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Adult burkitt lymphoma: Rare presentation

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Abstract

Burkitt lymphoma is a highly aggressive mature B-cell lymphoma, most frequently occurring in children. There are three clinical variants recognized: Endemic (African), sporadic (non-African) and immunodeficiency associated lymphomas (HIV/AIDS). Here, we describe a very unusual presentation of a sporadic Burkitt lymphoma case in a 62-year-old male with diffuse peritoneal and omental involvement, without lymphadenopathy.

Keywords

lymphadenopathy; B-cell neoplasm ; burkitt lymphoma

Background

Burkitt lymphoma is a fast-growing high grade B-cell neoplasm that rarely affects adults [1]. It consists of endemic, sporadic, and immunodeficiency-associated variants. All these subtypes possess chromosomal rearrangements of the c-myc oncogene, the genetic hallmark of Burkitt lymphoma most commonly resulting from t(8;14) [2]. Histologically, Burkitt's lymphoma presents with monomorphic, medium-sized B-cells with many mitotic figures and basophilic cytoplasm. High rates of apoptosis with benign histiocytic phagocytosis of apoptotic debris may lead to a starry-sky growth pattern. The neoplasms are negative for the following: CD5, CD23, BCL-2, and TdT. Ki-67 expression in these tumors, approaches to 100% [3,4]. Long-term survival with various combination chemotherapy including third generation protocol currently is approximately 40–80% [5].

Case Presentation

A 62 year old male presented to the medicine department with complaints of high grade fever, nausea, body aches along with arthralgia's for the past 15 days. There was positive history for weight loss and abdominal distention for past 2 months. There wasn't any associated complaints of vomiting or altered bowel habits.

EXAMINATION: On physical examination patient was pale, lethargic and well oriented. All other physical findings e.g. jaundice, posture, nail clubbing, palmar erythema, lymph nodes were found unremarkable.

Systemic examination including cardiovascular, pulmonary, gastrointestinal and nervous systems were normal.

Laboratory Findings:

Table 1: Shows laboratory findings of complete blood picture.

Sr. No.	Blood CP Parameters	Value	Normal Range
1	Hemoglobin	6.8 g/dl	14-18 g/dl
2	Red Cell Count	2.12	4.5-5.5 million/ul
3	White Cell Count	97.6	4-10 x 109/L
4	Hematocrit	19.2	30-40%
5	Platelet Count	15	150-450X1000/UL
6	Reticulocyte Count	0.5	0.1-4.0 %
7	Erythrocyte sedimentation rate (ESR)	40	0-20 mm

Peripheral film findings: it showed a hypochromic macrocytic picture with markedly decreased platelet count. There were only 4% neutrophils and 2% lymphocytes present and remaining 94% were blasts (atypical mononuclear cells with open chromatin and prominent nucleoli) as shown in Figure1.

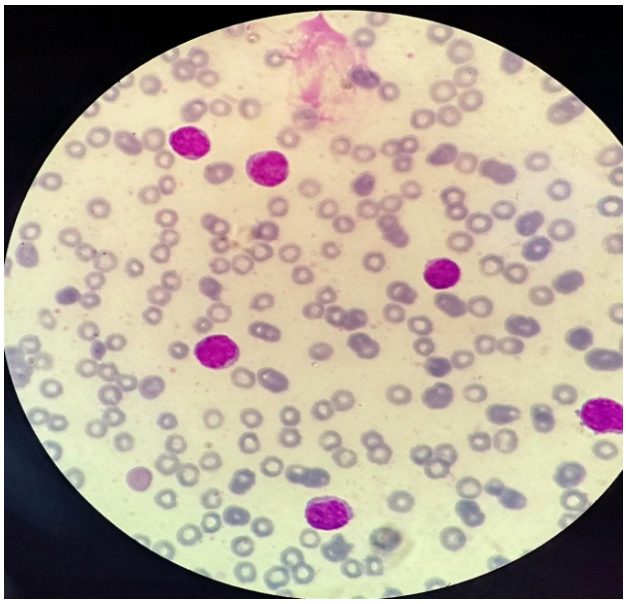


Figure 1: Shows atypical mononuclear cells in peripheral film.

Bone marrow biopsy was done which showed hyper-cellular smear with depressed erythroid, myeloid and megakaryocytic series and there were 96% blasts cells (mononuclear cells with open chromatin, prominent nucleoli and vacuolated basophilic cytoplasm) on the marrow aspirate (Figure2). Similar findings were observed in trephine biopsy was done (figure3 and 4). These cells were having characteristic appearance for Burkitt leukemia therefore, immunohistochemistry was performed on trephine biopsy.

Table 2: Shows routine biochemical laboratory tests along with coagulation profile.

Sr. No.	Parameters	Value	Normal Range
Liver function Tests			
1	Alkaline Phosphatase	126 IU/L	115-460 IU/L
2	Alkaline Transferase	163 IU/L	0-30 IU/L
3	Bilirubin	1.58 mg/dl	<0.1 mg/dl
RFTs			
4	Urea	55mg/dl	40-180 mg/dl
5	Creatinine	21 mg/dl	25-125 mg/Dl
Hepatitis screening			
6	HbsAg	NEGATIVE	
7	Anti HCV	NEGATIVE	
Electrolytes			
8	Calcium	1.97mmol/L	2.2-2.7mmol/L
9	Sodium	132mEq/L	135-147mmol/L
10	Potassium	2.5mmol/L	3.5-5.5 mmol/L
Others			
11	Blood Sugar Random	139mg/dl	100-180mg/dl
12	LDH	2189 U/L	140 – 280 U/L
13	Dengue serology	Negative	
14	ICT MP	Negative	
Coagulation Profile			
15	Prothrombin Time	12 sec	12 sec
16	Activated Partial Thrombin Time	33 sec	32 sec
17	International Normalized Ratio (INR)	0.94	1.0
18	Bleeding Time	3min	2-5 min

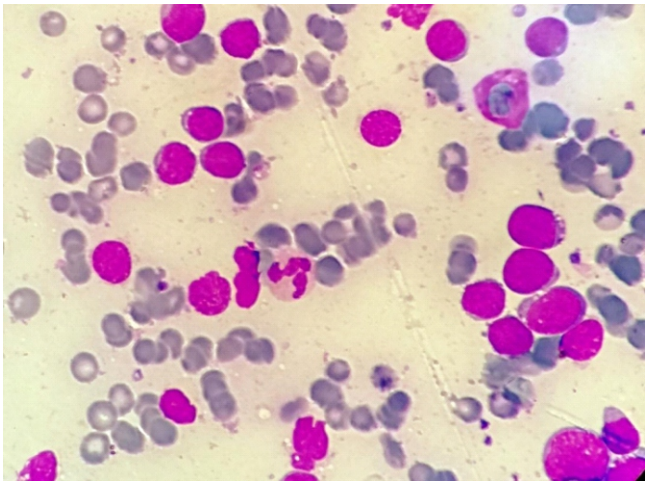


Figure 2: Shows atypical mononuclear cells with basophilic vacuolated cytoplasm.

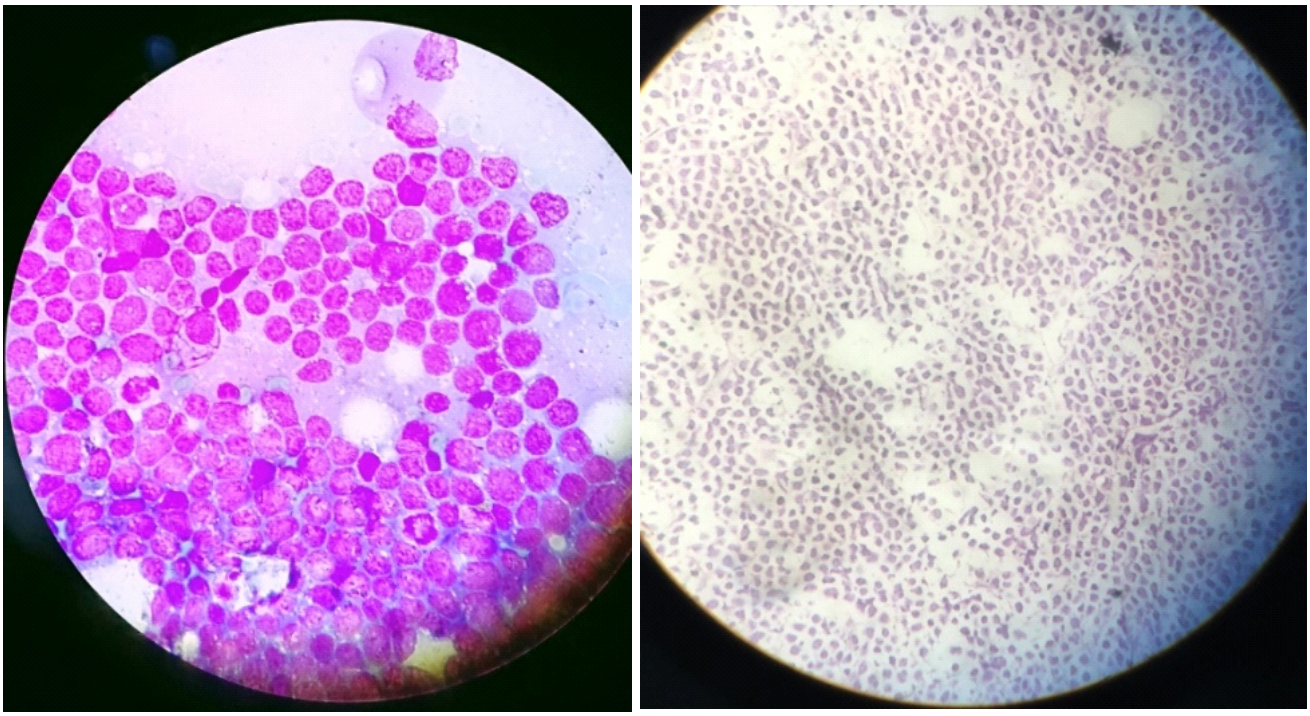
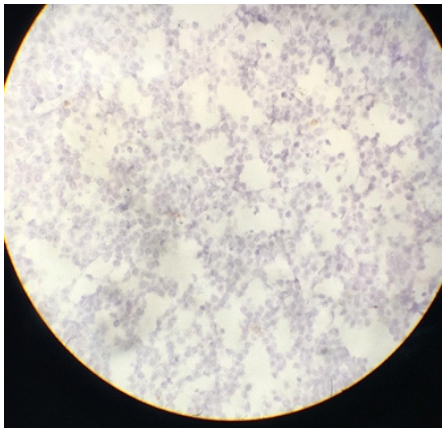
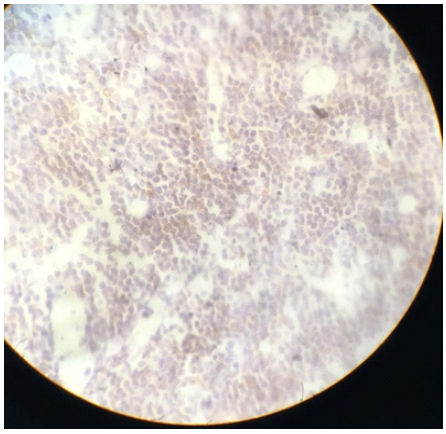
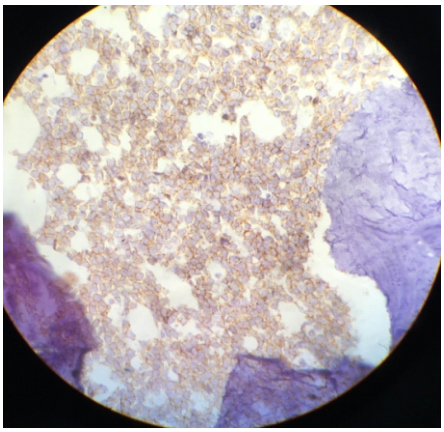
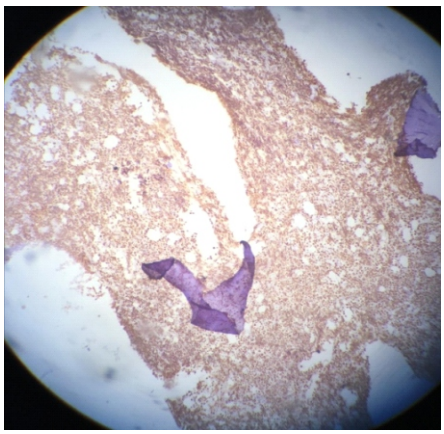
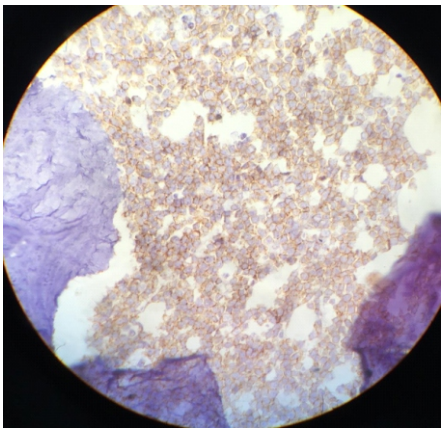
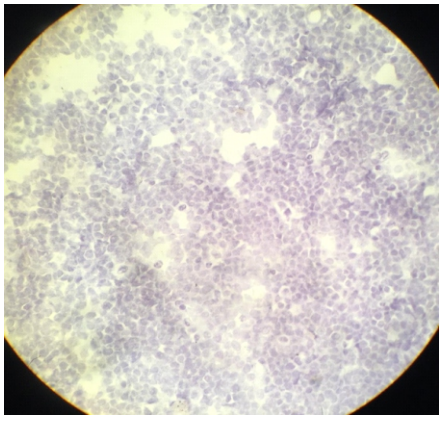


Figure 3,4: Figure 3 (left 100X) and Figure4 (right 40X) Shows diffuse infiltration by atypical mononuclear cells on trephine biopsy.

Immunohistochemistry markers were applied. Cells in the biopsy were negative for Tdt,CD3,and Pan CK, positive for LCA, CD20, and Ki67 was 100% positive which gave confirmation for Burkitt leukemia.

Table 3: Shows the immune histochemical stains applied on trephine biopsy at 40X magnification power.

	
Cells showing negativity for CD3 marker.	Cells showing positivity for CD20 marker.
	
Cells showing diffuse positivity (100%) Ki-67 marker.	Cells showing diffuse positivity (100%)Ki-67 marker. (10X)
	
Cells showing positivity for LCA marker.	Cells showing negativity for Pan CK marker.

Discussion

Burkitt lymphoma is a highly aggressive mature B-cell lymphoma that constitutes less than 2% of all NHL diagnoses. It is divided into three epidemiological categories: endemic, sporadic and immunodeficiency related Burkitt lymphoma [1, 6]. It has doubling time of 24 hours. Adult patients with sporadic Burkitt lymphoma are often presented with extra nodal disease with abdomen being most common site [2,6]. It is associated with immunocompromised patients usually HIV, in the non-endemic areas and also in association with Epstein Barr Virus infection. Involvement of more than 25% cells in the marrow transforms lymphoma into leukemia. Clinical features involve massive painless lymphadenopathy, jaw and facial bone involvement of lymphoma (endemic), abdominal tumors with bone marrow involvement (sporadic) [2,4,6]. Chemotherapy is the mainstay of treatment which include high-doses of methotrexate, cytosine arabinoside and cyclophosphamide. Approximately 50–80% of adult patients with Burkitt lymphoma can be cured with these intensive chemotherapy regimens [4,5].

We report a very unique case of Burkitt lymphoma in an old age group infiltrating blood and bone marrow and moreover, there was no associated lymphadenopathy or HIV-association. There was only abdominal (peritoneal) involvement and diagnosis was made morphologically on bone marrow biopsy and was confirmed through immunohistochemistry. Previously only two such cases have been reported in which there was no lymph node or HIV involvement.

Conclusion

Patients presenting with mild complaints but with a short history should be investigated thoroughly as these rare entities like leukemia's and lymphomas can be missed. A complete medical, radiological and hematological workup should always be done in patients presenting with acute symptoms regardless of there age.

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