
Burkitt's lymphoma-An unusual presentation in an Indian girl

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ABSTRACT

Burkitt's Lymphoma (BL) is a rare monoclonal proliferation of B- lymphocytes and is classified as a poorly differentiated lymphocytic lymphoma. It is highly aggressive. Symptoms are caused by rapid turnover of the mature B lymphocytes and the involvement of extranodal sites and invasion of adjacent organs. This tumor was first noted in Africans. The cause of this tumor is debatable, but strong evidence implicate Epstein- Barr virus(EBV) in its development. This tumor predominantly affects children and is probably the fastest growing tumor in humans, with exuberant proliferation. It is a very rare malignancy accounting to only 0.76% of solid malignant tumors among Indian children. A case of BL with unusual presentation as Krukenberg tumor presenting as bilateral ovarian masses in a 18 year old adolescent Indian girl is reported.

Key words: Burkitt's Lymphoma, Epstein – Barr virus, sporadic type, Adolescent, Krukenberg tumor

Introduction

Burkitt's lymphoma is a rare highly aggressive lymphoma identified and described by Denis Parsons Burkitt, a surgeon in 1958 in Africa, in areas endemic for malaria. BL occurring in non-endemic areas are histologically identical to those occurring in endemic areas[1]. There are three subtypes of BL. The Endemic variant (African Type) is strongly linked with Epstein – Barr virus. Patient presents with involvement of jaw, facial bones(orbit), cervical lymph nodes, distal ileum, caecum, ovaries, kidney, breast, skeletal.[2]The Sporadic type(Non African) is rarely associated with the EBV. Patient presents with abdominal tumors with bone marrow involvement, bowel obstruction secondary to ileo-caecal intussusception caused by tumor growth. Generalized lymphadenopathy is rare. Immunodeficiency – related BL associated with HIV infection, presents with nodal and bone marrow involvement [3].

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Case report

We report a case of adolescent 18 years Indian girl presenting with abdominal pain, nausea, weight loss, fatigue, night sweats, low grade fever, all suggesting B symptoms. Ultrasound and CT Scan of abdomen showed bilateral ovarian masses with spared fat planes (favouring Krukenberg tumor), right ovary measuring 96x70 mm, left ovary measuring 75x54 mm, multiple liver masses, gastric mass, peritoneal deposits, abdominal lymphadenopathy (portal, peripancreatic, pre-paraaortic, aortocaval, retrocaval, mesenteric) largest measuring 1.7x1.2 mm, ascites, Gall bladder wall thickening, multiple mixed osteolytic/sclerotic lesion in whole skeleton system, left pleural/effusion, right jaw swelling and right sided two cervical lymph nodes measuring 1.5 x1.5 cm and 1x1 cm. Overall radiological findings mimicking Krukenberg tumor of ovaries. Fine Needle Aspiration was done from right ovary, liver mass, right cervical lymph nodes (Fig.1). The smears were stained with MGG stain and studied for cytomorphology which showed monotonous population of cells having round intermediate sized nucleus, several or multiple small basophilic nucleoli, basophilic cytoplasm. Vacuolations were seen in cytoplasm and nucleus because of presence of lipid, starry sky appearance due to tingible body

macrophages. Thus a cytomorphological diagnosis of Burkitt's Lymphoma was made. Hematological tests revealed raised total count, raised ESR, Hemoglobin was within normal range. Liver function tests were unremarkable except LDH was raised to 1034 IU/L. Renal function tests were also unremarkable. Ascitic

fluid was examined but did not reveal presence of tumor cells. Peripheral blood film showed 5-6 % of lymphoma cells (Fig. 3). Bone Marrow aspirate also showed presence of morphologically similar lymphoma cells (Fig 2).

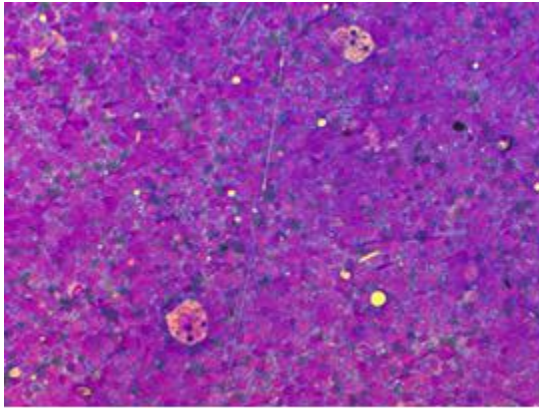


Fig1: Cytology smear (MGG stain) 40X

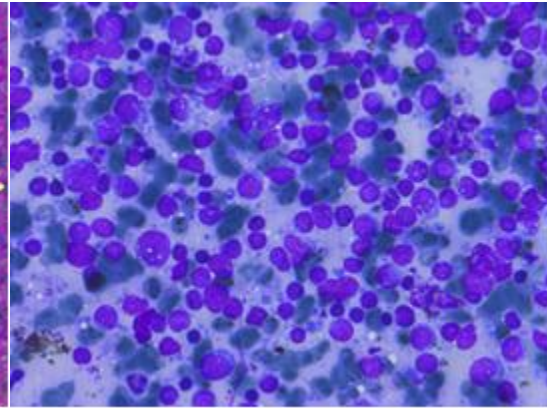


Fig 2: Bone Marrow aspirate smear (MGG stain) 40X

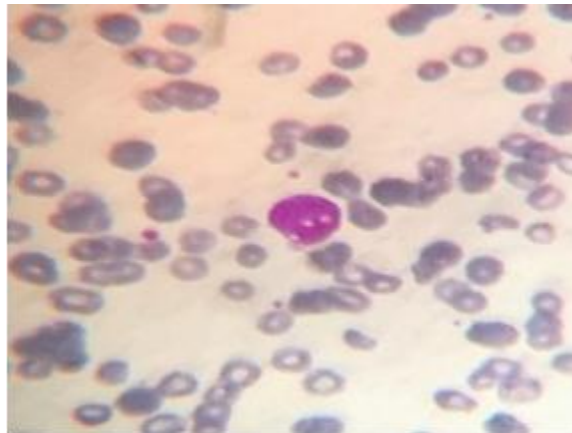


Fig 3: Peripheral Smear (Leishman stain) 100 X

S. EBV by ELISA method was done which showed positivity for IgG, IgM was negative. Anti-HIV was negative by ELISA method. Biopsy was done from cervical lymph node. Histologically on H&E staining the classical starry sky appearance along with monotonous population of intermediate sized round cells with vacuolations in cytoplasm & nucleus were seen (Fig. 4). Histomorphological diagnosis of Burkitt's

Lymphoma was made. Immunohistochemical markers for CD19, CD20 (Fig. 5), CD22, CD10, Bcl6 were positive. Ki67 (Fig. 6) was also positive in most of the cells. Henceforth the final diagnosis of Burkitt's Lymphoma was considered based on the clinical, hematologic, radiographic, cytomorphology, histopathology, serology and immunohistochemical investigations.

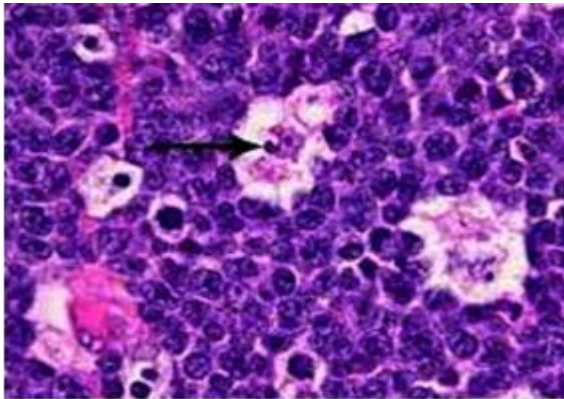


Fig 4: H&E histopathology of cervical lymph

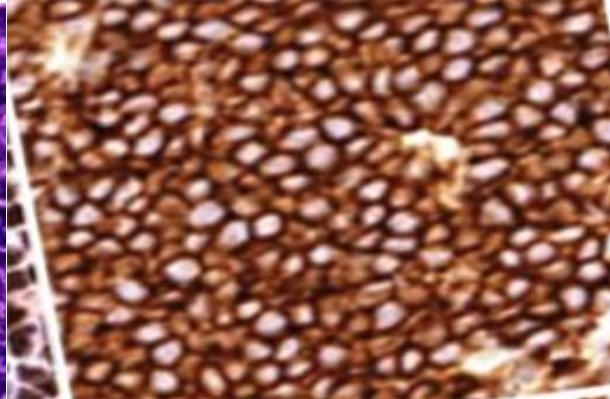


Fig 5: CD 20 , 40X node, 40X

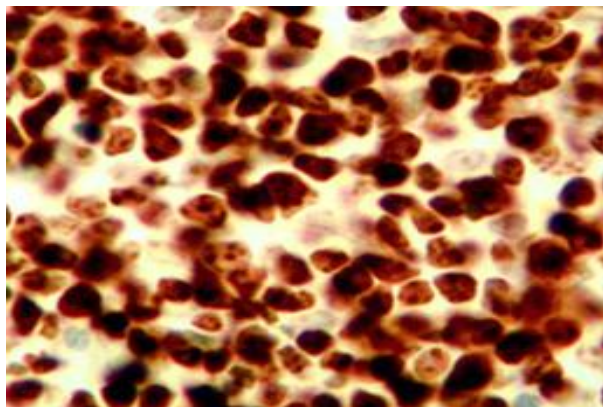


Fig 6: Ki-67 marker, 40X

Discussion

Burkitt's Lymphoma is rare in young adolescent 18 years Indian girl with clinical and radiological presentation as Krukenberg tumor. This is a high grade aggressive subgroup of B-cell Non Hodgkin's Lymphoma (NHL) composed of intermediate sized noncleaved, diffuse, undifferentiated malignant cells of lymphoid origin.[4]. It is the fastest growing human tumor with a doubling time of less than 24 hours [4][5]. In Africa, this tumor accounts for 50% of all childhood cancers. Outside Africa, it accounts for less than 2% of all cases of NHL. In an Indian series of solid malignant tumors in children, Pramanik et al, in 1997 studied 263 cases over a 10 year period and found only two cases (0.76%) of BL [6]. The role of EBV in BL is not well understood. This virus preferentially infects B cells via the C3d complement receptor CD21 [4].

Other co-factors may include chromosomal abnormalities, immune defects and protein energy deficits [4]. Burkitt's Lymphoma in India is intermediate between the sporadic and endemic types

in its clinical presentation. The current case too could be classified as intermediate exhibiting features of both endemic and sporadic forms.

A separate staging system for BL has been developed by Zeigler (1981), whereas Levine et al., (1982) classified the cases of the American BL as follows:[7] Stage I: single tumor mass (extra-abdominal 1A or abdominal 2A).

Stage II : two separate tumor masses on the same side of the diaphragm.

Stage III: involvement of more than two separate masses or disease on both sides of the diaphragm.

Stage IV: pleural effusion, ascites or involvement of the central nervous system or bone marrow.

On the basis of the staging proposed by Levine et al., the present case can be grouped as Stage IV due to the bone marrow involvement.

Conclusion

To conclude, Burkitt's Lymphoma intermediate between endemic and sporadic variety, Stage IV, associated with EBV, in a 18 year adolescent Indian girl with clinical and radiological presentation of Krukenberg tumor is an unusual presentation making the diagnosis challenging. BL should be considered in differential diagnosis of bilateral ovarian tumors. Characteristic morphological features supplemented by Immunohistochemistry helps in arriving at a diagnosis.

Acknowledgements

Patient and her relatives.

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Source of Support: Nil

Conflict of Interest: None