A palatal swelling transpires out as a nasal B-cell Non-Hodgkin's Lymphoma

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Abstract

Primary sinonasal Non-Hodgkin's Lymphoma's (NHLs) are quite rare and emulate the presentation of benign inflammatory diseases. It is challenging to distinguish them morphologically and radiologically from other malignant neoplasms.

We report a 37-year-old male patient who presented with obstruction of the nasal passages, rhinorrhoea, epistaxis, post nasal drip, facial swelling, orbital symptoms and pyrexia. The mass was a nasal diffuse large B-cell lymphoma confirmed by immunohistochemistry. After the first cycle of chemotherapy was started, the patient improved with resolution of the facial swelling, pain and visual defects.

A high index of suspicion is required to differentiate sinonasal lymphomas from other lesions.

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Case Report

Introduction

Lymphomas constitute a group of malignant neoplasms of the reticuloendothelial system which are divided into Hodgkin's disease and Non-Hodgkin's lymphomas (NHL).[1] NHL is a heterogeneous group of diseases with peculiar, morphological, molecular and phenotypic molecular features (B-cell neoplasms, T-cell neoplasms and putative natural killer (NK)-cell neoplasms). [2]

The nasal cavities and paranasal sinuses are rarely affected by primary NHL. The usual primary extra-nodal sites of lymphomas include bone marrow, bone, stomach, liver, soft tissue, meninges, lower gastrointestinal tract and others.[3] Geographical factors play an important role in the incidence and the histological subtype of sinonasal NHL. In Asian populations, nasal lymphomas are more commonly T-cell lymphomas, whereas B-cell subtypes are frequently more common among the sinonasal lymphomas seen in Western populations.[4]

They emulate the presentation of benign inflammatory diseases. It is challenging to distinguish them morphologically and radiologically from other malignant neoplasms.

Case Summary

A 37-year-old male patient who was referred to our hospital, presented with obstruction of nasal passages, rhinorrhoea, epistaxis, post nasal drip, facial and right hard palate swelling and orbital symptoms for 5 months and pyrexia. The symptoms did not resolve with antibiotics. His right ocular movements were restricted in all gazes.

Endoscopic examination revealed a narrow nasal passage and a mass covered with blood stained pus involving the right middle meatus and extending to the posterior choana of the right nasal passage. The right Fossa of Rossenmuller and nasopharynx were normal.

A CT Scan revealed a right sinonasal tumour with local infiltration seen in all sinuses as well as erosion of the hard palate.

A punch biopsy of the nasal mass was taken which was reported as chronic inflammation. Considering this histopathological finding, endoscopic sinus surgery was performed to eradicate the disease as well as obtain a definite histological diagnosis. The mass was histologically proven as a Nasal diffuse large B-cell lymphoma(DLBCL) [Figures 1 and 2] and confirmed by immunohistochemistry. Immunohistochemically, the cells were strongly positive for CD20 [Figure 3], CD79a, BCL2 [Figure 4], BCL6 [Figure 5] and MUM1 [Figure 6]. CD10 was focally positive. Ki-67 index was <99% [Figure 7].

After confirmation of the histological diagnosis, chemotherapy (R CHOP) was started and with the first cycle, the patient improved with resolution of the facial swelling, pain and visual defects.

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Discussion

These primary NHLs of the sinonasal tract develop in an anatomic space expanding towards the nasal cavity and sinus, without producing any symptoms at the early stages and hence an early diagnosis is unusual. The presenting symptoms appear only when adjoining anatomic structures are involved and the tumours reach a considerable size, and they may pose as other head and neck or nasal findings.

These lymphomas most commonly present with symptoms of unilateral nasal, facial or cheek swelling and obstruction of the nasal passages.[5] Other, uncommon symptoms include visual disturbances like diplopia or blurred vision and pain in the nasal or cheek regions. Sometimes, a patient will present with congestion of the nasal passages and discharge or a history of a problem resembling chronic sinusitis.

Our patient also presented with nasal congestion and discharge and was suspected to be suffering from chronic sinusitis. This led to him having a delay in treatment and worsening of his symptoms with the onset of facial swelling and visual difficulties.

The patterns on radiology are classically of large tumours with a locally infiltrative growth pattern resulting in the sinus walls showing prominent bony erosion. Destruction of the orbital soft tissue and osseous are usually seen in high-grade B-cell tumours. CT scans are especially useful in evaluating the involvement of the sinuses and orbits and extent of the destruction and should be regarded as the gold standard of management., There is a decrease in recurrence and metastasis with the use of combination chemotherapy, especially an anthracycline-based chemotherapy (i.e. CHOP), and local radiation. This results in an overall improved survival without notable side effects.[5]

Conclusion

Primary sinonasal NHLs are rare neoplasms. In order to achieve optimal treatment results, early diagnosis and staging of these tumours with appropriate treatment are vital, and it is crucial that the ENT surgeon be acquainted with their clinical presentation and management.

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Figures

Figure 1

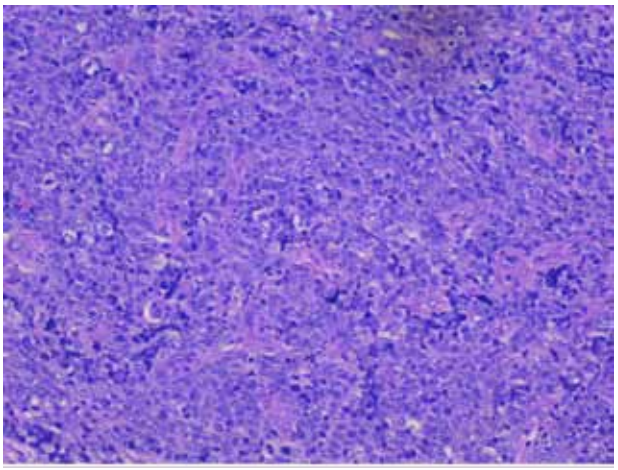


Figure 2

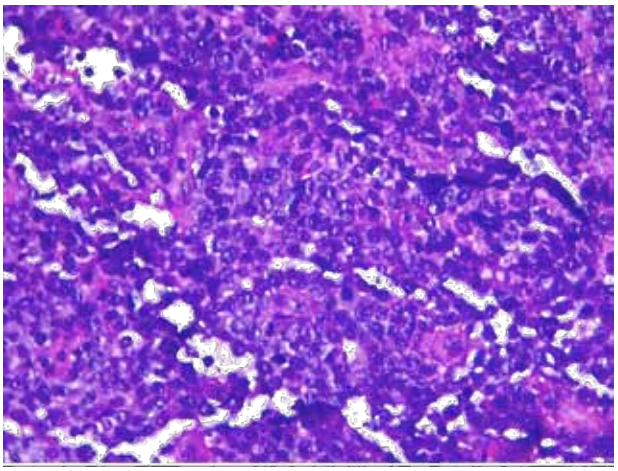


Figure 3

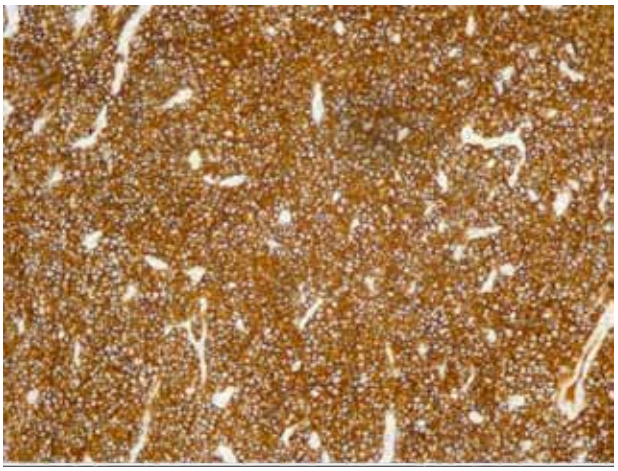


Figure 4

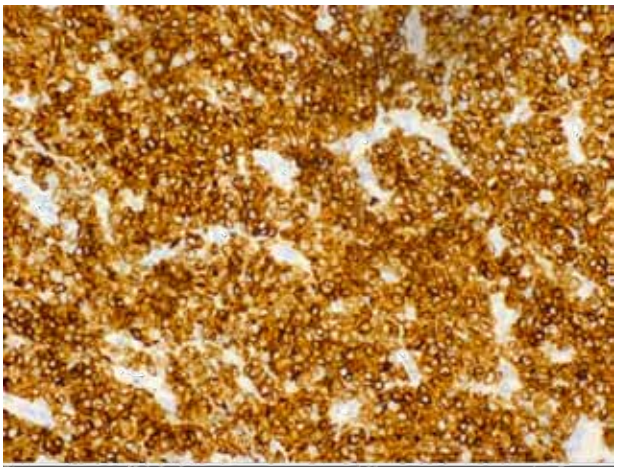


Figure 5

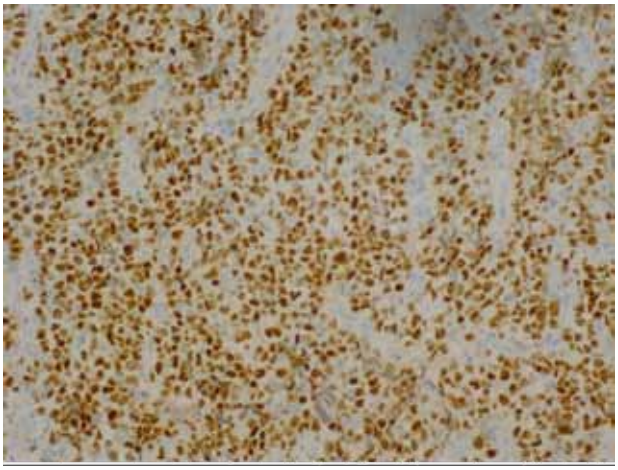


Figure 6

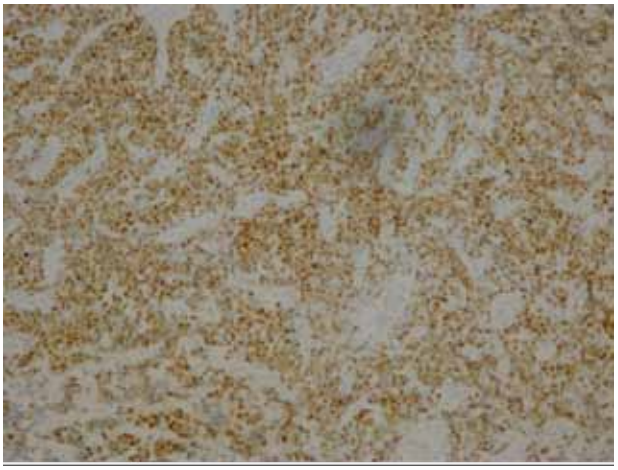
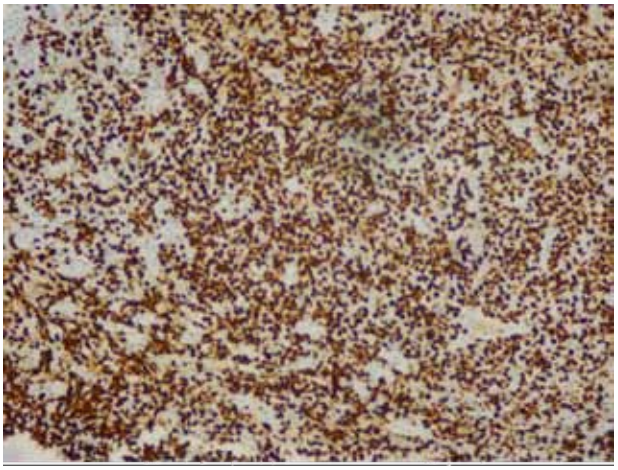


Figure 7



Legends to Figures:

Figure 1: Diffuse large B cell lymphoma. Diffuse infiltration of medium to large size atypical lymphoid cells. H&E stain x200.

Figure 2: Diffuse large B cell lymphoma. Diffuse infiltration of medium to large size atypical lymphoid cells. H&E stain x400.

Figure 3: Diffuse large B cell lymphoma. Immunohistochemistry for the B cell-marker CD20 shows B cell phenotype of the atypical cells. Magnification x 200.

Figure 4: Diffuse large B cell lymphoma. BCL2 staining shows strong cytoplasmic staining.
Magnification x 200.

Figure 5: Diffuse large B cell lymphoma. Neoplastic cells show nuclear staining for BCL6.
Magnification x 200.

Figure 6: Diffuse large B cell lymphoma. Neoplastic cells show nuclear staining for MUM 1.
Magnification x 100.

Figure 7: Diffuse large B cell lymphoma. Immunohistochemistry shows strong staining for Ki67.
Magnification x 100.