DOI: 10.7860/JCDR/2023/59601.17236

Case Report

Others Section

Primary Cardiac Burkitt's Lymphoma- A Deadlock

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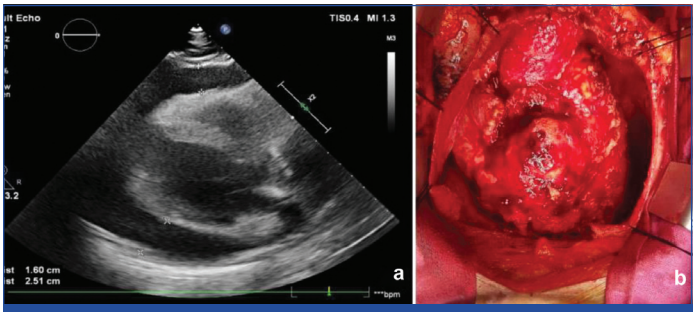
ABSTRACT

Burkitt's lymphoma is a high-grade, extremely aggressive form of B-cell non Hodgkin lymphoma with a very fast replication time (tumour doubling time of around 25 hours). The three forms of Burkitt's lymphoma are documented which include endemic, sporadic, and immunodeficiency associated. It is enormously rare with less than 30 cases reported till date. Primary cardiac lymphoma is a rare cardiac neoplasm. The most common type is diffuse large B-cell lymphoma. Diagnosis is usually late and with poor prognosis. Primary cardiac lymphoma is more common in immunodeficient patients. This is a case of primary cardiac Burkitt's lymphoma which was diagnosed in a 58-year-old immunocompetent male. The patient's two-dimensional echocardiography and plain Computed Tomography (CT) scan of the chest revealed presence of massive pericardial effusion with mild to moderate bilateral pleural effusion. Cardiac Positron Emission Tomography (PET) CT revealed a soft tissue mass at atrioventricular groove which indicated either pericardial lymphoma or inflammatory serositis. The patient was opted for surgery, but the mass was noted involving the free wall of right ventricle including the diaphragmatic surface which extended onto atrioventricular groove and base of right atrial appendage. Thus, a decision to not proceed further with surgical repair was made. The histopathological and cytogenetic findings confirmed the presence of Burkitt's lymphoma. Later, it was discovered that the patient died within one month of diagnosis.

Keywords: Cardiac neoplasm, Intracardiac tumour, Non hodgkin lymphoma, Primary lymphoma

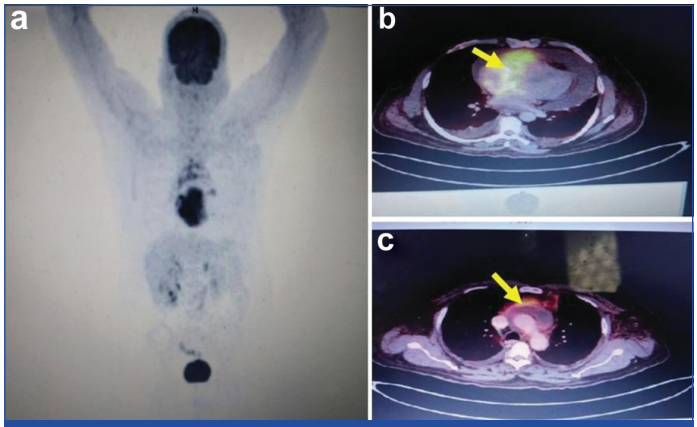
CASE REPORT

A 58-year-old male presented with a complaint of recent onset of dyspnea on exertion since one month (New York Heart Association class II). The patient was hypertensive and occasional alcoholic but non diabetic and non smoker. Two-dimensional echocardiography [Table/Fig-1] and plain Computed Tomography (CT) scan of the chest revealed presence of massive pericardial effusion with mild to moderate bilateral pleural effusion. The patient did not have signs of cardiac tamponade despite massive pericardial effusion.



[Table/Fig-1]: a) Transesophageal echocardiogram: Evidence of massive pericardial effusion causing patient's symptoms of impeding cardiac tamponade; and b) Massive tumour extending onto most of the free wall of right ventricle extending onto the inferior wall and atrioventricular groove.

Cardiac Positron Emission Tomography (PET) CT scan was done which revealed a soft tissue mass at Atrioventricular (AV) groove indicating either pericardial lymphoma or inflammatory serositis [Table/Fig-2]. A decision for pericardiocentesis and excision of mass was taken after a thorough discussion with medical oncologist [Table/Fig-1]. During surgery, pericardial and bilateral pleural fluids were drained for histopathological evaluation. The mass involved most of the free wall of right ventricle including the diaphragmatic surface and extending onto AV groove and base of right atrial appendage. After discussion with the oncologist and patient's family members, it was decided not to proceed further with excision and surgical repair. Frozen section was sent for analysis which was suggestive of lymphoma and revealed tumour with diffuse infiltration



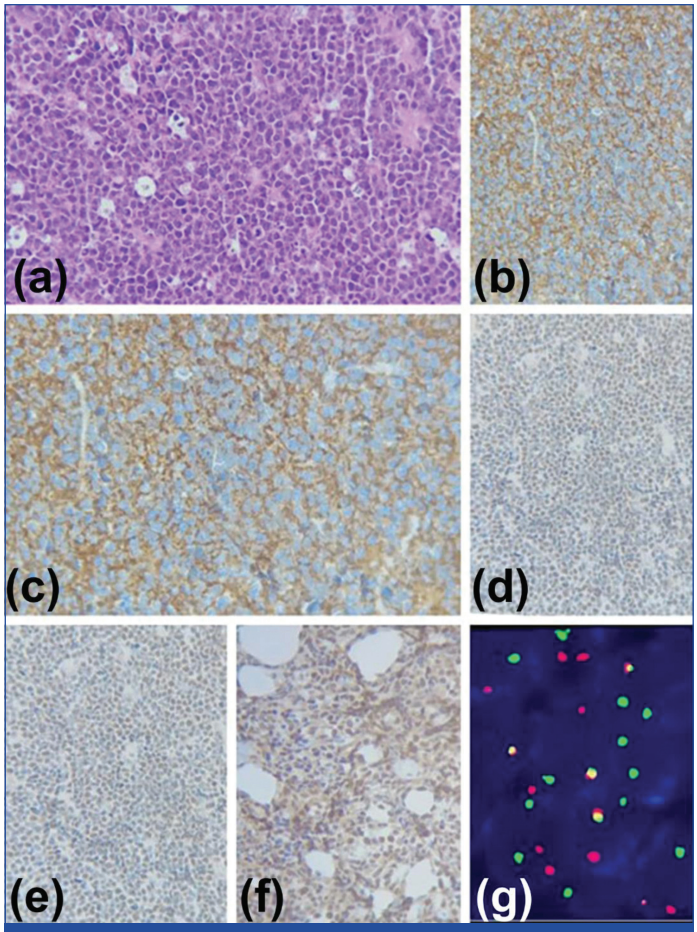
[Table/Fig-2]: Cardiac Positron Emission Tomography (PET) Computed Tomography (CT) images-(a) Maximum intensity projection showing intense FDG (Fluorodeoxyglucose) activity in the region of myocardium. Fused PET CT images localising the increased tracer uptake upto soft tissue mass along the right atrioventricular groove and soft tissue thickening along the superior pericardial sheath; (b) Soft tissue mass along the right atrioventricular groove; and (c) Soft tissue thickening along the superior pericardial sheath.

by medium sized lymphocytes with mild nuclear pleomorphism and scanty cytoplasm, and brisk mitosis with "starry sky" appearance [Table/Fig-3]. The tumour cells were positive for CD20, CD10, c-myc, BCL6 and Mib index (100%), and cytogenetic abnormality of t(8:14) was also detected. Therefore, based on histopathological and cytogenetic findings, Burkitt's lymphoma was confirmed.

Analgesics (tramadol and paracetamol) only if needed, antihypertensive drugs (metoprolol/telmisartan) and antacids (pantoprazole) were prescribed as a palliative care to the patient. The patient was supposed to consult an oncologist for further ement. But the patient did not come for the visit, and during telephonic conversation with his family member about his health, it was discovered that the patient died within a few days of diagnosis.

DISCUSSION

Primary cardiac lymphoma is a rare extra nodal lymphoma with a high degree of malignancy, mainly characterised by cardiac



[Table/Fig-3]: Histopathological evaluation-(a) diffuse infiltration by medium sized lymphocytes with mild nuclear pleomorphism and scant cytoplasm. Brisk mitosis with starry sky appearance noted. Tumour cells are positive for (b) CD20, (c) CD10, (d) cmyc, (e) BCL6, and (f) Mib index of 100%; (g) Cytogenetic abnormality of t(8:14) detected as highlighted by FISH study.

symptoms. The most common type is diffuse large B-cell lymphoma, which mainly involves the right atrium and right ventricle with non specific clinical manifestations [1,2]. The incidence of primary cardiac lymphomas involving only the heart and/or pericardium are <0.05% of which Burkitt's lymphoma is enormously rare with <30 reported cases till date [3]. Burkitt's lymphoma is a high-grade, extremely aggressive form of B-cell non Hodgkin lymphoma with a very fast replication time (tumour doubling time of around 25 hours). World Health Organisation (WHO) has classified Burkitt's lymphoma into three different clinical forms i) endemic (African), ii) sporadic (non endemic) and iii) immunodeficiency associated [3-5]. The endemic type originated in equatorial Africa and New Guinea and is responsible for up to 50% of all childhood cancer in equatorial Africa [5,7]. The sporadic form is generally observed in the United States of America, Western Europe and other parts of the world and is relatively more common in the paediatric population (30% of paediatric lymphomas); however, accountable for <1% of non Hodgkin lymphoma in adults [7]. Immunodeficiency-associated Burkitt's lymphoma is commonly observed in Human Immunodeficiency Virus (HIV)-positive patients, and rarely noted in patients with congenital immunodeficiency and allograft receivers [6,8,9]. The present report was that of the sporadic form of Burkitt's lymphoma in an Indian patient which is rare encounter.

Primary cardiac lymphoma usually involves right atrium and right ventricle, rarely left atrium and left ventricle, and can also involve the superior vena cava, inferior vena cava, and pericardium [2]. Primary cardiac lymphomas present with non specific symptoms as chest tightness, chest pain, dyspnea, arrhythmia, syncope, superior vena cava syndrome, heart failure and other symptoms. Atrial fibrillation, atrioventricular block, right bundle branch block, T wave inversion, and life-threatening ventricular tachycardia may be found on electrocardiogram [10,11]. Jimenez CAH et al., reported a case of Burkitt's lymphoma in a 69-year-old immunocompetent female, who presented with signs of heart failure and died during surgical intervention [3].

Burkitt's lymphoma is characterised by the translocation and deregulation between the long arm of chromosome 8, the site of the MYC oncogene (8q24), and one of three locations on immunoglobulin (Ig) genes which include i) Ig heavy chain gene on chromosome 14 (80%)-t(8;14); ii) kappa light chain gene on chromosome 2 (15%)-t(2;8) or; iii) lambda light chain gene on chromosome 22 (5%)-t(8;22) [6,8]. Furthermore, the lymphoma presents a classic "starry-sky" pattern during histopathological evaluation, and tumour cells commonly express IgM and kappa light chains, B cell-associated antigens (CD19, CD20, CD22, CD79a), and germinal centre-associated markers such as CD10 and BCL6 [5,8]. The histopathological and cytogenic evaluation, in the present case, also showed expression of CD20, CD10, c-MYC, BCL6 and Mib index (100%) in tumour cells, "starry sky" appearance, cytogenetic abnormality of Ig heavy chain gene on chromosome 14, and t(8:14) were also noted which confirmed the diagnosis of primary cardiac Burkitt's lymphoma.

In this patient, the surgical excision of the intracardiac mass was not opted based on oncologist's suggestion in view of the extensive involvement of the tumour and he was recommended chemotherapy. A rapid doubling time (approximately 25 hours), and intracardiac involvement can become life-threatening very quickly, as observed in the patient. Thus, a timely management, preferably with chemotherapy instead of surgery or radiation, should be opted for best outcomes in patient with Burkitt's lymphoma. However, there is no proved superiority of chemotherapy over surgery/radiation in these patients.

CONCLUSION(S)

Primary cardiac Burkitt's lymphoma is a rare and life-threatening disease due to its aggressive nature and quick tumour progression with a very low survival rate. Unfortunately, the patient died within one month of the diagnosis. However, this case is a prompt reminder about the importance of early diagnosis using interdisciplinary consensus, appropriate and timely management, and optimisation of cardiac symptoms in order to prevent unfortunate death in these patients. Currently, the most effective treatment is only chemotherapy.

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PLAGIARISM CHECKING METHODS: [Jain H et al.]
- Plagiarism X-checker: Aug 10, 2022
- Manual Googling: Nov 25, 2022
- iThenticate Software: Dec 02, 2022 (17%)

ETYMOLOGY: Author Origin

AUTHOR DECLARATION:
- Financial or Other Competing Interests: None
- Was informed consent obtained from the subjects involved in the study? Yes
- For any images presented appropriate consent has been obtained from the subjects. Yes

Date of Submission: **Aug 10, 2022**
Date of Peer Review: **Sep 17, 2022**
Date of Acceptance: **Dec 03, 2022**
Date of Publishing: **Jan 01, 2023**