Case report

Cardiogenic shock due to metastatic cardiac lymphoma: still a diagnostic and therapeutic challenge

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Abstract

Myocardial involvement by metastatic lymphoma progressively leads to severe contractile impairment and fatal outcome. Correct diagnosis is often late due to misleading presentation signs. We report on a case of extensive cardiac involvement of a T-cell thymic lymphoma in a young woman, necessitating emergent extracorporeal membrane oxygenation (ECMO) circulatory support, with satisfactory hemodynamic recovery and subsequent ECMO weaning. Unfortunately, the following clinical course was rapidly fatal. This case seems to confirm that early aggressive instrumental diagnosis is crucial before severe myocardial impairment can prevent any therapeutic option. Extensive use of transesophageal echocardiographic examination and early endomyocardial biopsy are highly recommended.

Keywords: Cardiac lymphoma; Extracorporeal membrane oxygenation support; Cardiac surgery; Transesophageal echocardiography; Endomyocardial biopsy

1. Case report

A 36-year-old woman presented with exertional dyspnea and recent syncope. Physical examination showed moderate rest dyspnea, lower limbs edema, mild hepatomegaly, and lowered cardiac tones. The blood pressure was 110/60 mmHg, ECG showed sinus tachycardia at 120 beats/min, and a chest X-ray indicated moderate cardiomegaly. Trans-thoracic 2D color-Doppler echocardiography (TTE) detected relevant pericardial effusion with signs of cardiac tamponade and quite normal biventricular function. Percutaneous pericardial drainage achieved relief of symptoms. The recurrence of pericardial effusion and the detection of immature lymphoid elements in a sample of pericardial fluid raised the suspicion of primary non-Hodgkin lymphoproliferative malignancy. However, a whole-body CT scan failed to reveal significant regional lymphadenopathy and bone stem cells examination from the iliac crest was negative. Evidence at subsequent TTEs of moderate interventricular septum hypertrophy, being higher with recurrence of pericardial effusion, coexistence of supraventricular tachyarrhythmias with variable incidence of atrioventricular (AV) block, and the cyclic clinical course with recurrent fever suggested the hypothesis of a myocarditis of undiagnosed etiology. Eventually the patient was referred to surgery for pleuro-pericardial window and pericardial tissue biopsy. Soon after anesthesia was induced, a complete AV block occurred, requiring pharmacologic resuscitation with temporary recovery of normal AV conduction and satisfactory hemodynamics. A few minutes later a similar episode occurred; surgery was aborted and continuous intravenous inotropic support was instituted due to persistent low output syndrome (LOS). An intra-aortic balloon pump (IABP) was percutaneously inserted. Transesophageal 2D color-Doppler echocardiography (TEE) showed a grossly dilated and hypokinetic right ventricle (RV), a quite normal left ventricle (LV), moderate mitral and massive tricuspid regurgitation, and trivial pericardial effusion. Several intracavitary masses were detected at both sides of the interatrial septum and the anterior mitral valve leaflet (Fig. 1). Emergent cardiac catheterization excluded massive pulmonary embolism and coronary artery disease. Transvenous endomyocardial biopsy was obtained from the RV. Due to the persistence of cardiogenic shock, biventricular mechanical support by means of centrifugal pumps was scheduled. After median sternotomy, the pericardial layers were found to be diffusely thickened. The epicardial surface of the RV was involved in a wide proliferation of polypoid, friable, hemorrhagic tissue extending to the ante-
rior aspect of the great arteries, which were strictly adherent to the thymus and the soft mediastinal tissue. Specimens of pericardium, RV epicardium, and thymus with gross lymph nodes were collected. Arterovenous extracorporeal membrane oxygenation (ECMO) (Ultrox III membrane oxygenator, Avecor Cardiovascular, Inc., Plymouth, MN; Bio-pump, Bio-medicus, Minneapolis, MN) was instituted via the femoral vessels with progressive recovery of satisfactory hemodynamics, reversal of acidosis and anuria within the following 48 h. The histology of endomyocardial biopsy (Fig. 2) and surgical samples from both heart and thymus was consistent with metastatic cardiac involvement by large T-cell lymphoma, originating from the thymus and the soft surrounding tissue. After 72 h the patient was progressively weaned from the mechanical support under moderate catecholamines and IABP. Unfortunately, she rapidly worsened to severe hypotension, bradyarrhythmia and acidosis. ECMO could not be resumed.

At autopsy, both interatrial and interventricular septum were found to be diffusely spread with metastatic masses with multiple localizations at the level of AV node and both bundle branches. AV valve apparatus were involved as well.

2. Comment

This case presents different clinically misleading features in the setting of cardiac involvement by lymphoproliferative malignancies [1] and once more highlights the frequent late diagnosis and often suboptimal therapy [2]. Recurrent pericardial effusion is a common presentation sign; however, cytological evidence of malignancy has been proven to be negative in almost one-third of cases [3]. Moreover, the finding of immature or atypical lymphoid cells is difficult to differentiate from benign reactive lymphocytosis or other neoplasms. In a review of primary cardiac lymphoma, Ceresoli et al. [3] found that none of their 48 patients had bone marrow involvement and only five of them had regional evidence of the disease. Immunocytochemical, cytogenetic and polymerase chain reaction studies have proved useful in cases of suspicious pericardial fluid cytology and should be highly recommended.

Supraventricular arrhythmias with complete AV block have been recently described in a case of cardiac lymphoma [4]. As noted at autopsy, they reveal the intracavitary spread of tumor with multiple metastatic localizations close to the conduction system and its partial destruction. Successful treatment with a progressive reduction in the size of the intracavitary masses and complete remission of the conduction disturbance have been proven possible by means of aggressive radiotherapy and polychemotherapy after early TEE detection of intracardiac neoplasm. In our case, repeated TTEs failed to clearly detect intracavitary masses. Indeed, the finding of a variable degree of ventricular hyper-
Transvenous endomyocardial biopsy has led to non-surgical diagnosis of primary or secondary cardiac tumors [5]. Despite its sensitivity being affected by a significant number of false negatives [3], its early use could result in the timely detection of the neoplastic disease [6], allowing for a prompt utilization of oncologic and surgical strategies. Few reports deal with the latter options and their effectiveness appears strongly related to the timing of the diagnosis. Surgical excision of intracavitary tumor masses followed by aggressive chemotherapy has led to complete remission in selected cases [7]. Orthotopic cardiac transplantation appears to be indicated only in the presence of primary cardiac tumor, without extracardiac metastasis, and has resulted in either recurrence of the neoplasm in the graft or prohibitive incidence of infections due to the low immunosuppressive regimen adopted [8]. ECMO support for cardiogenic shock has been reported mostly in the case of acute myocarditis, with good results in terms of both a bridge to transplantation and myocardial recovery [9]. Its use in the case of cardiac malignancies remains questionable and we are fully aware of this strong limitation. However, the young age of our patient and the otherwise rapidly fatal outcome prompted us to use it as rescue therapy and as a first necessary step for subsequent oncologic treatment, which unfortunately could not be instituted because of the rapid hemodynamic worsening early after ECMO removal.

In conclusion, recurrent pericardial effusion, right-sided intracavitary masses and supraventricular arrhythmias should raise the suspicion of primary or metastatic cardiac lymphoma and justify the use of specific and sometimes invasive diagnostic procedures, considering that combined aggressive surgical and chemotherapeutic protocols have proven effective, providing that early diagnosis could be obtained.

References


Fig. 2. RV endomyocardial biopsy. (A) Transvenous endomyocardial biopsy specimen taken from the RV showing a diffuse neoplastic myocardial infiltration (hematoxylin–eosin ×45). (B) Close-up of (A): the myocytes appear dissociated by massive infiltration of atypical lymphoid cells (hematoxylin–eosin ×240). (C) CD3 immunohistochemical staining reveals a T-cell lymphoma (ABC ×480).