma. Attualmente, il paziente è in trattamento chemioterapico.

References

1. Kurian KC, Weisshaar D, Parekh H, Berry GJ, Reitz B: *Primary cardiac angiosarcoma: Case report and review of the literature.* Cardiovasc Pathology, 2006; 15:110-12.

2. Strohl KP: *Angiosarcoma of the heart.* Arch Intern Med, 1976; 136: 928-29.

3. Bic JF, Fade-Schneller O, Marie B, Neimann JL, Anthoine D, Martinet Y: *Case report: Cardiac angiosarcoma revealed by lung metastases.* Eur Respir J, 1994; 7:1194-96.

4. Come P, Riley M, Markis J, Malagold M: *Limitations of echocardiographic techniques in evaluation of left atrial masses.* Am J Cardiol, 1981; 48:947-53.

5. Lo FL, Chou YH, Tiu CM, Lan GY, Hwang JH, Chern MS, Teng MM: *Primary cardiac leiomyosarcoma: Imaging with 2-D echocardiography, electron beam CT and 1.5-Tesla MR.* Eur J Radiol, 1998; 27:72-76.