

Primary Cardiac Lymphoma in a 62-Year-Old Man

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Abstract

A 62-year-old man was referred to our hospital with dyspnea. Preliminary studies revealed multiple masses in the right ventricle, epicardium, and pericardium; no lymphadenopathy or organomegaly was, however, detected.

The patient underwent a surgical operation with the diagnosis of a cardiac mass. Multiple, firm, whitish-yellow nodules with extension to the epicardial fat were excised, and the defect was repaired with an extensive pericardial patch. The myocardium was infiltrated by discohesive sheets of malignant round cells that had a high nucleocytoplasmic (N/C) ratio, scanty cytoplasm, and a coarse chromatin pattern.

The diagnosis was further confirmed by a panel of immunohistochemistry markers; the neoplastic cells were positive for CD 45 and CD 20.

Primary lymphomas originating from the heart and pericardium are extremely rare and constitute only 1.6 percent of cardiac neoplasms. They arise mainly from the right chambers and may be of low, intermediate, or high grade. The majority are of B-cell nature. No association with viruses has been established. Cytology is diagnostic in the effusions of the pericardium. Unfortunately, prognosis is grim due to delayed diagnosis (*Iranian Heart Journal 2008; 9 (4): 47-49*).

Key words: cardiac tumors ■ lymphoma ■ B-cell type

PPrimary cardiac tumors are rare, the most common being myxomas. Primary lymphomas originating from the heart and pericardium are extremely rare. They are usually diagnosed at autopsy and are responsible for only 1.6 percent of cardiac neoplasms. They arise chiefly from the right-sided heart chambers and may be of low, intermediate, or high grade. The majority are of B-cell nature. No association with any viruses has so far been noted. We present a case of primary cardiac lymphoma with the involvement of the right ventricle, epicardium, and pericardium in a 62-year-old man.

Case report

A 62-year-old man was referred to our hospital with dyspnea.

Preliminary studies revealed multiple masses in the right ventricle, epicardium, and pericardium, but no lymphadenopathy or organomegaly was detected. The patient underwent surgery with the diagnosis of a cardiac mass.

Multiple firm, whitish-yellow nodules with extension to the epicardial fat were sent to the pathology department.

The tumoral tissue was solid in appearance and firm to fragile in consistency.

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Microscopic findings included infiltration of the myocardium by discohesive sheets of malignant round cells that had a high N/C ratio, scanty cytoplasm, and a coarse chromatin pattern. The foci of necrosis, hemorrhage, and fibrin exudation were also seen. (Fig 1).

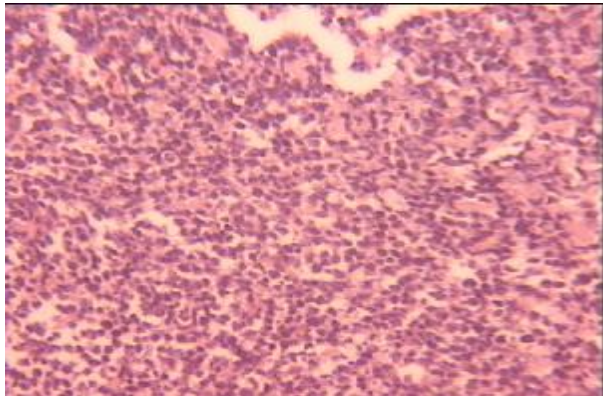


Fig 1. Photomicrograph of the tumor showing malignant round cells, foci of necrosis, hemorrhage, and fibrin exudation.

In view of the above histopathological findings, a diagnosis of malignant round cell tumor suggestive of lymphoproliferative disorders was established.

To further confirm the histological diagnosis, a panel of IHC markers was requested. The immunohistochemistry study included CD 45, CD 20, CD3, CK (for carcinomas), S100, and HMB45. The last two markers were used to exclude malignant melanoma. The tumoral cells were positive for CD 45 and CD 20, but negative for the rest of the markers. According to the WHO classification of lymphoid neoplasms, the final diagnosis was, therefore, Non-Hodgkin's lymphoma, diffuse large B-cell type.

It is also noteworthy that the pericardial fluid of the patient showed a lymphocytic infiltrate. The patient was discharged from the hospital but returned some two months later with worsening of his symptoms.

Discussion

Primary cardiac tumors are known to be rare. Be that as it may, primary lymphomas originating from the heart are even more rare, accounting for only 1.6 percent of cardiac neoplasms. Such lymphomas predominantly arise from the right chamber and are primarily of B-cell nature. No association has thus far been established with viruses.

It is important that primary cardiac lymphomas be included in the differential diagnosis of a right atrial mass.

This group of lymphomas is found mostly in the right ventricle with the involvement of the epicardium and pericardium, but there is usually no trace of lymphadenopathy or organomegaly.

Multiple firm, whitish-yellow nodules are seen grossly with a solid appearance and firm to fragile consistency.

Histopathological findings include infiltration of myocardium by discohesive sheets of malignant round cells that have a high N/C ratio, scanty cytoplasm, and a coarse chromatin pattern. Because these tumors are mainly diffuse large B-cell lymphomas according to the lymphoma classification, one expects IHC markers such as CD 45 and CD 20 to be positive.

Cytology is diagnostic in the effusions of the pericardium in such cases. Unfortunately, the prognosis is grim if diagnosis is delayed.

This entity, however, must be distinguished from two other similar conditions, i.e.

Epstein-Barr-related lymphoproliferative disorders seen in AIDS patient or post-transplant lymphomas. There is no evidence that cardiac lymphomas, which are seen in immunocompetent patients, contain genomic viral DNA.

Another important point in the differential diagnosis of primary cardiac lymphoma from post-transplant lymphoproliferative disorder lies in the fact that the latter is a heterogeneous infiltrate of reactive lymphocytes.

Clinical outcome varies; nonetheless, an early diagnosis in conjunction with effective treatment (surgery and/or chemotherapy) may result in an excellent prognosis.

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