



COMPLETE ATRIOVENTRICULAR BLOCK ASSOCIATED WITH PERICARDIAL EFFUSION REVEALING PRIMARY CARDIAC LYMPHOMA:

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Abstract : Primary cardiac lymphoma is a rare pathology, representing 1% of primary cardiac tumors. It often involves the right heart and preferentially the atria to the ventricles. Through our observation, we propose to remind a complication although exceptional which can reveal the primary cardiac lymphoma that is the auriculo ventricular block.

Key words : Complete atrioventricular block , cardiac lymphoma, pericardial effusion.

I. Introduction :

Primary cardiac tumors are rare, about 0.001-0.030% [1]. They are in the majority of cases benign, represented essentially by myxoma. When they are malignant, they are mainly sarcoma and rarely lymphoma.

Primary cardiac lymphoma is therefore a rare entity with a poor prognosis, requiring rapid diagnosis and appropriate management.

We report in this article the observation of a primary cardiac tumor probably lymphomatous revealed by a pericardial effusion and paroxysmal conduction disorders.

II. Case report :

The patient was 84 years old, with hypertension. She was admitted to the emergency room for the management of asthenia with weight loss.

The clinical examination on admission was unremarkable. The biology noted an inflammatory syndrome CRP 145, LDH elevated to 435, a normal troponin and a neutrophilic hyperleukocytosis with a slight anemia. Renal and liver function were normal.

Chest X-ray showed mediastinal enlargement with cardiomegaly.

The 12-lead electrocardiogram revealed a 1st degree atrioventricular block. The patient was monitored, after a moment it became complete then after a few hours it became BAV 2nd degree Mobitz 1.

The transthoracic echocardiography revealed a periaortic mass with a large pericardial effusion and collapsed right cavities. The effusion was drained and found to be serohematically exudative, with non-contributory culture.

CT scan showed a tissue mass encompassing the root of the aorta with endocavitary extension of both atria suggesting a lymphomatous origin.

Cardiac MRI revealed a mass invading the interatrial septum and surrounding the initial aorta with homogeneous irregular contours measuring 42x17 mm and heterogeneously enhancing after gadolinium injection.

The patient refused biopsy of the cardiac mass. She was therefore implanted with a permanent single-chamber pacemaker with close monitoring of the reconstitution of her pericardial effusion. The patient died 1 month after the diagnosis of her cardiac lymphoma.

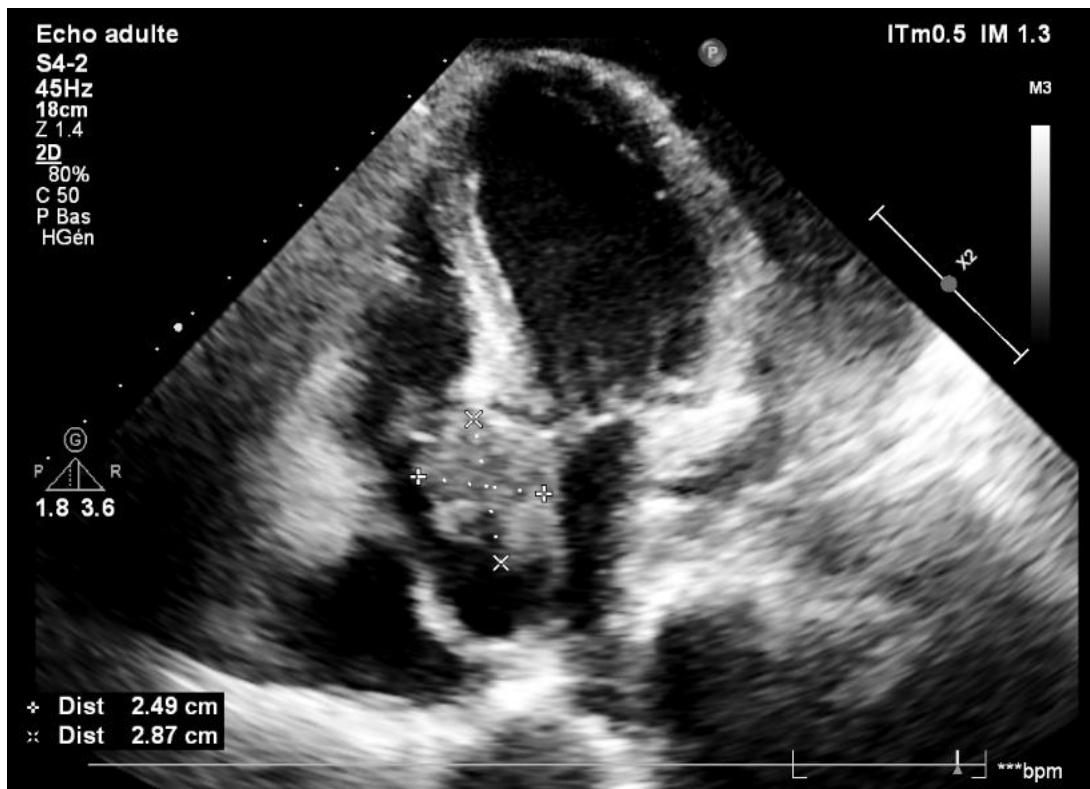


Figure 1 : Transthoracic echocardiography: apical 4-chamber section showing intra-OD periaortic mass with pericardial effusion.

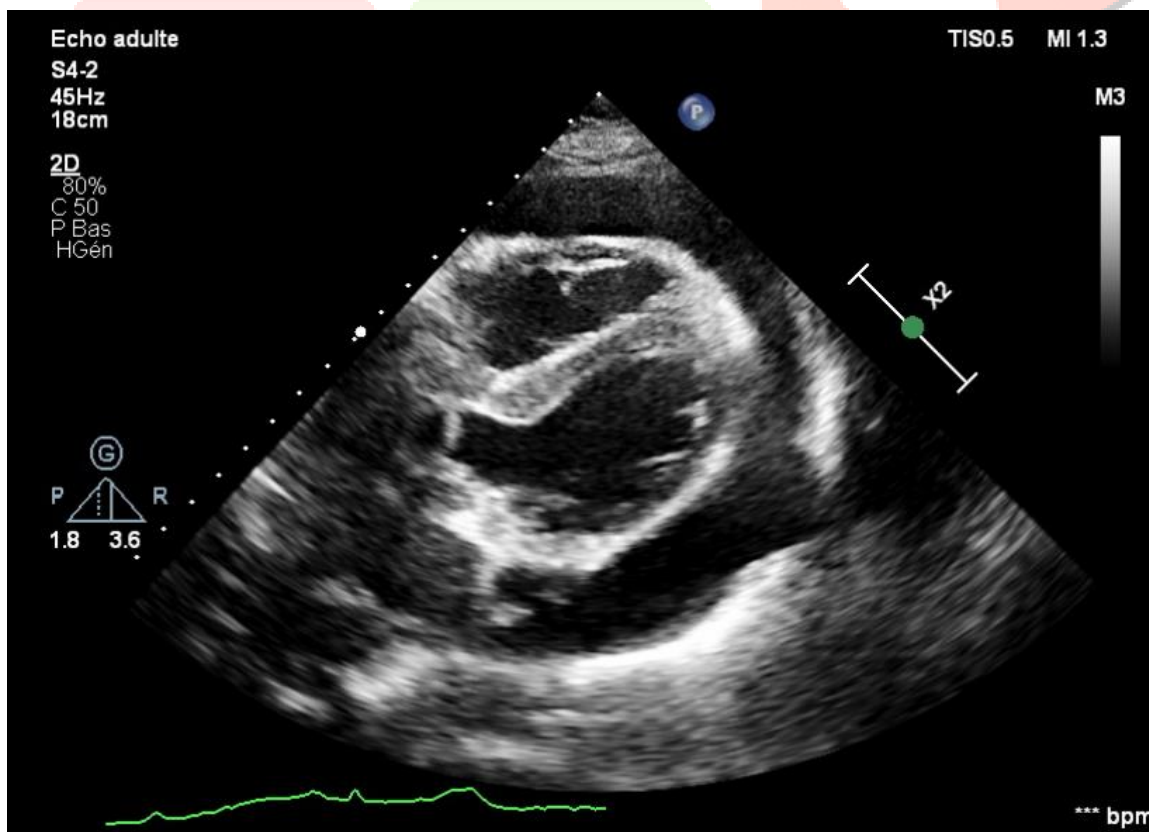


Figure 2 : Transthoracic echocardiography: Sub-costal section showing a circumferential pericardial effusion of great abundance.



Figure 3 : Thoracic CT: Axial section through the mediastinal window after injection of contrast medium showing a well-limited inter aortic-pulmonary mass with moderate enhancement measuring 80mm*60mm with pericardial effusion.

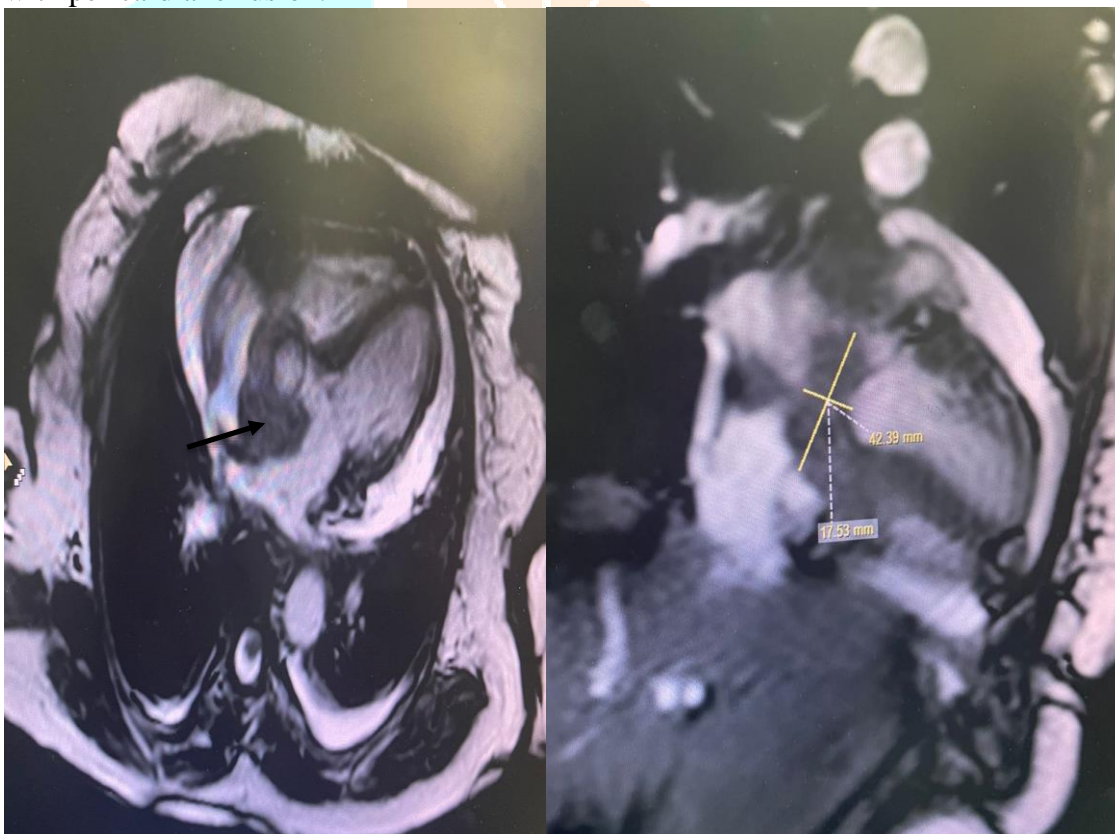


Figure 4 : Thoracic magnetic resonance imaging. Tumor infiltration of the interatrial septum and the initial aorta.

III. Discussion :

Primary cardiac lymphoma is a rare disease, representing 1 to 2% of primary cardiac tumors [2]. It mainly affects immunocompromised subjects, especially HIV-infected or transplanted patients, which is not the case in the reported observation.

PCL affects both sexes equally from the age of 50. It mainly affects the right cavities and more often the atria to the ventricles.

The clinical picture is variable. Dyspnea is in the first place [3] related to myocardial damage, then arrhythmias which could be due to tumor infiltration of the roof of the lateral wall of the right atrium and also conduction disorders related to tumor infiltration of the inter-atrial septum as in the case of our patient

where the diagnosis of PCL was suspected following paroxysmal AVB sometimes 1st degree, sometimes 2nd degree, sometimes complete. Pericardial effusion or tamponade can also be indicative of a neoplastic pathology, in particular PCL, especially if it is associated with an altered general condition [4,5,6].

The biological parameters are aspecific and not very contributory. An inflammatory syndrome with elevated CRP, LDH and ferritinemia is sometimes found [7,8]. Abnormalities of the haemogram have also been reported, notably anaemia with neutrophilic hyperleukocytosis [9,10]. A discrete disturbance of the liver balance has been reported in some observations of PCL [11,12].

Chest radiography is non-specific and of limited interest. It may be normal or show cardiomegaly [13, 14] or right atrial hypertrophy [15].

The ECG may reveal several abnormalities, including rhythm and conduction disorders, including complete atrioventricular block, which is most frequently found in patients with PCL and which is generally regressive after treatment of the tumor with chemotherapy [16,17].

Cardiac imaging plays an important role in the diagnosis of cardiac lymphoma.

Transthoracic echocardiography (TTE) is the first-line examination, and it allows to demonstrate the cardiac involvement, its characteristics and the tunica involved [4-18]. The tumor can be either single or multiple, endocavitary or located on the valvular endocardium. All three tunics can be affected, however pericardial involvement is the most common and often the right cavities are most affected [19-20-21]. Pericardial effusions are frequent, sometimes of great abundance, responsible for tamponade and may be indicative of the tumor [4-5-6-19]. TTE also plays an important role in monitoring the therapeutic response to judge its effectiveness. In addition, in certain conditions, TEE is used, which allows a more precise study and produces high-resolution images to better characterize the involvement [6].

The CT scan also contributes to the diagnosis but is especially useful in the extension assessment [9-22].

Finally, cardiac magnetic resonance imaging is currently the examination of choice for the diagnosis of cardiac lymphoma with a sensitivity of nearly 90% [20]. It allows a precise analysis of the tumor extension and a rigorous post-therapy follow-up [5-23].

However, the diagnosis of certainty relies on the biopsy of cardiac tissue, especially when the tumor is primary or when the cytological study of the pericardial effusion is not contributive. It can be done either by surgical sternotomy [2-5], by scannoguided transthoracic approach [24] or by endomyocardial approach [4-5]. Our patient refused biopsy and histological confirmation could not be obtained.

Treatment of primary cardiac lymphoma usually involves chemotherapy, but it is still not codified and depends on the experience of each team.

Surgery is indicated in rare cases, in the presence of mechanical complications or right heart failure [5-18] and as part of palliative treatment if the tumor is very large [25].

The prognosis of primary cardiac lymphomas is often poor, the main reason being the delay in diagnosis [1-18-24].

IV. Conclusion :

Cardiac lymphomas are rare, often diagnosed late due to their highly variable and non-specific clinical presentations. MRI is the reference examination allowing a better characterization of the cardiac involvement and also a monitoring of the response to treatment. The confirmatory diagnosis is anatomopathological.

Treatment is based on chemotherapy, but the short-term prognosis remains very poor because of the delay in diagnosis and the poorer response to treatment compared to other lymphomatous tumors.

Références :

- [1] Centofanti P, Di Rosa E, Deorsola L, Actis Dato GM, Patane F, La Torre M, et al. Primary cardiac neoplasms: early and late results of surgical treatment in 91 patients. *Ann Thorac Surg* 1999;68:1236–41.
- [2] Miguel CE, Bestetti RB. Primary cardiac lymphoma. *Int J Cardiol* 2011;149:358–63.
- [3] Petrich A, Cho SI, Billett H. Primary cardiac lymphoma. *Cancer*. 2011;117(3):581–589.
- [4] Mikdame M, Ennebi K, Bahrouch L, Benyass A, Dreyfus F, Touloune F. Localisations cardiaques du lymphome non hodgkinien : à propos de quatre cas. *Rev Med Interne* 2003;24:459–63.
- [5] Fuzellier JF, Saade YA, Torossian PF, Bæhrel B. Primary cardiac lymphoma: diagnosis and treatment. Report of 6 cases and review of the literature. *Arch Mal Cœur Vaiss* 2005;98:875–80.
- [6] Mioulet D, Bræm L, Heno P, Paule P, Peloni JM, Bonnet D, et al. Flutter atrial révélateur de l'extension cardiaque d'un lymphome malin non hodgkinien. *Ann Cardiol Angeiol* 2008;10:1016–20.
- [7] Sato Y, Matsumoto N, Kinukawa N, Matsuo S, Komatsu S, Kunimasa T, et al. Successful treatment of primary cardiac B-cell lymphoma: depiction at multislice computed tomography and magnetic resonance imaging. *Int J Cardiol* 2006;113:E26–9.
- [8] Hwang YY, Fan K, Lam YM, Chan WS, Cheung S, Wang EP, et al. Primary cardiac lymphoma presenting with right heart mass and bradycardia. *Ann Hematol* 2007;86:685–6.
- [9] Giunta R, Cravero RG, Granata G, Sellito A, Romano C, De Fanis U, et al. Primary cardiac T-cell lymphoma. *Ann Hematol* 2004;83:450–4.
- [10] Daus H, Bay W, Harig S, Schneider G, Feiden W, Schieffer H. Primary lymphoma of the heart: report of a case with histological diagnosis of the transvenously biopsied intracardiac tumor. *Ann Hematol* 1998;77:139–41.
- [11] Nart D, Nalbantgil S, Yagdi T, Yilmaz F, Hekingil M, Yüce G, et al. Primary cardiac lymphoma in a heart transplant recipient. *Transplant Proc* 2005;37:1362–4.
- [12] Trost B, Croft L, Nair A, Henzlova M. Primary cardiac lymphoma detected by myocardial perfusion imaging: case report. *J Nucl Cardiol* 2007;14:e6–10.
- [13] Medolago G, Virota G, Piti A, Tespili M, D'Adda F, Rottoli MR, et al. Abnormal uptake of technetium-99m hexakis-2-methoxyisobutylisonitrile in a primary cardiac lymphoma. *Eur J Nucl Med* 1992;19:222–5.
- [14] Sommers KE, Edmundowicz D, Katz WE, Hattler BG. Primary cardiac lymphoma: echocardiographic characterization and successful resection. *Ann Thorac Surg* 1996;61:1001–3.
- [15] Tai CJ, Wang WS, Chung MT, Liu JH, Chiang CY, Fan FS, et al. Complete atrioventricular block as a major clinical presentation of the primary cardiac lymphoma: a case report. *Jpn J Clin Oncol* 2001;31:217–20.
- [16] Deepti AN, Noone ML, Mahadevan A, Naresh KN, Yasha TC, Satishchandra P, et al. Primary cardiac cytotoxic T-cell lymphoma presenting with neurological deficits: a case report. *Cardiovasc Pathol* 2008;17:334–8.
- [17] Bulum J, Banfic L, Strozzi M, Aurer I, Jelasic D. Primary cardiac lymphoma presenting as atrial flutter and total heart block. *Heart Vessels* 2007;22:52–4.
- [18] Donal E, Coisne, Corbi P, Chritæns L, Menet E, Allal J, et al. Cardiac lymphoma disclosed by tamponade and complete atrioventricular block: apropos of a surgically treated case. *Heart and lymphoma. Ann Cardiol Angeiol* 1997;46:667–70.
- [19] Msaad S, Yangui I, Ketata W, Ayadi H, et al. Manifestations thoraciques révélatrices de lymphomes : a propos de 17 cas. *La Tunisie Médicale*. 2008; 86(2):105-13. PubMed | Google Scholar
- [20] Ceresoli GL, Ferreri AJ, Bucci E, Ripa C, Ponzoni M, Villa E. Primary cardiac lymphoma in immunocompetent patients: diagnostic and therapeutic management. *Cancer* 1997;80:1497–506.
- [21] Cracowski JL, Trémel F, Nicolini F, Bost F, Mallion JM. Myocardial localization of malignant non-Hodgkin lymphoma responsive to chemotherapy. *Arch Mal Cœur Vaiss* 1997;90:1527–31.
- [22] Binder J, Pflieger S. Schwarz. Images in cardiovascular medicine. Right atrial primary cardiac lymphoma presenting with stroke. *Circulation* 2004;110:e451–2.
- [23] O'Donnell DH, Abbara S, Chaithiraphan V, Yared K, Killeen RP, Cury RC, et al. Cardiac tumors: optimal cardiac MR sequences and spectrum of imaging appearances. *AJR Am J Roentgenol* 2009;193:377–87.
- [24] Roubille F, Massin F, Cayla G, Gahide G, Gervasoni R, Macia JC, et al. Intra-cardiac lymphoma with right heart failure: a therapeutic emergency in two patients. *Arch Mal Cœur Vaiss* 2007;100:1025–9.
- [25] Grantomo J, Pratita J, Rachmat J, Saraswati M. A rare case of primary cardiac lymphoma and the role of early surgical debulking: a case report. *Eur heart J Case Rep* 2018;2:yt116.