

Burkitt's Lymphoma of Right Maxillary Sinus: Case Report

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Abstract Introduction: Burkitt's lymphoma (BL) is an endemic malignant neoplasm with a mandibular localisation, first described, in 1958, in African children. In paranasal BL, maxillary sinus is most commonly involved and sphenoid-ethmoidal sinuses are less commonly involved. The aim of this study, we describe here a rare case of BL in the right maxillary sinus. **Case Report:** A 23-year-old female presented with a history of right nasal obstruction after pregnancy. In maxillary CT scan a heterogeneous large solid mass showed in the right maxillary sinus which expands this sinus and extends into the right nasal cavity and right ethmoid. She underwent surgical biopsy. In pathology of right maxillary sinus mass biopsy consistent with high grade diffuse small non cleaved NHL. She treated with combination chemotherapy regimen of CODOX-MVAC with triple intrathecal four courses suggested for her. Then she treated with maintenance therapy with thioguanin and MTX and now she is alive for 3 years after last treatment without any new drug and off all treatment and only is in follow up. **Conclusion:** BL in the right maxillary sinus is very rare but we will have a more successful treatment if it is diagnosed early.

Keywords: BL, CODOX-MVAC, intrathecal, maxillary sinus

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1. Introduction

Burkitt's lymphoma (BL) is an endemic malignant neoplasm with a mandibular localisation, first described, in 1958, in African children [1]. BL is a rare and rapidly progressive form of B-cell non-Hodgkin's lymphoma (NHL) that most commonly occurs in males during childhood and young adult life [2]. BL in the head and neck usually presents as lymphadenopathy, whereas primary involvement of the nasal cavity and paranasal sinuses is uncommon. In paranasal BL, maxillary sinus is most commonly involved and sphenoid-ethmoidal sinuses are less commonly involved [3]. The aim of this study, we describe here a rare case of BL in the right maxillary sinus.

2. Case Report

A 23-year-old female presented with a history of right nasal obstruction after pregnancy. In maxillary CT scan a heterogeneous large solid mass showed in the right maxillary sinus which expands this sinus and extends into the right nasal cavity and right ethmoid (Figure 1).

There is localized maxillary bone sinus rim destruction. The mass significantly extended into retrobulbar space of

the right orbit and leads to right sided proptosis. It also significantly involves the right optic and compresses the right optic nerve. She underwent surgical biopsy. In pathology of right maxillary sinus mass biopsy consistent with high grade diffuse small non cleaved NHL (Figure 2).

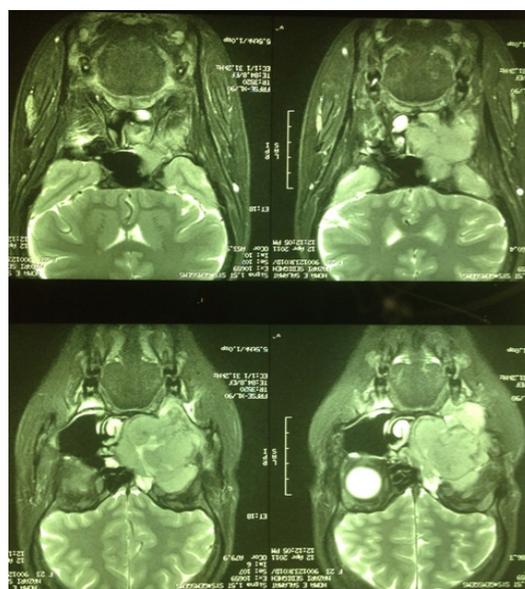


Figure 1. A localized heterogeneous mass in the right maxillary sinus (in the diagnosis)

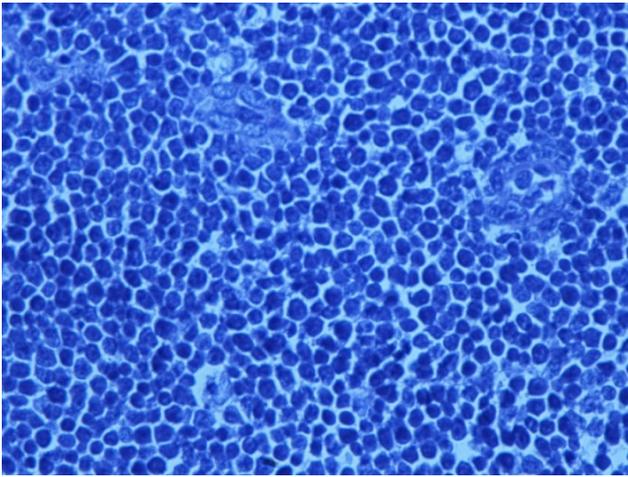


Figure 2. The neoplastic cells are medium-sized and show “squaring off” of the nuclear membrane accompanied by typical coarse chromatin, multiple distinct nuclei and frequent mitoses. Also there is starry sky appearance.

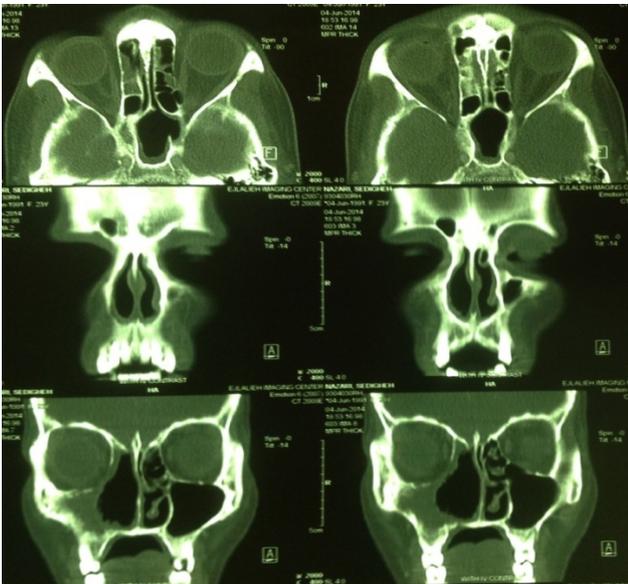


Figure 3. A heterogenous thickening seen in the right maxillary sinus (three months after treatment)

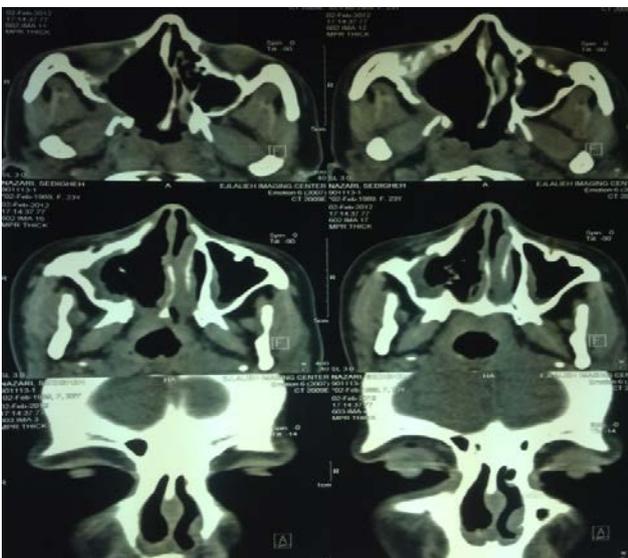


Figure 4. A mild thickness in the right maxillary sinus (six months after treatment)

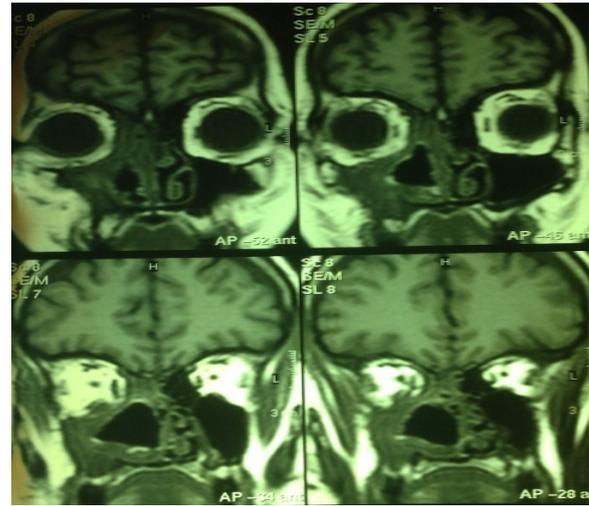


Figure 5. A mild thickness in maxillary sinus

Immunohistochemistry (IHC) is recommended for definite diagnosis. Markers in the IHC report consist of Ki67 (high proliferative) CD10, BCL-2, CD20 and CD99>90% were, positive, in tumor cells respectively. Hematoxylin and Eosin (*H&E*) and IHC studies are consistent with definit diagnosis BL. She treated with combination chemotherapy regimen of CODOX-MVAC with adriamycin, cyclophosphamide, vincristine, methotrexate, etoposide and dexamethasone and ifosfamide with triple intrathecal (methotrexate plus cytarabine plus hydrocortisone) for four courses suggested for her (Figure 3).

She had undergone twenty sessions radiotherapy (Figure 4). Now she had a slight haze in the ethmoid and sphenoid sinuses and swelling left eye and she treated with maintenance therapy with thioguanin and MTX and at now she is alive for 3 years after last treatment without any new drug and off all treatment and only is in follow up (Figure 5).

3. Discussion

NHLs of the sinonasal tract are uncommon malignancies representing 3% to 5% of all malignancies [4]. Histologically, “starry-sky” pattern is a microscopic hallmark. Immunophenotypically, Burkitt’s lymphoma expresses B-cell lineage markers, including CD19, CD20, CD22, CD74, and CD79a, and coexpressing CD10, Bcl-6, CD43, and p53, but not CD5, CD23, Bcl-2, CD138 or TdT. Almost all cells are positive for Ki-67. EBV titers are usually negative in patients with the sporadic type [5]. In our report Ki67 (high proliferative) CD10, BCL-2, CD20 and CD99>90% were, positive, in tumor cells respectively. Following the success and high cure rates with the CODOX-M/IVAC regimen in patients with similarly aggressive disease, we decided to investigate the use of a similar protocol in patients with relapsed or refractory NHL [6]. IVIC was designed as a salvage regimen which, when it proved moderately effective, was used as part of initial therapy [2]. Treatment consisted of 6 5-day chemotherapy cycles with high-dose methotrexate, high-dose cytosine arabinoside, cyclophosphamide, etoposide, ifosfamide, corticosteroids, and triple intrathecal therapy [7]. She treated with combination chemotherapy regimen of CODOX-MVAC with

adriamycin, cyclophosphamide, vincristine, methotrexate, etoposide and dexamethasone and ifosfamide with intrathecal. Early diagnosis of primary lymphoma in this region is challenging [4]. If treatment is delayed, Burkitt's lymphoma can spread to other parts of the body, leading to poor prognosis. If Burkitt's lymphoma involves the paranasal sinuses, it can cause facial deformity [5]. In most trials, the results for elderly Burkitt patients are inferior to those of the younger cohorts [7].

4. Conclusion

BL in the right maxillary sinus is very rare but we will have a more successful treatment if it is diagnosed early.

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