Primary Cardiac Lymphoma

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Abstract

A 4-year-old boy referred to our clinic with fever, abdominal pain and vomiting. He had a muffled heart sound and increased Jugular venous pressure on physical examination and cardiomegaly on the chest X-ray. Echocardiography showed a mass in the right ventricular outflow tract. A biopsy from the mass revealed primary cardiac lymphoma. He did not respond to chemotherapy and died.

Key words: Lymphoma, primary tumors, primary cardiac tumors, cardiac lymphoma

Introduction

Primary tumors of the heart are rare and primary lymphoma involving only the heart and pericardium is much less common, and there are few case reports mostly in adults (1,2). In this report, we present a four-year-old boy with primary cardiac lymphoma, who survived 19 months after chemotherapy.

Case report

A 4-year-old boy presented with fever, abdominal pain and vomiting for several days. His initial chest x-ray showed cardiomegaly. During hospital course he developed tachycardia and tachypnea. His jugular venous pressure was moderately increased and the heart sounds were muffled. An ejection systolic murmur (grade II/IV) heard in pulmonary area. His liver was palpable 2 centimeters below the costal margin. No lymphadenopathy and splenomegally detected. Hemoglobin level was 10g/dL, WBC count was 8350/ml with 48.9% lymphocytes. Platelet count was 324000/ml. BUN, creatinin, uric acid and liver function tests, all were normal. ESR showed increment and CRP was positive. Bone marrow aspiration, bone survey, and abdominal sonography were normal. CT scan of thoracic cavity showed cardiomegaly and pericardial effusion. Electrocardiogram showed typical signs of pericarditis with ST elevation in most leads. His echocardiography showed moderate pericardial effusion. There was a pedunculated mass in pericardial space and a 2x1.5cm mass in the right ventricular outflow tract. Pericardial tap showed a serosanguinous fluid with total cell count of 22,500/ml and WBC count of 2,000/ml with 90% PMN and 10% lymphocytes. Protein was 6.2g/dl and sugar was 105mg/dl. The cytology of pericardial fluid was negative for malignant cells. Pericardiectomy was done and a biopsy was taken from the right ventricular outflow tract which showed primary cardiac lymphoma (Fig.1 and 2).

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Fig 1 and 2: Indicating uniform round cells with deep basophilic cytoplasm and numerous vacuoles, diagnostic for Burkitt's lymphoma. (The biopsy taken from the outflow tract mass of the patient)
Systemic chemotherapy including cyclophosphamide, Adriamycin, vincristine and prednisolone was started. Serial echocardiography showed a decrease in the size of the right ventricular outflow tract mass.

After six courses of systemic chemotherapy, echocardiogram and physical findings were completely normal. Three months after the last course, a low grade ejection systolic murmur was heard in the left sternal border, and a small mass was detected in pulmonary area. Another course of chemotherapy was started. After six courses of chemotherapy, signs of pulmonary insufficiency and pulmonary hypertension were appeared. His parents refused radiotherapy and follow-up, and unfortunately he expired three months later.

**Discussion**

Primary cardiac lymphoma is rare. Most of the cases occur in older age group, and there is no report of such a young patient in the literature. However secondary lymphomatous involvement of the heart is more common, with an estimated incidence of 20%.14

Most primary cardiac lymphomas are proved at the time of autopsy, because it is very difficult to confirm it clinically. Numerous modalities have been employed for diagnosis of primary cardiac lymphoma such as echocardiography, g XM scan, and cytological examination of pericardial fluid. However, only few cases were positive for cytology of pericardial fluid. The definitive diagnosis by pathological report at biopsy or autopsy.6-13

The clinical presentation depends on the location of the tumor. Pain, arrhythmia, signs of heart failure, cardiac tamponade, pulmonary emboli and sudden death, all are described.9

**References**