Primary Extranodal Non-Hodgkin's Lymphoma of Maxillary Sinus: Rare Incident

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Abstract

Non-Hodgkin lymphoma (NHL) commonly arises from lymphoid organ and tissues. But up to 35% of cases have been reported to primarily originate from extranodal sites. Primary paranasal sinus lymphoma accounts for 8% of all paranasal malignancies and a mere 2% of all primary extranodal lymphoma. We described a 72-year-old woman with primary extranodal diffuse large B-cell lymphoma (DLBCL) originated from maxillary sinus extended into adjacent sinuses and structures. She presented with painful, numb, vague, diffuse, hard swelling over right medial canthus since two months. There was no diplopia. She had right epiphora since two weeks. Minimal proptosis was seen in the right eye. There was no history of foul smelling nasal discharge or obstruction. There was no neck swelling. She did not have underlying comorbid illnesses. Rigid nasal endoscopy revealed a friable mass arising from the right osteomeatal complex (OMC) occupying the entire right side of nasopharynx. Computed tomography (CT) revealed a soft tissue mass occupying the entire right maxillary sinus extending into adjacent structures. Treatment was completed and patient remained in remission. Features pertaining to demographic, markers, treatment regimes, and outcomes are discussed in detail.

Keywords: Maxillary diseases, maxillary sinus neoplasms, lymphoma, Non-Hodgkin

INTRODUCTION

Non-Hodgkin lymphoma (NHL) typically arises from lymphoid tissues and organs: lymph nodes, spleen, and thymus. But 25%–35% of NHL have been reported to be originated from extranodal organs, with gastrointestinal tract being the most common (1). Primary paranasal sinus lymphoma accounts for 8% of all paranasal malignancies and a mere 2% of all primary extranodal lymphoma (2, 3).

We describe a patient with primary extranodal diffuse large B-cell lymphoma (DLBCL) originated from maxillary sinus extended into adjacent sinuses and structures.

CASE PRESENTATION

A 72-year-old woman presented to ENT department with painful, numb, vague swelling over right medial canthus since two months (Figure 1). The swelling was diffuse and hard in consistency. There was no diplopia, and ocular movements were normal. She had right epiphora since two weeks. Minimal proptosis was observed in the right eye. There was no history of foul smelling nasal discharge or obstruction. There was no neck swelling. She did not have underlying comorbid illnesses.

Rigid nasal endoscopy revealed a friable mass arising from the right osteomeatal complex (OMC) occupying the entire right side of nasopharynx.

Computed tomography (CT) revealed a soft tissue mass occupying the entire right maxillary sinus extending to right anterior ethmoid, right frontal sinus, right nasolacrimal duct, right preseptal region, and medial extraconal space of right orbit resulting in right globe being abutted anteriorly. The mass extended into right osteomeatal complex and right infratemporal space. There was no intracranial extension but bilateral fossa of Rusenmuller fullness seen. The preseptal component of the mass measured 2.2 cm × 3.1 cm × 2.4 cm (Figure 2, 3). CT staging revealed no distant metastasis. Blood investigations including lactate dehydrogenase were normal.

Histopathology of the tissue from the right nasopharynx and OMC revealed lymphoid cells with positive immunohistochemical stain for CD20 but negative for CD3, CD10, CD30, CKMNF, and CKAE1 indicating NHL-DLBCL subtype.

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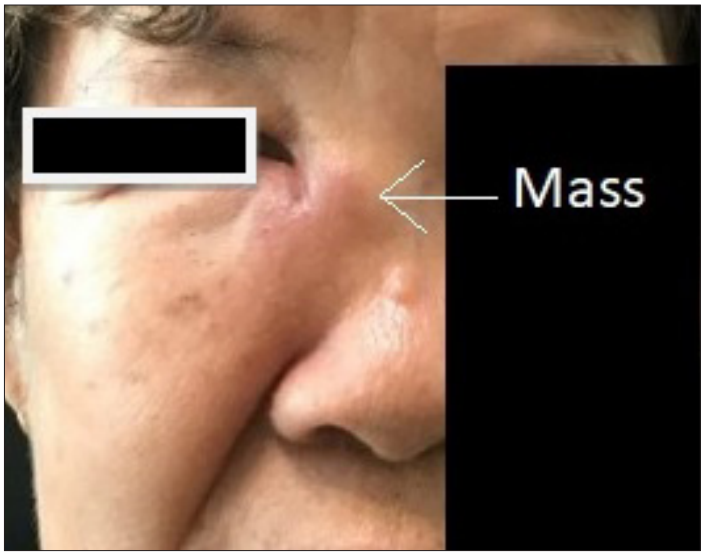

Figure 1. Mass at right medial canthus

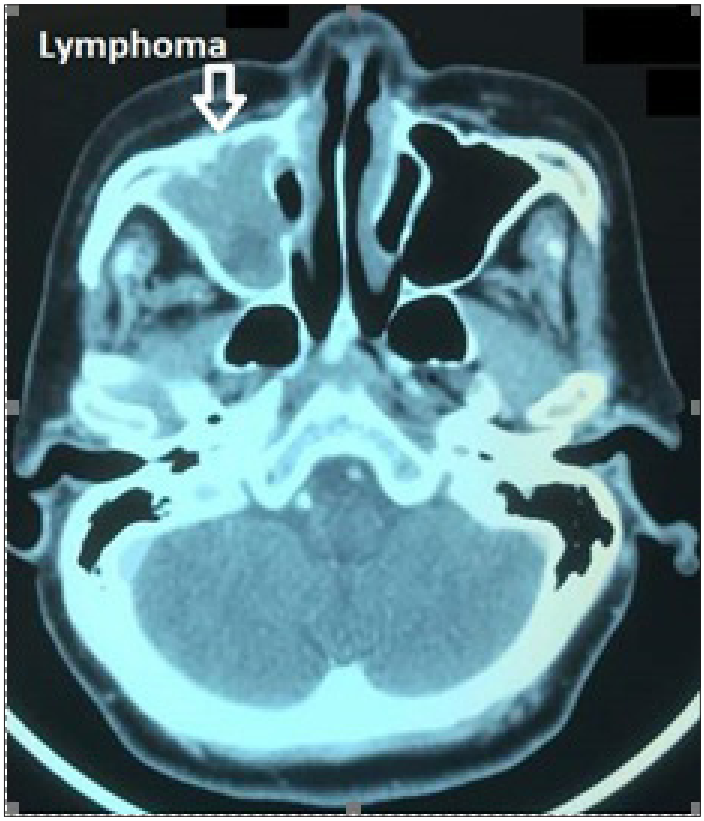

Figure 2. CT axial of skull revealed right maxillary sinus mass

Her disease was stage IE according to Ann Arbor staging. She is currently in complete remission since three months after six cycles of rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisolone (R-CHOP) chemotherapy regime. No significant debilitating side effects of chemotherapy were observed in this patient. Verbally informed consent was obtained from the patient who participated in this study.

DISCUSSION

Non-Hodgkin lymphoma of the head and neck peak incidence between the ages of 50 and 60 years, predominantly in males (3, 4). Maxillary sinus

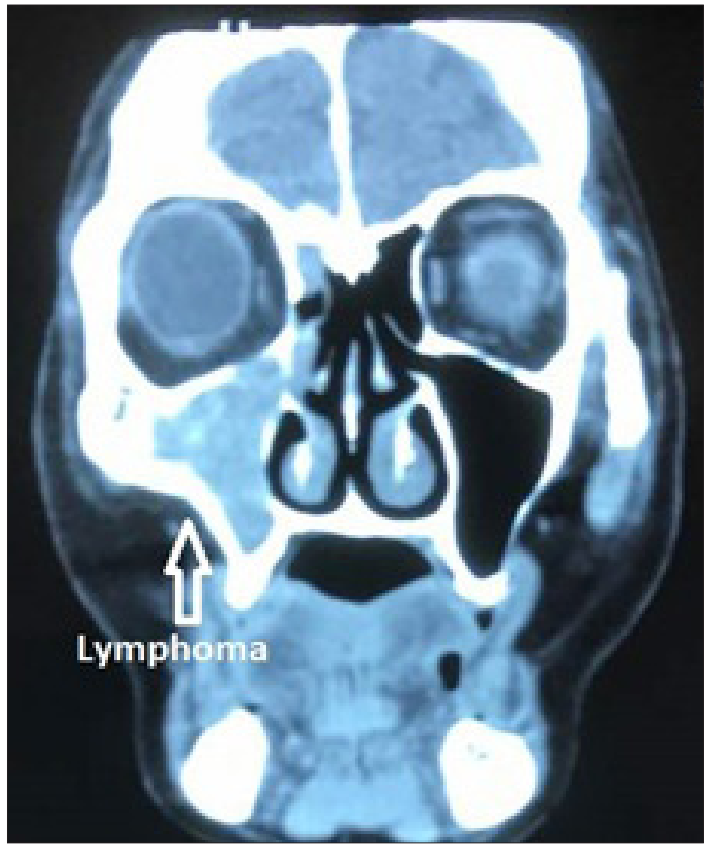

Figure 3. CT coronal skull with right maxillary sinus mass

was reported to be most affected among the paranasal sinuses by NHL, and the highest incident is DLBCL histologic subtype (3-5). B-cell lymphoma are generally less aggressive and therefore prognosis is better (2). Similar cases reported in international literatures featured local disease invasion but no distant metastasis or lymphadenopathy at time of initial diagnosis (2, 3, 6).

High expression of CD20 on immunohistochemical staining has been observed in several reported cases (2, 4-6). This phenomenon has also been observed in non-paranasal sinuses DLBCL. This indicates a significant diagnostic marker for DLBCL.

The LDH levels are raised in only a very small fraction of patients with lymphoma, and this casts doubt over its role in diagnosis and as a prognostic factor (7). LDL was not raised in our patient as well, which resulted in the lack of suspicion of a valid differential diagnosis endoscopically and radiologically.

There is no different treatment regime for extranodal NHL of paranasal sinuses in origin as compared to those of primary nodal disease. Similar cases in literature also reported success with the same chemotherapy regime used in our case (2, 3, 6, 7). Although uncommon, concomitant radiotherapy and radiotherapy alone are practiced as well, but no data are available to determine if they are superior to the R-CHOP chemotherapy regime (3). Complete remission was observed in approximately 70% of patients treated with a CHOP regimen, while overall survival is approximately 60% at five years in a series of non-maxillary sinus specific paranasal sinus DLBCL (4, 7). Local and central nervous system relapse of the disease was at about 40% with the earliest was at eight months after therapy completion (7). Majority of these relapse cases presented with early stages of disease initially (7).

CONCLUSION

R-CHOP chemotherapy regime is the most common first-line treatment globally for primary paranasal sinus DLBCL. Paranasal sinus masses should warrant lymphoma as a differential diagnosis especially if dermographic fits.

Informed Consent: Verbally informed consent was obtained from the patient who participated in this study.

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Conflict of Interest: The authors have no conflicts of interest to declare.

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REFERENCES

1. Newton R, Ferlay J, Beral V, Devesa SS. The epidemiology of non-Hodgkin's lymphoma: Comparison of nodal and extra-nodal sites. Int J Cancer 1997; 72: 923-30. [CrossRef]
2. Catalfamo L, Nava C, Matyasova J, De Ponte FS. Primary Diffuse Large B-Cell Lymphoma of the Ethmoido-Orbital Region. J Craniofac Surg 2013; 24: 602-4. [CrossRef]
3. Yoon YH, Park WY, Choi YJ, Cho KS. Synchronous, primary, diffuse, large B-cell lymphomas involving the ethmoid sinus and epiglottis: a rare clinical entity. Laryngoscope 2013; 123: 702-4. [CrossRef]
4. Doumas S, Sakkas L, Panayiotidis P, Wozniak G, Vlychou M, Vassilopoulos G. Favorable outcome in non-Hodgkin lymphoma of the maxillary sinus treated with R-CHOP. Arch Med Sci 2014; 10: 406-9. [CrossRef]
5. Buchanan A, Kalathingal S, Capes J, Kurago Z. Unusual presentation of extranodal diffuse large B-cell lymphoma in the head and neck: description of a case with emphasis on radiographic features and review of the literature. Dentomaxillofac Radiol 2015; 44: doi: 10.1259/dmfr.20140288. [CrossRef]
6. Adwani DG, Arora RS, Bhattacharya A, Bhagat B. Non-Hodgkin's lymphoma of maxillary sinus: An unusual presentation. Ann Maxillofac Surg 2013; 3: 95-7. [CrossRef]
7. Oprea C, Cainap C, Azoulay R, Assaf E, Jabbour E, Koscielny S, et al. Primary diffuse large B-cell non-Hodgkin lymphoma of the paranasal sinuses: a report of 14 cases. Br J Haematol 2005; 131: 468-71. [CrossRef]