Primary cardiac lymphoma complicated by cardiogenic shock: successful treatment with chemotherapy delivered under extracorporeal membrane oxygenation support

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Abstract

Primary cardiac lymphomas (PCLs) are rare in immunocompetent patients. Their clinical presentation is highly variable and in case of cardiogenic shock, death is often inevitable with a diagnosis made post-mortem. We report the case of a 65-year old immunocompetent man with cardiogenic shock requiring emergent extracorporeal membrane oxygenation (ECMO). Soon after, a diagnosis of PCL was given and chemotherapy was delivered under ECMO support. The patient was progressively weaned from the mechanical support. Six months later, he had fully recovered.

Keywords: Primary cardiac lymphoma • Heart failure • Extracorporeal membrane oxygenation

INTRODUCTION

Primary cardiac lymphomas (PCLs) are rare in immunocompetent patients and in case of cardiogenic shock, death is often inevitable with a diagnosis made post-mortem. We report the case of a 65-year old immunocompetent man with cardiogenic shock requiring emergent extracorporeal membrane oxygenation (ECMO). Soon after, a diagnosis of PCL was given and chemotherapy was delivered under ECMO support. The patient was progressively weaned from the mechanical support. Six months later, he had fully recovered.

CASE REPORT


A few weeks later, the effusion had been reconstituted (Fig. 1A). In addition, a chest computed tomography revealed a pleural effusion and mediastinal nodes. The patient was then referred to our hospital.

The day after his transfer, he worsened haemodynamically because of rapid atrial fibrillation. A control transthoracic echocardiography confirmed recurrence of the circumferential pericardial effusion (Fig. 1B). Percutaneous pericardiocentesis was unsuccessful. It was then decided to carry out pleuro-pericardial window procedures through left thoracotomy.

Soon after anaesthesia was induced, 1 min of cardiac arrest occurred. The patient recovered following injections of adrenalin and cardiopulmonary resuscitation. Left thoracotomy evacuated 1 l of pleural effusion. The pericardium was thickened, and adhered to the myocardium. There was no fluid pericardial effusion. Pericardial biopsies were carried out for aetiological diagnosis. Despite intravenous inotropic support using dobutamine 15 gamma/kg/min, the patient’s haemodynamic status remained precarious. Arterial gazometry revealed a metabolic acidosis (pH = 7.17, $p_{O_2}$ = 158 mmHg, $p_{CO_2}$ = 62 mmHg; HCO$_3$ = 22 mmol/l, base excess = −7.3). A trans-oesophageal echocardiography did not find dilatation of the right heart, and excluded massive pulmonary embolism. Because of persistent cardiogenic shock with a subaortic velocity time integral of 8 cm, arterio-venous ECMO was applied to the femoral vessels by surgical approach to allow a distal perfusion of the leg. Emergent cardiac catheterization excluded a coronary aetiology.

The course of ECMO was uneventful. The patient did not present any respiratory problems. Unfractionated heparin anti-coagulation was conducted according to the department protocol with an activating clotting time of between 2 and 3. It is of note that despite the restoration of a satisfactory haemodynamic status without organ malperfusion signs, lactate levels remained high all along ~6 mmol/l. Finally, the persistence of hyperlactataemia without other signs of circulatory failure was related to the high-grade lymphoma.

While histological analysis of pericardium showed a dense lymphoid population B leading to suspicion of phenotype B small cell lymphoma, it did not allow for definitive diagnosis. Biopsies of mediastinal lymph nodes (Fig. 2A) were then performed by
mediastinoscopy under ECMO support. The diagnosis of high-grade B lymphoma was confirmed (Fig. 2B and C).

Emergent chemotherapy with cyclophosphamide, vindesine and solu-medrol was started on Day 8 of ECMO support. After 11 days, the patient was progressively weaned from the mechanical support. He was extubated the 20th day and left the intensive care unit after 26 days. A control transthoracic echocardiography found recovery of cardiac function exceeding 50%. The patient went home after 60 days. Six months later, he had fully recovered. Chemotherapy is still ongoing.

**DISCUSSION**

While cardiac involvement occurs in ~25% of patients with malignant lymphomas, PCLs are rare in immunocompetent patients, accounting for 1.3% of primary cardiac tumours and 0.5% of extranodal lymphomas [1].

According to Ceresoli et al. in 1997, PCL is defined as a lymphoma involving only the heart and/or pericardium. In the authors’ opinion, ‘only a single and asymptomatic extracardiac site of disease or minimal locoregional disease should be accepted as PCL’ [1].

The clinical presentation is highly variable. The most common presenting symptoms include precordial pains, cardiac tamponade and unresponsive heart failure [1, 2].

In case of cardiogenic shock, death is often inevitable as reported in several case reports [1, 2]. Rapid unfavourable evolution of the disease usually does not leave time to deliver diagnosis, which is usually given post-mortem [1].

Only in 67% of PCL patients with a clinically relevant pericardial effusion is a diagnostic cytological sample generally obtained [1].

When cytology is not available, less invasive diagnostic approaches such as endomyocardial transvenous biopsies or computed tomography-guided biopsies have been reported. Mediastinoscopy is another less invasive procedure, carrying out surgical macrobiopsies and facilitating histological analysis with a positive result of 100% [1].

The use of mechanical circulatory assistance in patients with lymphoma has been successfully reported in children in cases of post-chemotherapy toxicity cardiogenic shock [3] or in case of cardiopulmonary collapse in the setting of a diagnosis of an anterior mediastinal mass [4]. Aboud et al. [5] also reported the successful treatment of a lymphoma complicated by acute respiratory insufficiency due to compression of central airways using venovenous ECMO. As regards cardiogenic shock due to myocardial infiltration by lymphoma, the use of ECMO support was reported only once in adults, in 2001, by Testolin et al. [2]. Circulatory assistance was discontinued after 72 h, but without aetiological treatment using chemotherapy, the patient rapidly died from hypotension, bradycardia and acidosis.

Figure 1: (A) Chest computed tomography showing circumferential pericardial thickening and left pleural effusion. (B) Subcostal view in transthoracic echocardiography showing pericardial effusion of 2.17 cm (1) next to the right ventricle. RV: right ventricle; LV: left ventricle.

Figure 2: (A) Chest computed tomography showing the mediastinal node biopsied by mediastinoscopy (white arrow). (B) Haematoxylin-eosin stain showing plasmablastic cells (magnification ×40). (C) CD138-positive stain confirming the plasmablastic type of the lymphoma (magnification ×10).
As soon as the diagnosis of lymphoma was established, we decided to administer chemotherapy under ECMO. It allowed for successful withdrawal of assistance and patient survival with satisfactory recovery of cardiac function. Needless to say, ECMO in patients with uncontrolled neoplasia must be carried out with caution; in our case, however, we had already provided effective assistance before the diagnosis of lymphoma. Without the combination of the two therapeutics, haemodynamic support and chemotherapy, the patient probably would not have survived.

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REFERENCES