Burkitt’s Lymphoma with an Unusual Cardiac Involvement: A Case Report

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ABSTRACT

Burkitt’s lymphoma (BL), a variety of non-Hodgkin’s lymphoma, is uncommon in India. Cardiac involvement in sporadic BL is rare. Cardiac involvement may be primary or a part of a systemic disease process. It affects the endocardium, myocardium, or pericardium. Cardiac symptoms may or may not be present in the early clinical stages. We are presenting a case of sporadic BL in a 13-year-old child with cardiac and systemic involvement.

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INTRODUCTION

Burkitt’s lymphoma, a monoclonal proliferation of B lymphocytes, is uncommon in adults but common in children with a doubling time of less than 24 hours.1 Males are affected predominantly. BL may be endemic (eBL, African), non-endemic (sBL, sporadic), or immunodeficiency related. eBL typically involves the orofacial region (jaw and facial bone) which may spread to extranodal sites like genitalia, kidney, breast, meninges, and bone marrow. sBL commonly presents with abdominal mass, ascites, genital involvement, bone marrow, and central nervous system involvement. Lymphadenopathy is more related to immunodeficiency cases.2 Cardiac involvement of lymphoma may be primary or as a part of the systemic disease process. Disseminated non-Hodgkin’s lymphoma is more common than primary cardiac lymphoma. Intracardiac mass caused by BL is extremely rare, and young males are commonly affected.3 We are presenting here a case of BL in a young male with cardiac and systemic involvement.

CASE DESCRIPTION

A 13-year-old Muslim boy of Malda (West Bengal) attended the outpatient department of general medicine of our hospital on 9th January 2019 with complaints of insidious onset dull aching intermittent abdominal pain and a right-sided neck swelling for 3 months. There was no associated history of fever, rash, anorexia, weight loss, chest pain, palpitations, shortness of breath, or syncope. There was no history of similar illnesses or any chronic diseases in the past. The boy had a normal developmental and birth history. There was no history of consanguinity in the family.

On examination, his higher functions were normal, pulse 98/min—regular, symmetrical, BP 100/66 mm Hg in supine position with an absence of pallor, cyanosis, icterus, and clubbing. No deformities or skin manifestations could be seen. He was afebrile. Enlargement of right submandibular and supravcircular lymph nodes of >2 cm size was noticed. They were firm, non-tender, and mobile without any overlying skin changes. Abdominal examination revealed hepatosplenomegaly (17.8 cm), splenomegaly (3 cm below the left costal margin), and an intra-abdominal epigastric mass with extension to the left hypochondrium (5 × 7.5 cm, tender, mobile, and nonpulsatile). Bilateral testicular swellings were noted which were non-tender, nonpulsatile, and negative for transilluminescence with a normal cord. No other significant clinical findings were found. The patient was admitted in the general medicine ward for a workup.

Investigations

Baseline investigations revealed normocytic normochromic anemia, neutrophilic leukocytosis, thrombocytosis, high ESR, and decreased hematocrit (Hb:9.8/ WBC:14,500/platelet:5.3 lakh/PCV:32.5/ N77,L08,M10,E05,ESR-85). Renal and liver function test reports were normal. Lipid profile was mildly deranged (total cho:218/ TG:304/HDL:60), CRP (106 mg/L) and LDH (1320 U/L) were raised. Chest X-ray, blood cultures, and urinalysis were normal. The peripheral smear did not reveal any abnormal cells. ECG was normal. Malaria parasites could not be detected; tests for EBV virus and HIV I and II were negative. Abdominal sonography revealed a hypoechoic SOL (7 × 5 cm) occupying the left flank of retroperitoneal origin and multiple enlarged para-aortic lymph nodes. Mild ascites were present. Another (2 × 1.4 cm) hypoechoic SOL was present at level III of the right side of the neck with partial necrosis and extending up to the right parotid gland. A (2.8 × 1.4 cm) hypoechoic SOL was incidentally detected at the right atrial wall. Scrotal sonography showed bilateral testicular enlargements (right: 8 × 5.4 × 4.5 cm, left: 6.2 × 3.6 × 3.5 cm) with multiple, vascular, and hypoechoic lesions (average size: 1.4 cm) with distortion of the testicular architecture. Echocardiography was done showing multiple heterogeneous SOLs arising from the interatrial septum (IAS); one in the left atrium (1.4 × 1.4 cm) and three in the right atrium (2.0 × 2.0 cm) being the largest. Ventricles were normal. No vascular extension or spread was found (Fig. 1).

18FDG PET-CT revealed metabolically active multiple lymphadenopathies in the right supraclavicular, mediastinal, abdominal pelvic, and left inguinal region along with the involvement of solid organs such as the stomach, bilateral testis and peritoneum, and active multiple metastatic signals in bone marrow (Figs 2A and B).

Bone marrow aspiration and biopsy showed hypercellular marrow without any lymphoid infiltration. Incisonal biopsy from the patient’s right neck gland revealed monomorphic, medium-sized round cells, with a high nuclear–cytoplasmic ratio with infiltration of the soft tissue and striated muscle bundles (Figs 3A and B). Immunohistochemistry showed diffuse round cells and strongly immunoreactive for LCA, CD20, CD10, and moderate immunoreactivity for bcl6 with 80–90% Ki67 index; suggesting non–Hodgkin’s lymphoma—B cell type, CALLA positive with immunoprofiling favoring BL.

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FIGURE

1 A 13-year-old Muslim boy of Malda (West Bengal) attended the outpatient department of general medicine of our hospital on 9th January 2019 with complaints of insidious onset dull aching intermittent abdominal pain and a right-sided neck swelling for 3 months. There was no associated history of fever, rash, anorexia, weight loss, chest pain, palpitations, shortness of breath, or syncope. There was no history of similar illnesses or any chronic diseases in the past. The boy had a normal developmental and birth history. There was no history of consanguinity in the family.
**Discussion**

Burkitt’s lymphoma is a type of non-Hodgkin’s lymphoma, a high-grade B cell malignancy with the fastest doubling time. It is common in children. eBL is related to EBV infection and *Plasmodium falciparum*-mediated immune dysregulation which provokes EBV infection. Mean age of presentation of sBL is 11 years and less likely to involve the jaw. Abdominal presentations like mass, ascites, lymphadenopathy, and genital involvement are more common in sBL. Translocation of MYC proto-oncogene on chromosomes 8–14 occurs in BL. eBL commonly starts with jaw swelling whereas B symptoms are more common in sBL and HIV-associated BL. Cardiac involvement of lymphoma may occur. It is reported in 16% of cases of Hodgkin’s disease and 18% of cases of non-Hodgkin’s disease on autopsy findings. Echocardiography is useful and transesophageal echocardiography is more sensitive to assay cardiac masses. MRI and FDG-PET scan are useful alternatives. Cardiac involvement in BL is rare but more common in HIV-infected cases. Fresneau et al. reported BL arising in the right atrium associated with cervical and mediastinal lymphadenopathy in a 17-year-old female. Meshref et al. also reported a 10-year-old boy with multiple intracardiac masses. Histopathologically, tumor cells typically have round and oval nuclei and two to five distinct nucleoli. The cytoplasm is basophilic or amphophilic containing small lipid-filled vacuoles. High rates of proliferation and apoptosis are characteristic. Macrophages containing ingested nuclear debris are surrounded by a clear space creating a “starry” sky pattern. BL cells express surface IgM light chains, B cell markers—CD19, CD20, CD22, and CD79a, and germinal center-associated markers—CD10 and bcl6. Translocation of t(8,14)q(24,32) was seen in about 80% of cases. Tumor cells are negative for CD5, CD23, and TdT.
Clinical features and investigations are correlating our case as sBL but cardiac involvement as interatrial septal mass (one in LA and three in RA) is rare. With chemotherapy, the patient is responding well.

REFERENCES