Primary cardiac Burkitt-like lymphoma of the right atrium

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A 74-year-old man presented for shortness of breath. Echocardiography revealed the presence of a large pericardial effusion with signs of tamponade. A right atrial mass was suspected and later confirmed by transesophageal echocardiography. The mass was attached to the right side of the interatrial septum. Surgical resection was performed. Histology was compatible with a diagnosis of undifferentiated B-cell non-Hodgkin's (Burkitt-like) primary cardiac lymphoma. The present report provides the first description of a Burkitt-like primary cardiac lymphoma. The presence of a mass in the right atria should raise the possibility of a malignant cardiac tumour. Transesophageal echocardiography should be considered as the initial diagnostic procedure to be performed. Rapid histological diagnosis is important because systemic therapy can influence prognosis in the presence of a primary cardiac lymphoma.

Key Words: Cardiac lymphoma; Cardiac tumours; Transesophageal echocardiography

Un lymphome cardiaque primaire de Burkitt atypique dans l'oreillette droite

Un homme de 74 ans a consulté pour cause de dyspnée. L'échocardiographie a révélé la présence d'une large effusion péricardique accompagnée de signes de tamponnade cardiaque. On a présumé la présence d'une masse auriculaire droite qui a ensuite été confirmée par échocardiographie transœsophagienne. La masse était fixée au côté droit de la cloison interauriculaire et a été réséquée par voie chirurgicale. L'histologie était compatible avec un diagnostic de lymphome cardiaque primaire non hodgkinien à cellules B non différenciées (lymphome de Burkitt atypique). En autant que nous le sachions, ce rapport est la première description d'un lymphome cardiaque primaire de Burkitt atypique. La présence d'une masse dans l'oreillette droite devrait soulever la possibilité d'une tumeur cardiaque maligne. Il faut envisager une échocardiographie transœsophagienne comme première intervention diagnostique. Il est important de poser un diagnostic histologique rapide parce que la thérapie systémique peut influencer sur le pronostic en présence d'un lymphome cardiaque primaire.

The following case report is the first description of a primary cardiac Burkitt-like lymphoma of the right atrium.

CASE PRESENTATION

A 74-year-old man with no history of cardiac disease presented to a local hospital for shortness of breath. He was referred to the Quebec Heart Institute, Laval Hospital (Quebec City, Quebec) after echocardiography revealed the presence of a large pericardial effusion. Initial transthoracic echocardiography at the hospital confirmed the presence of a pericardial effusion with signs of tamponade. A right atrial mass was suspected on initial imaging. The patient underwent urgent pericardiocentesis.

Transesophageal echocardiography (TEE) revealed the presence of a large mass in the right atrium. The mass had a broad base, was heterogeneous, measured 4 cm × 5 cm and was attached to the right side of the interatrial septum (Figures 1 and 2).

Analysis of the pericardial fluid showed atypical lymphoid cells. Surgical resection of the mass was performed for debulking and diagnostic

purposes. Surgical removal was laborious, necessitating reconstruction of the interatrial septum with a pericardial patch and was complicated by a complete atrioventricular block that required permanent pacemaker implantation.

The resected tumour was submitted to the pathology laboratory as white and red soft fragments measuring 8.0 cm × 8.0 cm × 2.5 cm. Histologically, it consisted of a diffuse proliferation of small to intermediate size lymphocytes with one to four prominent nucleoli and high mitotic activity, accompanied by scattered tingible body macrophages (Figure 3), giving the tumour a starry sky appearance at low magnification (Figure 4). Periodic acid-Schiff stain with and without diastase revealed no cytoplasmic accumulation of glycogen in the tumour cells. Immunophenotypically, the tumour cells were reactive with antisera directed against CD20, CD10, CD43, bcl-2 and CD99 but not terminal deoxynucleotidyl transferase, CD45RO, Epstein-Barr virus latent infection membrane protein 1, CD30, bcl-1, bcl-6, Wilms' tumour 1, thyroid transcription factor 1, CK7,

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Figure 1) Transesophageal echocardiogram at 0° showing a large heterogeneous mass attached to the right side of the interatrial septum

Figure 2) Transesophageal echocardiogram at 130° showing a large mass attached to the right interatrial septum and almost completely filling the right atrium

CK20, AE1/AE3, CK5/6, CAM5.2, CD56 or desmin. The proliferation index assessed by Ki67 was 60%. The pathological diagnosis was made according to the World Health Organization criteria and interpreted as undifferentiated B-cell non-Hodgkin's lymphoma (Burkitt-like).

The patient tested negative for HIV. No other organ involvement was noted. Standard chemotherapy with cyclophosphamide, adriamycin, vincristine and prednisone (the protocol known as CHOP) was initiated and the patient was transferred back to the referring hospital. The patient did not respond to chemotherapy and palliative care was offered; the patient died six weeks following surgery.

DISCUSSION

Primary tumours of the heart are extremely rare. Atrial myxoma is the most common type, representing approximately 50% of tumours. Approximately 25% of primary cardiac tumours are malignant. Almost 95% of malignant tumours are sarcomas. Primary cardiac lymphoma (PCL) represents 1.3% of primary cardiac tumours and is defined as a lymphoma in which the bulk of the tumour is located within the heart and pericardium (1). Recently, the incidence of lymphoma associated with immunosuppression by HIV or in transplant recipients has increased. Fewer than 100 immunocompetent patients with PCL have been reported to date (1). PCL usually occurs in adults with a male-to-female ratio of 2:1. Symptoms associated with PCL are nonspecific, making early diagnosis difficult. In their series, Fuzellier et al (1)

Figure 3) Histology at high magnification showing diffuse proliferation of small to intermediate size lymphocytes, with one to four prominent nucleoli and high mitotic activity, accompanied by scattered tingible body macrophages

Figure 4) Histology at low magnification resembling a starry sky

reported right-sided heart failure, dyspnea, tamponade and rhythm disturbance as the most frequent manifestations.

Right-sided chambers are usually involved in PCL, with two-thirds of cases involving the right atrium. Only one patient has been reported to have PCL isolated in the left heart chambers (1,2). This is in contrast to cardiac myxoma, in which 75% of cases are located in the left atrium and only approximately 15% of cases arise from the right atrium (3). Most sarcomas involve the left atrium but angiosarcoma has a predilection for the right atrium. In contrast to PCL, angiosarcomas are usually well defined, originate on the atrial free wall near the inferior vena cava and do not involve the interatrial septum.

TEE should be considered as an initial imaging method when an intracardiac mass is suspected (4,5). The sensitivity of TEE for the detection of PCL approaches 100% in some series (2). Cytology in the pericardial fluid is often nonspecific, demonstrating atypical lymphoid cells (2). Only approximately 19% of pericardial fluid samples are diagnostic; the majority of cases require biopsy or surgical excision for diagnosis (1). In the presence of a right-sided cardiac mass, an aggressive approach to obtaining a rapid histological diagnosis is important. Less invasive procedures, such as TEE-guided biopsy, endomyocardial transvenous biopsy, mediastinoscopy and thoracoscopic pericardial window, have been performed with success (2).

Over 70% of PCL cases described so far have been diffuse, large B-cell lymphomas. T-cell lymphomas are extremely rare, with only two cases reported to date (1). To the best of our knowledge, the present

report is the first description of a Burkitt-like primary cardiac lymphoma. Burkitt-like lymphomas are high-grade aggressive lymphomas associated with a poor short-term prognosis.

If a PCL diagnosis is documented, a curative surgical procedure should be discouraged. Surgical resection of PCL is often difficult and incomplete, and should be reserved for patients with life-threatening hemodynamic compromise (1). Early systemic treatment appears to be the only chance for cure. Chemotherapy remains the preferred initial treatment; the CHOP protocol is most often employed. Complete remission has been achieved in less than 60% of cases (1).

CONCLUSION

The presence of a mass in the right atria should raise the possibility of a malignant cardiac tumour. TEE should be considered as the initial diagnostic procedure to be performed. Rapid histological diagnosis is important because systemic therapy can influence the prognosis in the presence of a primary cardiac lymphoma.

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