

Adult Burkitt Lymphoma: An Unusual Presentation

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Abstract: Burkitt lymphoma is a disease of young population; especially children. It rarely occurs and presented in elderly group. World Health Organization (WHO) divided Burkitt lymphoma in endemic, sporadic and immunodeficiency – associated. The non-endemic variant occurs in children with abdominal involvement. We report a sporadic case presented at the age of 61 years old male with irregular thickening of parietal peritoneum and omentum without involving any lymph nodes.

Case Report

A 61 year old male presented to department of medicine with complaints of fever and weight loss for 2 months, anorexia and melena for 1 month; and abdominal distention and burning micturation. There was no history of nausea, vomiting or change in bowel habits.

Physical examination: Patient was pale looking, lethargic and well oriented. There was mild abdominal distention. All other physical findings e.g. jaundice, posture, nail clubbing, palmar erythema, pigmentation contractures, hairs, lymph nodes, and were found unremarkable. Systemic examination including cardiovascular, pulmonary, alimentary, nervous, genitourinary, endocrine, integumental and musculoskeletal systems was normal.

Laboratorial findings: Hemoglobin was 6.5 g/dL, red blood cell count was $2.33 \times 10^6/\mu\text{L}$, white blood cell count was $15.9 \times 10^3/\mu\text{L}$, hematocrit was 20.7%, mean corpuscular volume (MCV) was 88.8 fL, mean corpuscular hemoglobin (MCH) was 27.9 pg, mean corpuscular hemoglobin concentration (MCHC) was 31.4 g/dL, red cell distribution width (RDW) was 17.7% and platelet distribution width (PDW) was 13.2%. On peripheral blood smear, there was hypochromic anemia with moderate anisocytosis and poikilocytosis; and predominant neutrophils with left shift and few atypical mononuclear were also seen. Platelets count was $13 \times 1000/\text{UL}$ (N = 150 – 400 X 1000/ UL). Reticulocyte count was 2.2%. (N = 0.5 – 2.5% in adults).

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Erythrocyte sedimentation rate (ESR) was 112mm (N = 0 – 22mm). Serum albumin level was 3.1 g/dL (N = 3.5 – 5.5 g/dL), urea was 44 mg/dL (N = 5 – 20 mg/dL), phosphorus was 6.5 mg/dL (N = 2.5 – 4.5 mg/dL), sodium was 144 mEq/L (N = 135 – 145 mEq/L), lactate dehydrogenase (LDH) was 2535 U/L (N = 140 – 280 U/L), uric acid was 18.5 mg/dL (N = 3.4 – 7.0 mg/dL in male), total protein was 5.2 g/dL (N = 6.4 – 8.3 g/dL), amylase was 22 U/L (N = 23 – 85 U/L), lipase was 30 U/L (N = 0 – 160 U/L), parathyroid hormone level was 6.9 pg/mL (N = 10 – 55 pg/mL), vitamin D level was 121 ng/mL (N = 20 – 50 ng/mL), ascitic fluid protein was 4 g/dL (N = <3 g/dL) (transudate), ascitic fluid adenosine deaminase (ADA) was 53 IU/L. On ascitic fluid examination 95% atypical mononuclear cells were seen. Serology for dengue, hepatitis and HIV were negative.

Radiological findings: On ultrasound abdomen and pelvis, there was mild pelvic ascites and renal parenchymal changes grade I were noted. There was no visceromegaly. All other findings were unremarkable. On CT scan abdomen and pelvis with contrast, there was gross ascites on abdominopelvic cavity. There was thickening of omentum on the right anterolateral side of abdomen causing displacement of the bowel loops from normal anatomical location representing omental caking. Irregular thickening of the parietal peritoneum was also noted. No lymphadenopathy was found on imaging. There were degenerative changes presented in bones. All other structures including liver, spleen, kidneys and urinary bladder were normal. Concluded picture of CT scan was irregular thickening of omentum and parietal peritoneum likely representing metastatic disease process.

Bone marrow biopsy findings: Bone marrow biopsy from posterior superior iliac spine was done. Bone

marrow aspiration was diluted as there was not significant cellularity, but there were blasts cells present in aspiration; with moderate - sized cells having basophilic cytoplasm and vacuolation. The nuclei were regular and round - oval in shape. Nucleoli were prominent (Figure 1).

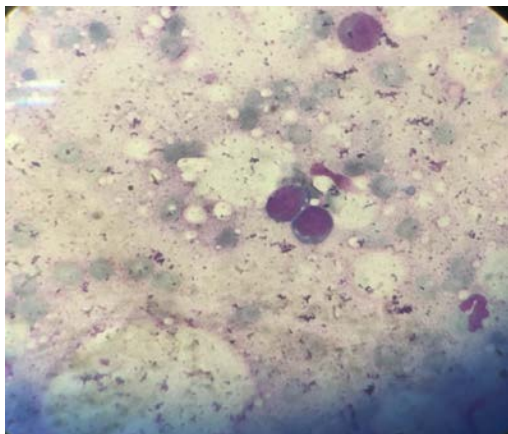


Figure 1: Bone Marrow Aspirate (40x) showing blasts with prominent nucleoli and basophilic cytoplasm with vacuolation

Imprint of trephine biopsy showed monogenous population of blasts cells with basophilic cytoplasm and vacuolation (Figure 2).

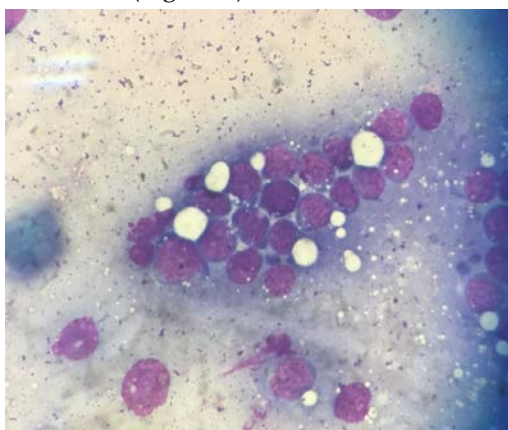


Figure 2: Bone marrow imprint of trephine biopsy (100x) showing infiltrate by atypical mononuclear cells having cytoplasmic vacuolation

Morphologically the findings were in the favor of Burkitt leukemia (ALL - L3). Bone marrow trephine biopsy of patient showed hypercellular marrow with decrease in all cell lines (myeloid, erythroid, and megakaryocytes) and infiltration by homogenous population by atypical mononuclear cells (Figure 3).

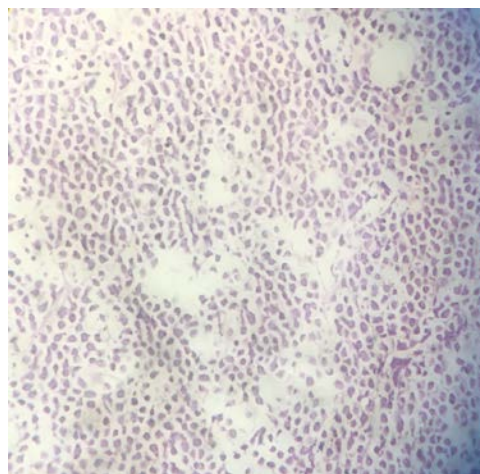


Figure 1: Bone Marrow Trephine Biopsy (40x) showing infiltration by homogenous population of atypical mononuclear cells

Immunophenotyping was performed on cut section of bone marrow trephine biopsy. Specific panel of markers was performed consisting of Pan - CK, LCA, CD3, CD20 and Ki - 67. Pan - CK (Figure 4) was negative so tumors of epithelial origin were excluded.

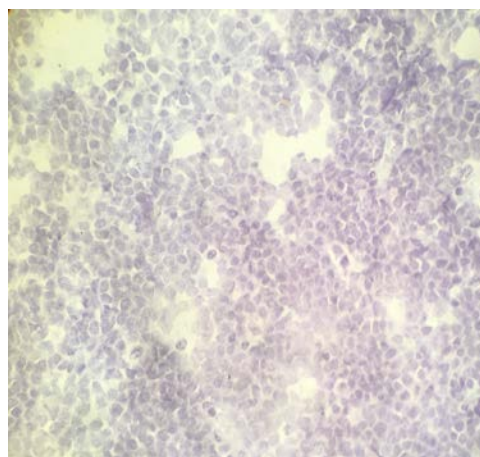


Figure 2: Immunophenotyping (40x) showing negativity for Pan CK marker

LCA (Figure 5) showed positive stain and was in favor of tumor of common lymphoid origin. CD3 (Figure 6) was negative so tumor of T cell origin was less likely to present. CD20 (Figure 7) was positive suggestive its origin from B cell. Ki - 67 showed 100% positivity (Figure 8); strongly suggested of the presence of Burkitt lymphoma.

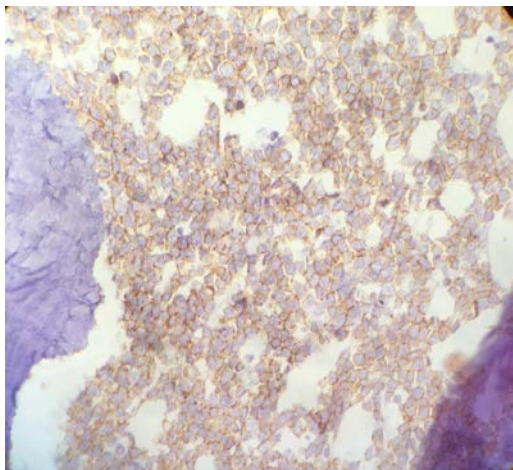


Figure 3: Immunophenotyping (40x) showing positivity for LCA marker

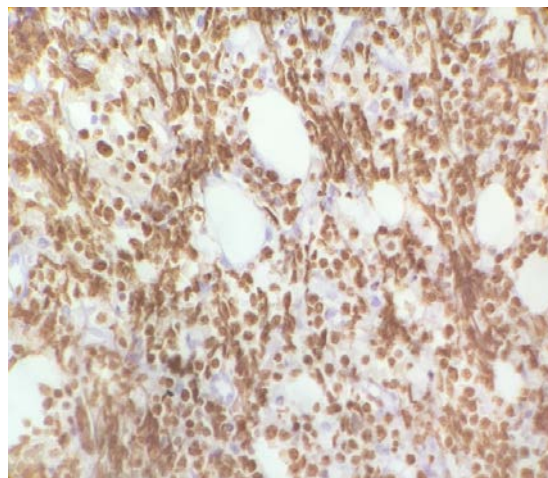


Figure 8: Immunophenotyping (40x) showing 100% positivity for Ki - 67 marker

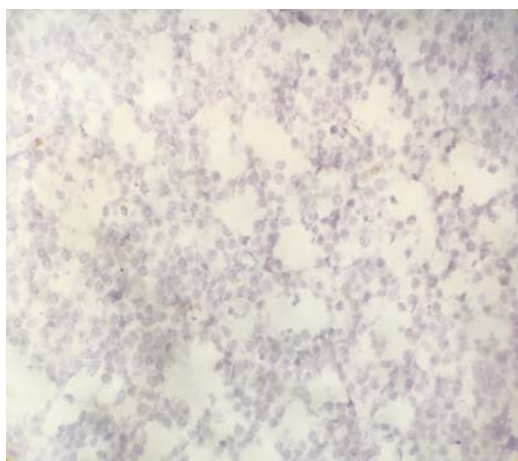


Figure 4: Immunophenotyping (40x) showing negativity for CD3 marker

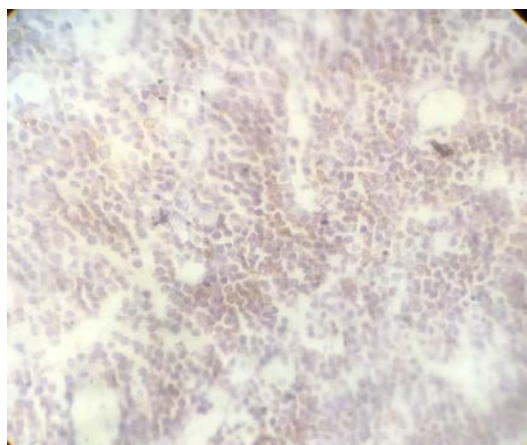


Figure 7: Immunophenotyping (40x) showing positivity for CD20 marker

Discussion

Burkitt lymphoma is very aggressive B cell non - Hodgkin lymphoma, consisting of 1 - 2 % of adult lymphomas. It is divided into three epidemiological categories: endemic, sporadic and immunodeficiency related Burkitt lymphoma (1-3). It has doubling time of 24 hours. Adult patients with sporadic Burkitt lymphoma are often presented with extranodal disease with abdomen being most common site, followed by retroperitoneum, kidney, ovary and testis (4-6). The disease is associated with Epstein - Barr virus (7). The incidence is very high in immunosuppressant patients in non - endemic areas, especially when associated with human immunodeficiency virus (HIV) (8,9). Endemic Burkitt lymphoma occurs in Africa, while sporadic cases are reported from all over the world. Children are most affected age group (5). World Health Organization (WHO) classification of lymphoid diseases recognizes lymphoma and leukemia phase of Burkitt lymphoma as single entity. Morphological features of Burkitt lymphoma are medium sized - cells with abundant basophilic cytoplasm; often containing lipid vacuoles; round nuclei with clumped chromatin and multiple nucleoli; and a diffuse monotonous pattern of infiltration. Immunophenotyping includes expression of surface IgM, Bcl - 6, CD19, CD20, CD22, CD10 and CD79a and negative for CD5, CD23 and TdT (10-13). Patient did not survive as disease is very progressive and having bad prognosis.

We present very unique and extremely rare picture of Burkitt lymphoma in old age group without any lymphadenopathy and HIV - association. There was

only diffuse peritoneal and omental involvement in this patient. Only one case was reported on radiological findings, but there was no bone marrow involvement (14). In our patient, diagnosis was made on bone marrow biopsy and immune-phenotyping with some evidence of carcinoma on radiological findings.

Conclusion

With some features related to abdomen and weight loss only, patient of Burkitt lymphoma may present with very unusual and rare manifestations. So Burkitt lymphoma should also be considered while examining the patients with same clinical features, as rare entities may be present and can confuse with other related conditions. So detailed medical history, physical examination, along with laboratory and radiological workup should be done to reach the final diagnosis.

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