Original Research Article

Aggressive orbital Burkitt's lymphoma with rapid onset proptosis

Jayashri Thorat[1*], Ranjana Bindu[2], Sujata Chahande[3], Smita Harne[4]

Department of Ophthalmology, Dr DY Patil Medical College Kadamwadi, Kolhapur, Maharashtra, INDIA.
Email: dr.kiranpatil84@gmail.com

Abstract

Burkitt's lymphoma is rare B cell monoclonal malignant lymphoma with short doubling time (less than 3 days) occurring due to deregulation of c-myc oncogene. Most Burkitt's lymphomas (80% to 90%) carry a translocation of the proto-oncogene c-myc from chromosome 8 to the immunoglobulin heavy chain region on chromosome 14 [t(8;14)]. In 10% to 20% of cases the c-myc gene is translocated to the kappa light chain loci on chromosome 2 [t(8;2)] or to the lambda light chain on chromosome 22 [t(8;22)]. Translocation leads to over-expression of the c-myc oncogene. Increased incidence is found in AIDS patients. In the sporadic form of the disease that occurs worldwide in non-endemic areas, mostly in developed countries, patients present with an abdominal mass that frequently involves the ileocecum in the bowel, whereas ocular or orbital involvement is rare.

Key Words: Burkitt's lymphoma.

[*]**Address for Correspondence:**
Dr. Jayashri Thorat, Assistant Professor, Department of Ophthalmology, Dr DY Patil Medical College Kadamwadi, Kolhapur, Maharashtra, INDIA.
Email: dr.kiranpatil84@gmail.com

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CASE REPORT

12 year old female patient came in outpatient department with chief complaints of painful rapidly progressive Protrusion of left eye and diminution of vision in left eye since 15 days. Patient was known case of HIV on ART since 1 year. There was History of weight loss associated with abdominal pain. On examination right eye was normal with best corrected visual acuity of 6/6 while left eye had proptosis of 6mm on hertel exophthalmometry. Eye ball was displaced downwards and outwards with fullness in upper lid. Left eye movements were restricted superomedially. Snellen's Best corrected visual acuity in left eye was 6/60. On fundus examination chorioretinal folds were present. On general examination pallor and icterus was noted. systemic examination revealed hepatomegaly. Appearance at presentation chorioretinal folds at posterior pole.

Figure 1:

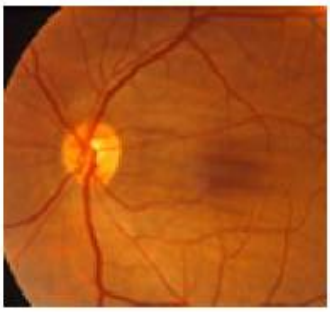

Figure 2:

Figure 3:

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Proptosis increased in size rapidly in span of 4 days. Patient also developed exposure keratopathy due to severe proptosis in spite intensive management with lubricating drops and ointment and lid taping. blood investigations of patient showed raised serum bilirubin-6.2mg /dl.(direct -5.8) and cd4 count-393.CSF was clear indicating no Central Nervous System involvement. MRI BRAIN AND ORBIT -Well defined soft tissue intensity lesions involving extraconal compartment of left orbit superomedially with intracranial extension suggestive of neoplastic etiology mostly lymphoma.

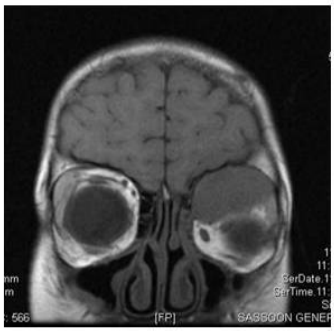

Figure 4:

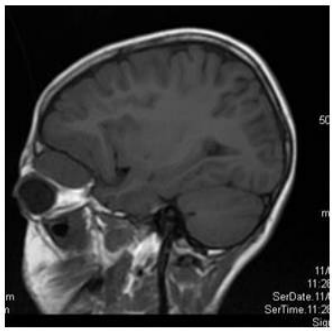

Figure 5:

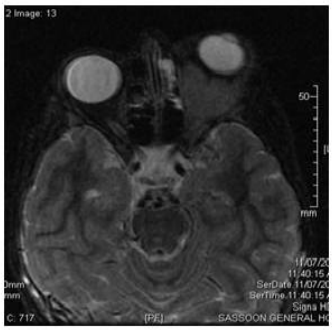

Figure 6:

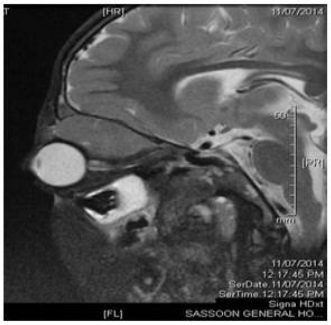

Figure 7:

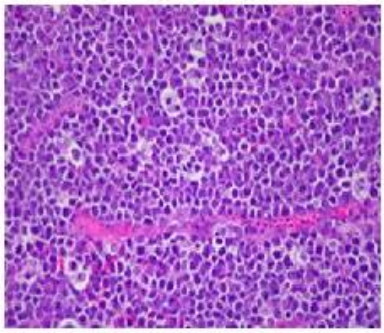

Figure 8:

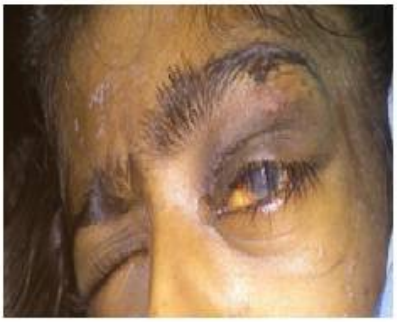

Figure 9:

CT ABDOMEN: Peripancreatic lesion compressing Central bile duct with intrahepatic biliary radicles dilatation suggestive of lymphomatous deposits in right adrenal,mesenteric nodes,peripancreatic and internal iliac nodes. Abdominal lymph node biopsy suggestive of Burkitt's lymphoma. A "starry sky" pattern was apparent in the section of the lymph node mass Of abdomen. The tumor cells are uniform in size and shape with small centrally situated nucleoli (hematoxylin–eosin, original magnification 3400). After consultation with oncologist, patient was adviced chemotherapy for burkitts lymphoma. Patient was started initially on CHOP regimen (cyclophosphamide+hydroxydaunorubicin+oncovin+pred nisolone) for first cycle. Then COMP regimen (cyclophosphamide+oncovin (vincristine) +methotrexate+prednisolone) was given for second cycle. Post chemotherapy appearance After 2 chemotherapy cycles proptosis is resolved completely. But vision dropped to perception of light due to exposure keratopathy.

CONCLUSION

There are 3 clinical subtypes of Burkitt's lymphoma.

1. Endemic subtype -The highest incidence of Burkitt's lymphoma (50 to100 cases per million) occurs in equatorial Africa, Endemic (African) BL primarily affects African children aged 4–7, is twice as common in boys compared with girls where it is highly associated with the Epstein–Barr virus, a human lymphotropic herpes virus. In the endemic subtype- host is unable to generate an adequate T-cell cytotoxic response to Epstein–Barr virus due to a concomitant infection with malaria or another infection results in ongoing B-cell proliferation leads to gene translocation and monoclonal proliferation. Endemic Burkitt's lymphoma most commonly presents in the jaw, abdomen, orbit and paraspinal area.[1-5]
2. Sporadic form - is relatively rare (2 to 3 cases per million). Less associated with Epstein–Barr virus infection and commonly presents in children with abdominal tumors, causing swelling, pain, and obstruction. Orbital involvement is rare.(6)

Sporadic (non-African) BL accounts for 1–2% of adult lymphoma cases worldwide. In the US and Western Europe, 40% of paediatric lymphoma cases are reported as sporadic BL[8]

3. Immunodeficiency associated- which is presented in our case. Immunodeficiency-associated BL is the most prevalent form of BL occurring in individuals with HIV/AIDS, accounting for 30–40% of non-Hodgkin lymphoma in these patients and being described as an AIDS-defining disease. It also can occur in people with congenital immune deficiency disorders and patients receiving immunosuppressive drugs after organ transplant.[9]

On Histology- Burkitt's lymphoma shows small homogenous lymphoid cells with basophilic cytoplasm, coarse chromatin, round to oval nuclei, and an increased number of mitotic figures. Interspersed macrophages with phagocytized cellular debris yield a "starry sky" appearance. Diagnosis of Burkitt's lymphoma should be considered in patients presenting with painful proptosis. Because of the rapid progression and good response to treatment, diagnostic work-up including biopsy of tissue, should be initiated immediately. Burkitt's lymphoma is becoming common cause of proptosis in immunocompromised patients specially in early age, so possibility of it should be kept in mind while evaluating such patients with proptosis.

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