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Obstructive shock secondary to an unusual cause: primary cardiac lymphoma

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Abstract

Background The medico-surgical management of cardiac tumors when there is a suspicion of malignancy is complex. Moreover, in a critically ill setting, the choice of diagnostic tools seems crucial.

Case presentation We present the case of a sixty-four-year-old patient with no prior medical history who was admitted to the intensive care unit with obstructive shock secondary to a right heart mass and pulmonary embolism. Clinical and biological features revealed secondary hemophagocytic lympho-histiocytosis (HLH). The patient underwent a diagnostic procedure with dual thoracoscopic and mediastinoscopy with biopsies. Following the diagnosis of primary cardiac lymphoma, the patient received sequential chemotherapy without undergoing cardiac surgery leading to initial improvement, with reductions in intracardiac obstruction and in biomarkers associated with hemophagocytic lympho-histiocytosis.

Conclusion When a cardiac mass is associated with extracardiac symptoms indicative of a hematological malignancy, the preferred treatment is chemotherapy, and cardiac surgery should be avoided.

Background

Primitive cardiac tumors (PCTs) are rare pathologies (30 cases/million people/year) [1] and are predominantly benign (90% of cases). In PCT, the diagnostic procedure is challenging and should follow a thorough process. PCT clinical images can be separated into (1) general or extracardiac symptoms, (2) cardiovascular symptoms, and (3) embolic events [1]. Clinical manifestations are highly related to the underlying disease. The diagnostic procedure for mediastinal and cardiac tumors should be multimodal, based on imaging (echocardiography, Computed Tomography and Magnetic Resonance Imaging) and surgical thoracic procedures [1–3] for anatomopathological analysis. Currently, minimally invasive surgical endoscopic techniques (mediastinoscopy or thoracoscopy) are considered the gold standard. PCTs are malignant in only 10% of cases, with sarcoma as the predominant disease [4]. Cardiac localization in lymphoma is a rare extra nodal involvement, which may occur as part of either disseminated or isolated disease [5]. In a North American cohort from a thirty-year study period, in ten million cancer patients, 184 had primary cardiac lymphoma. The management of critically ill patients with severe clinical manifestations of PCT is poorly reported, and there are no guidelines. We present here the case of a critically ill patient with obstructive shock due to primary cardiac lymphoma initially successfully managed by diagnostic thoracoscopy and chemotherapy.

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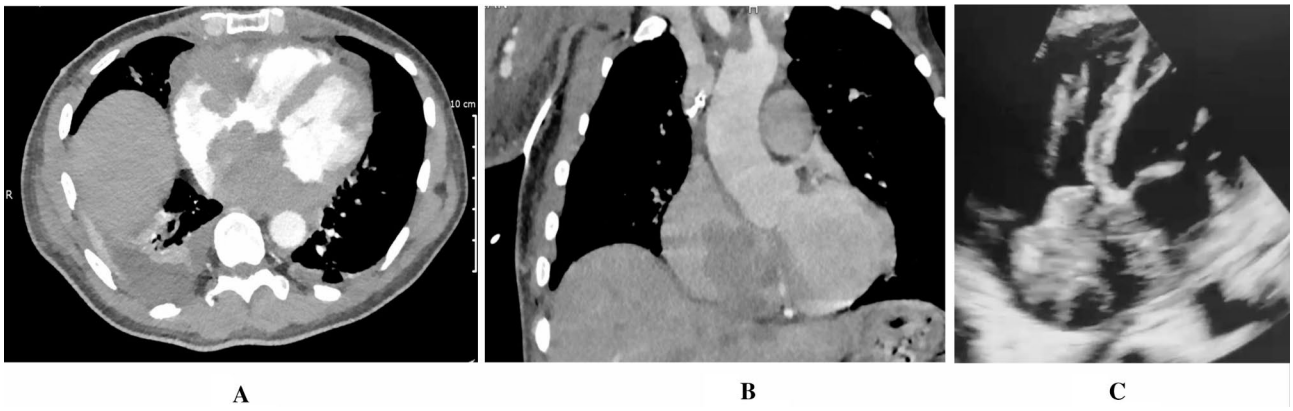

Fig. 1 1/A et B Thoracic CT demonstarating a voluminous intra cardiac mass 1C/ Transthoracic echocardiography view depicting the intra right atrium tumour with tricuspid and septal involvement

Case presentation

A 64-year-old patient without a significant medical history was admitted for hypotension (93/55mmHg), tachycardia (blood pulse 145/min), and acute dyspnea in our intensive care unit. In the previous weeks, the patient had general symptoms consisting of fever, night sweats, and weight loss (above 10% of his weight), without syncope or dyspnea. Initial clinical examination revealed symptoms of systemic inflammatory response syndrome (tachycardia, polypnea and hyperthermia) and both right and left heart failure (jugular venous distension, cool extremities, and thready pulse). Electrocardiogram revealed a tachycardia at 150/min, with Atrial fibrillation and a right bundle branch block. At ICU admission, an electrocardiogram revealed sinus rhythm with right bundle branch block. Blood test showed, leukocytosis (16G/L), anemia (7.6 g/dl), hyperlactatemia (4.6mmol/L), normal kidney function, normal hepatic function. Elevated levels were observed for (i) Protein C reactive (343 mg/L), (ii) liver enzymes (AST:234UI/L, ALT:123UI/L), (iii) troponin (0.163ng/ml), (iv) lactate deshydrogenase (234UI/L) and (v) ferritin (29 106 ng/ml). Thorax CT (Fig. 1A and B) revealed unilateral right segmentary pulmonary embolism without right ventricular distension, cardio-phrenic mediastinal infiltration, and an intracardiac mass with pericardial infiltration. No thoracic or abdominal tumoral syndrome was observed. Echocardiography (Fig. 1C) revealed a heterogeneous right atrial mass measuring 60×40 mm with endocardial extension and tricuspid invasion. The patient had a dilated inferior vena cava (24 mm), Right Ventricular (RV) impairment with underfilling and intracardiac-obstructive features (elevated right atrium–RV gradient>10 mmHg, mean pulmonary arterial pressure at 40 mmHg and no signs of tricuspid regurgitation) (Fig. 1B). The mass extensively infiltrated the atrial septum with RV prolapse, without any visible thrombus. Pulmonary embolism was considered non massive based on the unilateral status and the absence of RV distension. Blood cultures and virological serologies were negative. Hemophagocytic lympho-histiocytosis (HLH) suggested by biological test was confirmed by the presence of hemophagocytic figures on the myelogram cytological analysis (Fig. 2). Notably, no tumor cells were observed. We therefore concluded that the patient had a mixed shock consisting of obstructive shock and vasoplegia associated with HLH). The patient was initially treated with broad-spectrum antibiotics, vasopressors, curative anticoagulation, and amiodarone. On day 2, the patient underwent a dual surgical procedure involving video thoracoscopy and mediastinoscopy with paracardial mediastinal biopsies. Anatomopathological tests yielded the diagnosis of primitive cardiac anaplastic T-cell lymphoma with mediastinal involvement. Following the diagnostic procedure, the patient was treated with sequential chemotherapy, leading to hemodynamic improvement (reduce need for vasopressors

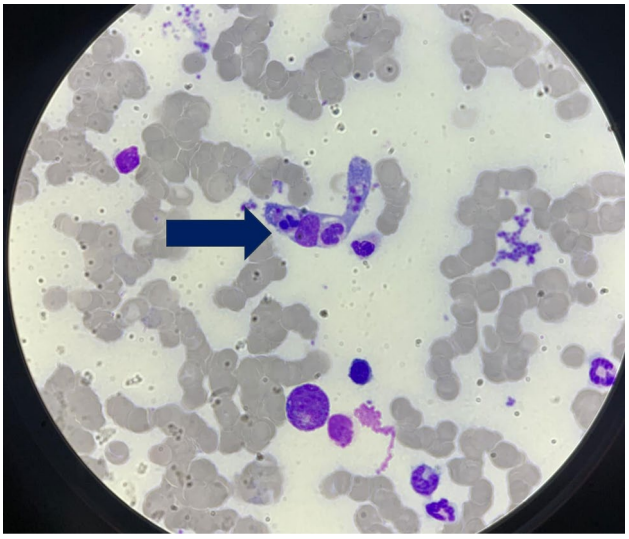

Fig. 2 Bone marrow aspirate stained with Wright-Giemsa of intracellular red blood cells macrophage (black arrow)

and decreased intracardiac gradient) and a reduction in biological markers of HLH. The patient was weaned from vasopressors on day 5 and discharged from the ICU on day 8. At 30 days after the first chemotherapy, echocardiographic control revealed a decrease in the size of the right heart tumor. However, he finally died 6 weeks later of a refractory septic shock.

Discussion and conclusions

Diagnostic management of cardiac tumors is multimodal and is based on the location of the tumor, clinical pictures and imaging data (transthoracic echocardiography, cardiac MRI, positron emission tomography) [1]. Therapeutic management is based on the suspected diagnosis. Primary cardiac lymphoma (PCL) is a rare form of extranodal lymphoma (1% PCT), with only 490 described cases worldwide [4–6]. In the last twenty years, PCL has been used for patients with PCTs with general symptoms and features of hematological malignancy (HLH). In PCL, echocardiography revealed mostly homogeneous infiltrating masses with predominant right heart locations.

HLH is a life-threatening disorder characterized by high fever, biological abnormalities and vasoplegic shock due to cytokine storms [7], with hematological malignancies as the main etiologies. The therapeutic efficacy of HLH is related to the underlying disease.

In the absence of extracardiac tumoral syndrome, endoscopic thoracic procedures (dual thoracoscopy and mediastinoscopy) were necessary for histological diagnosis [8] and staging in our patient. The patient had simultaneous obstructive shock due to right heart lymphoma obstruction and vasoplegia related to HLH [7]. Emergency chemotherapy is expected to improve HLH vasoplegia and tumoral obstruction [9]. The management of critically ill patients with PCL has rarely been described [10], and the described therapeutics are highly variable. Histological diagnosis is crucial, as chemotherapy is the cornerstone of aggressive lymphoma treatment [9].

In summary, we reported a complex case of shock in a patient with PCL. Despite high severity at admission, medical treatment based on supportive care and emergency chemotherapy was initially successful in our patient. In PCL, chemotherapy is the only factor associated with survival [7, 9], and aggressive surgical management is associated with poor outcomes.

The therapeutic effects on cardiac parameters should be cautiously monitored via repeated echocardiography.

Author contributions
GO, LC and JDP participated in the study design, research, and manuscript writing. LDT and CF were involved in editing the manuscript and performing the research. All the authors have read and approved the final version of the manuscript.

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Data availability
No datasets were generated or analysed during the current study.

Declarations

Ethics approval and consent to participate
Informed consent for participation was obtained from the patient and the next of kin.

Consent for publication
Consent was obtained from the patient for the publication of this report and any accompanying images.

Competing interests
The authors declare no competing interests.

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