

Primary cardiac lymphoma (PCL) – diagnostic difficulties



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Abstract

Primary cardiac lymphoma (PCL) is the very rare disease that is associated with a high mortality rate. A prompt and proper diagnosis may affect the prognosis, and proper treatment may improve life expectancy. This report documents the case of a 74-year-old female with primary cardiac lymphoma. Unfortunately, the patient died from heart failure on her 23rd day in hospital.

Key words: primary cardiac lymphoma, heart tumour, echocardiography.

Streszczenie

Pierwotny chłoniak serca jest rzadkim schorzeniem, charakteryzującym się wysoką śmiertelnością. Rokowanie poprawia szybka i właściwa diagnoza oraz leczenie. W pracy przedstawiono przypadek 74-letniej pacjentki z pierwotnym chłoniakiem serca. Niestety, kobieta zmarła w 23. dobie pobytu szpitalnego w przebiegu niewydolności serca.

Słowa kluczowe: pierwotny chłoniak serca, guz serca, echokardiografia.

Introduction

Primary cardiac neoplasms are rare, with an incidence of 0.056% of all tumours seen in autopsies. Twenty-five percent of these cardiac neoplasms are malignant [1, 2]. Primary cardiac lymphoma (PCL) constitutes 5-6% of primary cardiac malignant neoplasms [1, 3]. Because of its rapid progress, late diagnosis due to its non-specific symptoms and variable patient responses to treatment, the prognosis for patients with PCL remains poor. Complete remissions have been reported, although the longest period of survival has not exceeded 144 weeks [4]. Approximately 80% of patients live less than 12 months after diagnosis [5].

Case report

A 74-year-old woman was admitted to the Department of Internal Medicine because of exertional dyspnoea and a fever which had lasted for two months. On admission, the physical examination revealed nothing remarkable except for an elevated body temperature (37.6°C). Abnormalities in the laboratory test results included: Hb 10.5 g/dl, WBC 10.6 x 10⁹/l, CRP 71 mg/l, ESR 47/86 mm, total bilirubin

1.89 mg/dl, AST 46 IU/l, ALT 74 IU/l, and GGTP 128 IU/l. An electrocardiogram showed a sinus rhythm of 90/min, and negative T-waves in II, III, aVF, and V3-V6. The chest X-ray revealed cardiomegaly. Both the transthoracic (TTE) and the transoesophageal (TEE) echocardiographs indicated the presence of a large right-atrial tumour (Fig. 1). A computed tomography (CT) of the heart and large vessels revealed a polycyclic, infiltrating nodular mass with non-homogenous enhancement after contrast administration encompassing the right atrium, right ventricle, and in the pericardial sac with pericardial effusion, with an associated mediastinal lymphadenopathy (Fig. 2A and 2B). Three months earlier the patient had been hospitalised in the Department of Cardiology due to atrial fibrillation. At that time, the TTE did not show any of the right-sided masses.

With a presumptive diagnosis of a cardiac sarcoma or lymphoma, the patient was disqualified from pericardiocentesis due to the presence of the tumour mass at the puncture site, and from a transjugular biopsy due to the extensive thrombosis of both jugular veins. A transcervical mediastinoscopy was performed under general anaesthetic, and several lymph node stations (3, 4R, 4L and 10 R)

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were biopsied. The pathological examination only revealed chronic lymphadenitis. The patient's condition gradually deteriorated and she died from heart failure on her 23rd day in hospital. An autopsy was performed and the post-mortem examination of the tumour revealed a cardiac B-cell lymphoma.

Discussion

A primary cardiac lymphoma is defined as a non-Hodgkin's lymphoma involving only the heart and/or pericardium or a lymphoma with the bulk of the tumour within the heart [6, 7]. More than 50% of patients with PCL suffer from heart failure that is unresponsive to therapy. The classic symptoms of PCL include dyspnoea, oedema, weakness and fatigue, pericardial effusion, arrhythmia, chest pain, weight loss, fever, and night sweats [4, 7]. Due to the low prevalence of PCL, its treatment is not established. Treatment approaches include combinations of: chemotherapy, radiotherapy, autologous stem cell transplant, and tumour mass resection [4]. The median of the survival range is from 1.5 months with the surgical approach to 26.5 months with chemotherapy combined with radiotherapy [4]. The average survival rate is extremely short in untreated patients [4].

The procedures aimed at establishing a diagnosis vary considerably in sensitivity and invasiveness. In the case documented here, a mediastinoscopy with lymph node sampling was performed, but it only revealed chronic inflammation. In view of the high risk of heart failure, other procedures could not be performed. There are two descriptions of a similar diagnostic path; in both cases a mediastinoscopy yielded the diagnosis [7, 8]. Some procedures, such as an open-heart biopsy, have 100% sensitivity but are associated with a high risk of serious complications in patients with uncontrolled heart failure [4, 7]. Pericardial



Fig. 1. Transoesophageal echocardiography – obliteration of the right atrium by the structure of echogenicity similar to the myocardium, causing obstruction of the tricuspid valve and descending to the right ventricle

cytology is diagnostically effective in 60-67% of cases, but pericardial effusion is only present in 12-49% of patients [4, 7, 9]. A pericardial biopsy has a reported diagnostic yield of 38-50% [7, 9]. There are individual reports of other procedures used to diagnose PCL such as transoesophageal ultrasound-guided biopsy, transvenous endomyocardial biopsy, percutaneous intracardiac biopsy with combined fluoroscopy, and transoesophageal echocardiographs [6, 9]. Their sensitivity remains unproven.

Conclusions

In conclusion, prompt diagnosis and treatment management maximise prolonged survival in PCL. Therefore, the appropriate choice of diagnostic procedure influences the patient's prognosis [2, 4, 9]. Unfortunately, the range of

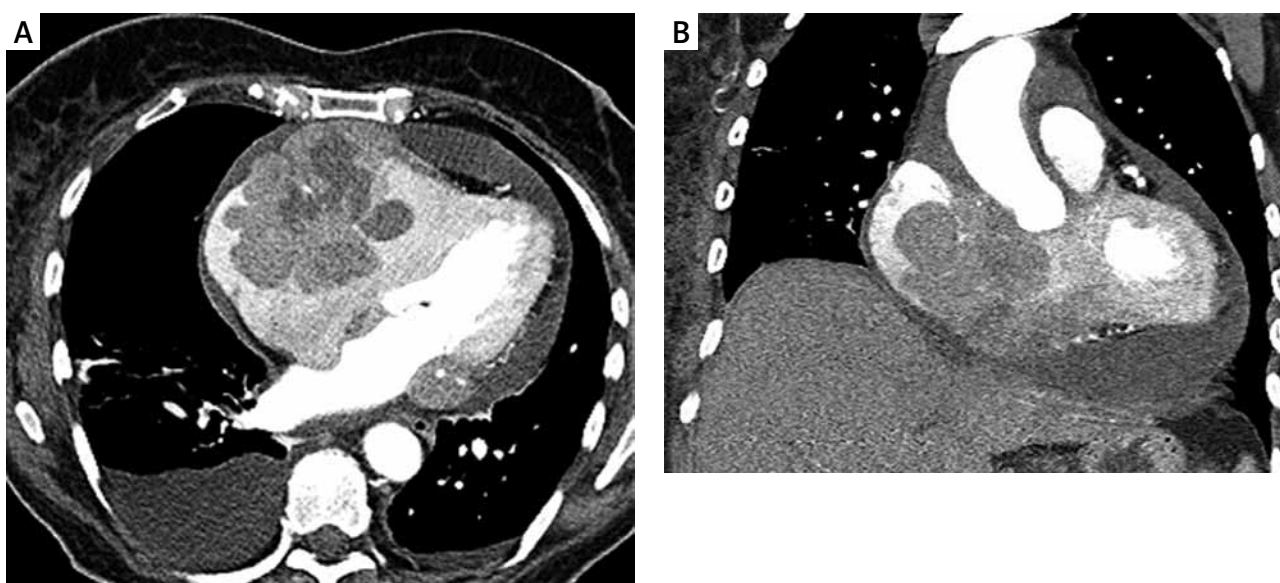


Fig. 2. Computed tomography of the heart and large vessels: a polycyclic, non-measurable, infiltrating nodular mass undergoing non-homogenous enhancement in the cavity of the right atrium, extending into the right ventricle, pericardium, and myocardium of the left atrium. There is also a pericardial effusion. **A)** Multiplanar reconstruction – axial view. **B)** Multiplanar reconstruction – anterior view

modern diagnostic approaches may be seriously limited in an advanced stage of PCL and when a patient's condition on admission is critical.

Disclosure

Authors report no conflict of interest.

References

1. Neragi-Miandoab S, Kim J, Vlahakes G. Malignant tumours of the heart: a review of tumour type, diagnosis and therapy. *Clin Oncol* 2007; 19: 748-756.
2. Nascimento AF, Winters GL, Pinkus GS. Primary cardiac lymphoma: clinical, histologic, immunophenotypic, and genotypic features of 5 cases of a rare disorder. *Am J Surg Pathol* 2007; 31: 1344-1350.
3. William CR. Primary and secondary neoplasms of the heart. *Am J Cardiol* 1997; 80: 671-682.
4. Anghel G, Zoli V, Petti N, Remotti D, Feccia M, Pino P, Majolino I. Primary cardiac lymphoma: report of two cases occurring in immunocompetent subjects. *Leuk Lymphoma* 2004; 45: 781-788.
5. Ikeda H, Nakamura S, Nishimaki H, Masuda K, Takeo T, Kasai K, Ohashi T, Sakamoto N, Wakida Y, Itoh G. Primary lymphoma of the heart: case report and literature review. *Pathol Int* 2004; 54: 187-195.
6. Rolla G, Bertero MT, Pastena G, Tartaglia N, Corradi F, Casabona R, Motta M, Caligaris-Cappio F. Primary lymphoma of the heart. A case report and review of the literature. *Leuk Res* 2002; 26: 117-120.
7. Ceresoli GL, Ferreri AJM, Bucci E, Ripa C, Ponzoni M, Villa E. Primary cardiac lymphoma in immunocompetent patients. *Cancer* 1997; 80: 1497-1506.
8. Bambury R, Gallagher F, Dodd JD, Fennelly D. Primary cardiac lymphoma: diagnostic tools and treatment challenges. *Ir J Med Sci* 2011; 180: 271-273.
9. Chalabreysse L, Berger F, Loire R, Devouassoux G, Cordier J, Thivolet-Bejui F. Primary cardiac lymphoma in immunocompetent patients: a report of three cases and review of the literature. *Virchows Archiv* 2002; 441: 456-461.