Case report

Burkitt's lymphoma of medial part of clavicle: Case report of unusual localization

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ABSTRACT

Introduction: Burkitt lymphoma is a form of non-Hodgkin’s lymphoma in which cancer starts in immune cells called B-cells. Recognized as a fast-growing human tumor, it is considered as a medical emergency requiring immediate diagnostic and therapeutic intervention. However, intensive chemotherapy can achieve long-term survival in more than half the people with Burkitt lymphoma.

Case report: We report a case of Burkitt’s lymphoma of medial part of clavicle was diagnosed in a 60 years year-old man and review the literature concerning the clinical features, radiological appearance, histopathological findings and treatment options.

Discussion: Clinical course of Burkitt lymphoma is aggressive and rapid, commonly occurs in children and young adults with frequent involvement of Bone Marrow, associated with impaired immunity and is rapidly fatal if left untreated, early diagnosis can be life saving for Burkitt lymphoma. As per the literature available this is the second case of Burkitt’s lymphoma presenting primarily in medial part of clavicle.

1. Introduction

Dennis Burkitt first described Burkitt's lymphoma in 1956 in equatorial Africa. Burkitt's lymphoma is highly invasive B-cell Non-Hodgkin’s lymphoma with aggressive and rapid progression of the disease and high death rate. It is the fastest growing cancers in human.

Management of Burkitt lymphomas in adults is inspired by pediatric protocols given the good results of these. Efforts have been made to minimize the toxicity of chemotherapy. Burkitt's lymphoma have rapid proliferation capacity, therefore the proposed protocols are based on a combination of high-intensity and short-duration chemotherapy that has shown extremely effective for a high proportion of patients with Burkitt's lymphoma.

In the present case, Burkitt lymphoma of medial part of left clavicle is being presented. Primary lymphoma of bone is an extremely rare form of extranodal lymphoma, which is limited to the bone or bone marrow without any systemic involvement [1]. 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 [1]. As per the literature available, this is second case of Primary Burkitt lymphoma of clavicle. The work has been reported in line with the SCARE 2020 criteria [9].

2. Case summary

We report a case of a 60-year-old man, seen in our institution for swelling on the left side of the neck of one-year duration; the swelling had increased in size over that period of time. There was no history of trauma, local, systemic infection, or surgical intervention. He had no shortness of breath, no dysphagia, or hoarseness. The patient reports a slow increase in size over the last few months. A physical examination revealed a firm and tense mass on the lower left of the neck in the clavicular and sub clavicular region measured approximately 5.0 × 5.0
cm (Fig. 1). There was no abnormal pulsation or bruit. The rest of the clinical exam was normal CT scan; MRI showed heterogeneously enhancing large soft tissue density mass lesion in left medial clavicular region with area of necrosis and internal vascularity (Fig. 2). His hematology, serum chemistries, and lactate dehydrogenase were normal. A biopsy from the lesion under general anesthesia showed sheets of round cells with scanty cytoplasm, round nuclei, and immature chromatin (Fig. 3). Immunohistochemistry was positive for CD20, CD10. Negative for CD3, CD5, a diagnosis of Burkitt's lymphoma was made. Further, staging work-up including a CT scan of the neck, chest, abdomen, and pelvis, a bone marrow biopsy, did not reveal any disease elsewhere. Lumbar puncture was performed and showed no infiltration of central nervous system (CNS). Serology of HIV was negative. Pet scan showed increased uptake at the primary site only. Using Murphy staging system, he is included to Group A. He received intense chemotherapy according to LMB regimen associated to Rituximab; complete remission was achieved after RCOPADM1, and continues to be in remission at 6 months.

3. Discussion

Primary bone lymphoma is characterized as a lymphoma of the bone or bone marrow without proof of any simultaneous systemic involvement and no evidence of disease elsewhere for at least 6 months after diagnosis [1]. 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 [1,2].

Burkitt's lymphoma is highly aggressive B-cell NHL, characterized by the translocation and deregulation of the c-myc gene on chromosome 8 [3]. Common sites of Burkitt lymphoma are jaw, facial bones, distal ileum, caecum, ovaries, kidney, and breast. Three variants of Burkitt lymphoma are described; sporadic and endemic variant. Third form of BL is now found in Africa, HIV-associated Burkitt's lymphoma, occurring specially adults [3]. Separate staging system for BL has been developed by Ziegler (1981), Levine et al. (1982), classified the cases of American BL as follows [4].

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.

Precise diagnosis of BL is based on histologic, immunophenotypic, and genetic features. 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 (the proliferation fraction is closely 100%), CD-19, CD-20, CD-22, CD-79 which is useful for diagnosis. Cell express also CD10, Bcl-6, cd43 but not CD5, CD23, Bcl-2, CD138 OR TdT, the time of doubling of tumors is very short, between 24 and 48 h [5]. A key feature in the determination of Burkitt's lymphoma is the presence of a translocation between the c-myc gene and the IgH gene. This anomaly is found in 80% of cases: t (8; 14), sometimes it is a translocation between c-myc and the kappa or lambda light chain (Igl) gene in the remaining 20%: t (2; 8) or t (8; 22), respectively. Bone marrow and CNS involvement are reported in 30–38% and 13–17% of adults with Burkitt lymphoma [6]. Prognostic factors with direct impact on the evolution of the disease are not yet determined, such as, some characteristics that have been associated with less favorable outcomes in adults and children include advanced age, advanced stage, index poor performance, bulky disease, elevated LDH, and CNS or bone marrow involvement [7]. For our patient, age is the unique prognosis factor, he had no advanced stage, LDH rate was normal, no bulky disease, the bone marrow and CNS were not infiltrated. A 2 year survival of 70% was reported in adult patients with Burkitt's lymphoma by Divine et al. [6].

Massive acute destruction of the tumor cells during initial chemotherapy due to rapid growth rate may result in tumor lysis syndrome. Intensive chemotherapy inspired by pediatric regimen make possible to achieve complete remission for adult patients. The association of the chemotherapy to a monoclonal anti-CD20 antibody (Rituximab) improved outcome for BL's patients, combination of chemotherapy and rituximab for adults' patients treated for BL became usually used by many centers, association. Can include R/LMB regimen, R-HyperCVAD or other regimen, but the real role of rituximab is not yet determinate and required more investigations [8]. In the present case, patient was given chemotherapy associated to antiCD20 Antibody. Complete remission was achieved and 6 months after of the end of chemotherapy, complete remission is maintained and patient is doing well with no residual disease.

4. Conclusion

This case demonstrates that even though rare, clavicle can be the primary site for Burkitt's lymphoma and it should also be considered in the differential diagnosis of bone tumors, our patient is probably the second reported case of primary Burkitt's lymphoma involving the medial part of clavicle as the only site of disease. He received intense treatment and continues to be in remission at 6 months.
Ethical approval

Written informed consent was obtained from the patient for publication of this case report and accompanying image.

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CRediT authorship contribution statement

Said Anajar: Corresponding author
Mounia bandari and Ahnach maryame: writing the paper
Fouad benariba: study concept

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Registration of research studies

Not “First Man” studies, this is the second case of Burkitt’s lymphoma presenting primarily in medial part of clavicle.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying image.
Provenance and peer review

Not commissioned, externally peer-reviewed.

Declaration of competing interest

All the authors have no personal or financial conflicts of interest regard this case report.

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