Document heading doi: 10.21276/apjhs.2019.6.1.22 **Case Report**

Primary Burkitt's lymphoma of medial part of clavicle- an extremely unusual site

Nupur Rastogi*, M. Saxena, R.K. Tanwar, H. Goyal,B.Saxena

Private Diagnostic Centre, Dr. Nupur's Lab, Kota, Rajasthan, India

Received: 05-02-2019 / Revised: 25-02--2019 / Accepted: 15-03-2019

Abstract

Burkitt's lymphoma is a subgroup of Non-Hodgkin's lymphoma of high grade which is aggressive and composed of diffuse, undifferentiated malignant cells of lymphoid origin which are small and non cleaved. Clinical course is aggressive and rapid, commonly occurs in children and young adults with frequent involvement of Bone Marrow and CNS. It is considered as a medical emergency requiring immediate diagnostic and therapeutic intervention. We report a case of Burkitt's lymphoma of medial part of clavicle extending into overlying pectoralis muscles, intercostal spaces, superior mediastinum, supraclavicular region. As per the literature available this is the first case of Burkitt's lymphoma presenting primarily in medial part of clavicle.

Keywords: Medial part of clavicle, Burkitt's lymphoma

Introduction

Dennis Burkitt first described Burkitt's lymphoma in 1956 in equatorial Africa. It is the fastest growing cancers in human, growth fraction close to 100%. Burkitt's lymphoma and Burkitt like lymphoma are highly invasive B-cell Non Hodgkin's lymphoma with a aggressive and rapid progression of the disease and high death rate. For the past few years, the prognosis of the children with Burkitt's lymphoma or BLL has improved, and 2 year disease free survival rate in advanced stage has attained 75-89 % [2]. In the present case, Burkitt lymphoma of medial part of right clavicle is being presented. Primary lymphoma of bone is an extremely rare form of extranodal lymphoma, originally described in 1928 by Oberling, which is limited to the bone or bone marrow without any systemic involvement [3]. It represents around 7 % of all primary malignant bone tumors and less than 1 % of malignant lymphomas. It usually emerges from the medulla, presents as a localized, single lesion and can involve any part of the skeletal body [3].

*Correspondence
Dr. Nupur Rastogi,
31-Royal Town, Khedli Phatak, Kota – 324 001, Rajasthan, India.
E-Mail: nupurkota123@gmail.com

The present case is of Primary Burkitt lymphoma in 17 year old Indian female arising in medial part of right clavicle. As per the literature available and searched on internet, this is the first case of Primary Burkitt lymphoma of clavicle.

Case report

A 17 year Indian female presented with swelling in right supraclaviclar region (Fig 1). She was subjected to Fine needle aspiration. Giemsa stained cytosmears revealed large to medium sized round cells with open nuclear chromatin, irregular nuclear contours, conspicuous nucleoli, vacuolations in cytoplasm. Mitotic figures were seen. Cytological diagnosis of Burkitt's lymphoma was given (Fig 2). She was then sent for CT scan which showed heterogeneously enhancing large soft tissue density mass lesion in right medial clavicular region with poor fat planes involving medial clavicle, infiltrating into overlying pectoralis muscles and with obliteration of subcutaneous spaces and minimal cutaneous edema (Fig 3).

Lesion infiltrating into intercostal spaces, superior mediastinum, abutting SVC without significant luminal narrowing. Lesion show area of necrosis and internal vascularity.

Multiple variable sized axillary, supraclavicular, superior mediastinum, pre, paratracheal, subcarinal and AP window lymphnodes were seen, largest measuring 12 mm in size. H&E sections of the histopathological

tissue showed large round cells with irregular nuclear contours,open nuclear chromatin , conspicuous nucleoli, vacuolated cytoplasm. Mitotic figures and tingible body macrophages giving starry sky appearance were seen (Fig 4). S. LDH level was raised. All other laboratory investigations were within normal range. Immunohistochemistry was done for confirmation . It was positive for CD20, CD10. Bcl-6.Ki-67 > 80 %.Negative for CD3, CD5, CD34. Bone marrow aspiration was done for staging which also revealed singly scattered cells suggestive infiltration of similar lymphoma cells (Fig 5). Thus diagnosis of Primary Burkitt's lymphoma of medial part of right clavicle was established.

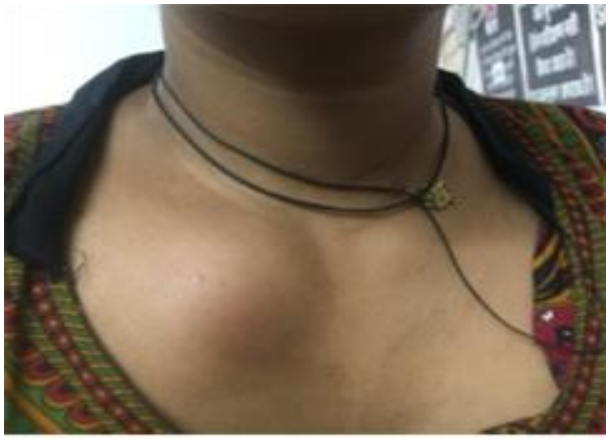

Fig 1: Clinical photograph of swelling in right supraclavicular region

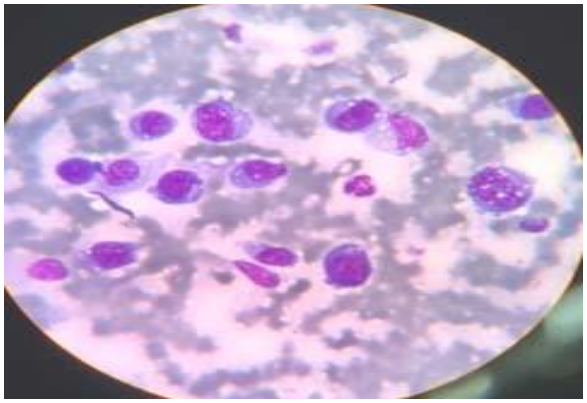

Fig 2:Cytosmears,40X,show round cells with high N:C ratio, vacuolated cytoplasm

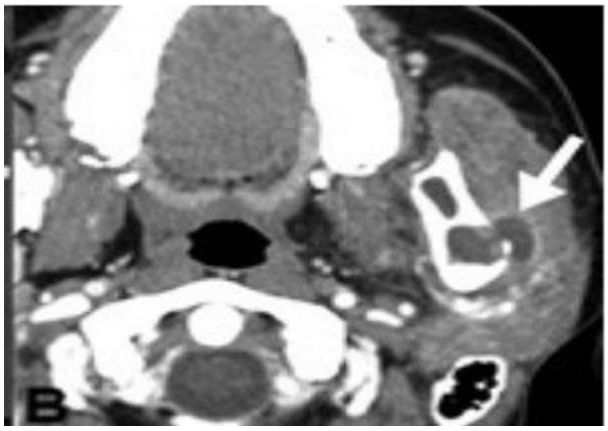

Fig 3.CT scan showing lesion arising from medial part of right clavicle

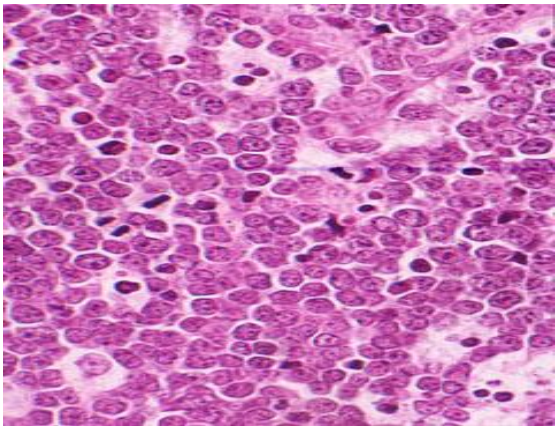

Fig4: H&Esection showing starry sky appearance.

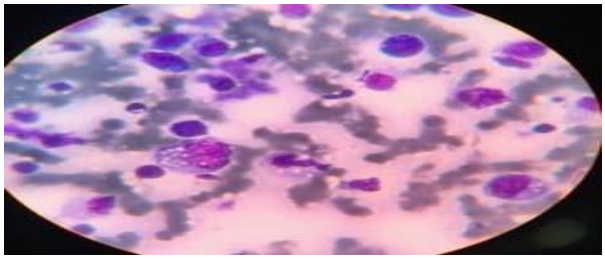

Fig 5: Bone Marrow ,Giemsa stain,40 X showing presence of lymphoma cells

Discussion

Primary bone lymphoma is characterized as a lymphoma of the bone or bone marrow without proof of any simultaneous systemic involvement[3]. As per Coley's criteria, 1950, Peripheral blood smear is diagnosed by lymphoma in an osseous site with no evidence of disease elsewhere for atleast 6 months after diagnosis. Presence of regional lymph node without histological examination, does not exclude diagnosis of PBL [4]. Ostrowski et al in 1987, reclassified bone lymphoma into 4 subgroups: Group 1- solitary primary bone lymphoma, Group 2- more than one bone is affected by no nodal or visceral disease, Group 3- distant nodal disease, Group 4- Visceral disease. According to WHO classification of tumors of soft tissue and bone, 2002, the criteria for a diagnosis of PBL are (1)- a single skeletal tumor with or without regional lymph node involvement (2)- multiple bone lesions without visceral or lymph node involvement [3,5].Burkitt's lymphoma is highly aggressive B-cell NHL, characterized by the translocation and deregulation of the c-myc gene on chromosome 8[6].
Common sites of Burkitt lymphoma are jaw, facial bones, distal ileum, caecum, ovaries, kidney, breast. Two variants of Burkitt lymphoma are sporadic and endemic. Separate staging system for BL has been developed by Ziegler (1981), Levine et al (1982), classified the cases of 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 2 separate masses or disease on both sides of the diaphragm.Stage IV- pleural effusion, ascites or involvement of the lymph nodes (malignant cells in the CSF) or bone marrow. There is no racial predilection, males are affected 2-3 times more than females. Primary Burkitt's lymphoma of bone is uncommon. Primary BL of medial part of clavicle is not been reported as per the literature available. BL is one of the most rapidly proliferating neoplasm with doubling time of as short as 25 hours [8]. Cytologically and histologically Burkitt's cells are homogenous in size and shape, with round to oval nuclei and slightly coarse chromatin with multiple nucleoli and intensely basophilic vacuolated cytoplasm containing neutral fat. The hallmark of BL is the presence of starry sky appearance due to presence of scattered macrophages phagocytizing cell debris and apoptotic cells. Immunohistochemistry is positive for Ki-67, CD-19, CD-20, CD-22, CD-79a which is useful for diagnosis. Bone marrow and CNS involvement are reported in 30-38 % and 13-17 % of adults with Burkitt lymphoma, respy [9]. With aggressive chemotherapy, complete remission rates are 75-90 % and overall long-term survival rate is 50-70 % in adults [8]. Massive acute destruction of the tumor cells during initial chemotherapy due to rapid growth rate may result in tumor lysis syndrome. In the present case, patient was given chemotherapy, taking care of tumor lysis syndrome. After 6 months of chemotherapy, patient is well with no residual disease.

Conclusion

Fine needle aspiration cytology is the best mode of investigation for early, accurate, minimally invasive for diagnosis of Burkitt' lymphoma. Histopahtology and IHC are further diagnostic tools for confirmation of the diagnosis. Early diagnosis can be life saving for Burkitt lymphoma. As per the available literature, this is the FIRST case of Primary Burkitt's lymphoma involving the medial part of right clavicle.

Acknowledgement

Patient and his relatives.

References

1. S.K. Verma, Burkitt lymphoma: Thorax to Pelvis, Austin J Clin Case Rep, 2014;1(9):1044
2. N Jing LIN, W Zheng, et al. Clinical Analysis of 13 cases with Burkitt Lymphoma or Burkitt-like Lymphoma, ClinOncol Cancer Res.2010;7:110-115
3. Bhagavathi S, Fu K. Primary Bone Lymphoma. Archives of Pathology & Laboratory Medicine.2009;133:1868-1871.
4. Rahmat K, Wastie M, Abdullah B.Primary bone lymphoma: report of a case with multifocal skeletal involvement. Biomedical Imaging and Intervention Journal.2007: 3.
5. Unni KK, Hogendoorn PCW (2002) Malignant lymphoma. In: Fletcher CDM, Unni KK, Mertens F. eds. Pathology and Genetics of Tumors of Soft Tissue and Bone. Lyon, France: IARC Press. World Health Organization Classification of Tumors
6. Ali H Kanbar, In: DeVita VT Jr, Hellman S, et al. Cancer: Principles and Practice of Oncology,2.5thedu.Philadelphia, Pa:Lippincott Williams 7a Wlikins, 1997;2157-9
7. Patton LL, McMillan CW, Webstar WP. American Burkitt's Lymphoma: a 10 year review and case study, Oral Surg Oral Med Oral Pathol, 1990; 69: 307-316

8. K.A. Blum, G. Lozanski and J.C. Byrd, Ädult Burkitt leukemia and lymphoma", Blood 2004;104(10):3009-3020

9. M.L. McMaster, J.P. Greer, F.A. Greco, D.H. Johnson, et al, " Effective treatment of small noncleaved-cell lymphoma with high intensity, brief duration chemotherapy, "Journal of Clinical Oncology,1991;9(6):941-946

Conflict of Interest: None
Source of Support: Nil