

Mantle Cell Lymphoma of Nasal Cavity: A Rare Case Report

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Abstract: A rare case of Mantle Cell Lymphoma of nasal cavity. Index patient is a 50-year-old male presented to us with chief complaints of nasal bleeding, nasal blockage and swelling of cheek following which CECT face and neck was done and biopsy was taken from nasal mass and on histopathology and immuno-histochemistry the diagnosis of Mantle Cell Lymphoma was made. Mantle cell lymphoma of nasal cavity is very rare and only few case reports are there in literature.

Keywords: Cyclin-D1, CD5, CD20.

1. Introduction

Mantle cell lymphoma (MCL) is a type of non-Hodgkin's lymphoma (NHL) which is not so common usually presents at an advanced stage with systemic spread. Mantle cell lymphoma was recognized as a distinct type of NHL by an international consensus conference [1] in 1992 and represents approximately 6% of all non-Hodgkin's lymphomas [2]. Non-Hodgkin's lymphoma of the sinonasal tract is uncommon, accounting for 0.2% to 2.0% of all NHL. [3], [4] However primary nasal lymphomas comprise 3%-10% of cases of NHL in the Asian population.

2. Case report

A 50-year-old male presented in Radiotherapy outpatient clinic with chief complaints of right sided nasal blockage for 2 months, bleeding from the right nostril for one month and right sided maxillary swelling for one month. Initially he received some nasal steroids and topical decongestants by his general practitioner but there was no improvement in his symptoms. He had no history of orbital symptoms such as redness, diplopia, epiphora. There was no past medical history of any significant illness. On examination there was a significant reduction of nasal airway patency in the right nostril. Anterior rhinoscopy revealed a friable mass filling the right nostril completely. Flexible or rigid nasal endoscopy was not possible due to complete occlusion of the nostril. The left nasal airway was patent and appeared normal on rigid endoscopy. The rest of the ENT examination was unremarkable.

Ophthalmological examination did not reveal any evidence

of ophthalmoplegia and the patient had normal visual acuity and colour vision. A computed tomography (CT) scan of the nose and paranasal sinuses demonstrated an elongated soft tissue mass in the right inferior meatus inseparable from the inferior turbinate. The lesion extended into the right maxillary antrum through a defect in its medial wall. The lesion also appeared to have involved the nasolacrimal duct and was extending into the right lacrimal sac (Figure 1).

An examination and biopsy of this right nasal mass was performed under general anesthesia. Histological examination showed mucosal tissue lined by stratified squamous epithelium with orderly maturation. Subepithelium zone shows a tumor in sheets composed of monomorphic small to medium sized atypical lymphoid cells. These cells have round to oval nuclei, clumped chromatin, inconspicuous nucleoli, and scanty cytoplasm (Figure 2).

Immuno-histochemistry staining demonstrated a strong diffuse positivity with B-cell markers CD 20 (Figure 3). CD3 was positive in background cells (Figure 4) and negativity with synaptophysin, CD10, CD30, CD56, panCK. Ki67 (a marker for cell proliferation) showed 40-50% proliferation index (Figure 5). Cyclin D1 was positive (Figure 6) and CD5 was weak positive (Figure 7) confirming the diagnosis as Mantle cell lymphoma. Initially pre chemotherapy PET-CT scan could not be done due to some logistic issues and chemotherapy was started in view of heavy symptom burden.

The patient was referred to radiotherapy department in view of nasal bleeding and received palliative radiotherapy 8Gy in single fraction following which bleeding was stopped and after that patient treated with remaining cycles of intensive immune-chemotherapy with rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone (R-CHOP) for total of 6 cycles and is currently in complete remission on PET-CT scan. Following this treatment, the patient has been assessed and found to have complete resolution of his nasal symptoms and is in routine follow-up.

3. Discussion

Mantle cell lymphoma usually presents with lymph node enlargement. It can spread to other tissues such as bone marrow, liver and gastrointestinal tract. Cyclin D1 expression is a hallmark of MCL. This protein that promotes cell division and growth is detected by immune-histochemistry to confirm the diagnosis of MCL. Over-expression of cyclin D1 is usually caused by a translocation between chromosomes 11 and 14. Detecting this chromosomal translocation can also be used to establish the diagnosis. The classical immune-phenotypes of MCL are CD5+, CD19+, CD10-, CD23-/ weakly +, FMC7+.

Most patients with MCL present with advanced stage extranodal disease involving the bone marrow, spleen, gastrointestinal tract or circulating lymphoma cells in peripheral blood. Untreated MCL tends to progress rapidly.

Mantle cell lymphoma was considered aggressive and incurable as it typically relapses after conventional chemotherapy regimens with a median survival of three to five years. Recent clinical trials have been performed using dose-intensified immune-chemotherapy with peripheral blood stem cell transplantation to achieve a high proportion of responses and complete remission.

Treatment options for MCL also depend on the age and the general condition of the patient. Patients who are unable to tolerate aggressive treatment either because of age or of comorbidities, are treated with palliative chemotherapy of reduced intensity, usually with single agent. In conclusion, we report a rare case of an extra nodal presentation of MCL as an intranasal mass which has not been reported commonly previously in the literature. Although MCL is considered to be incurable, recent trials with intensive immune-chemotherapy have shown encouraging results.

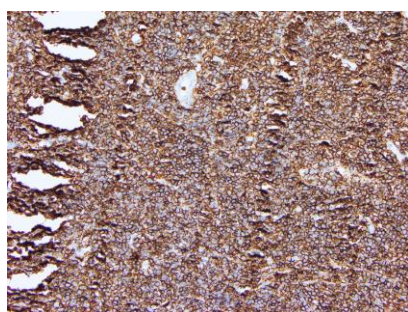


Fig. 1. CD 20

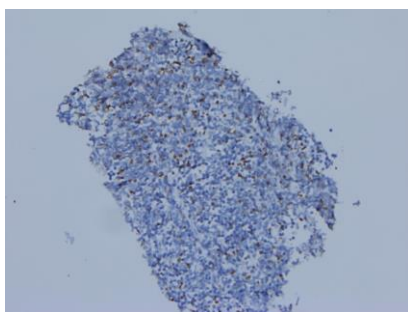


Fig. 2. CD3

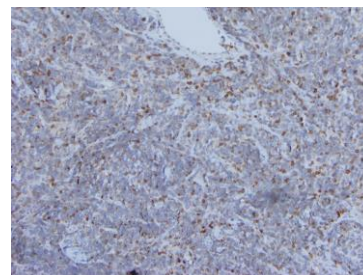


Fig. 3. CD5

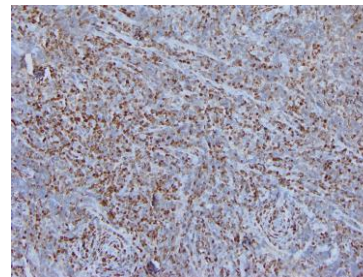


Fig. 4. Cyclin D1

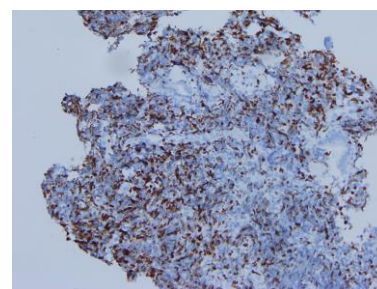


Fig. 5. Ki67

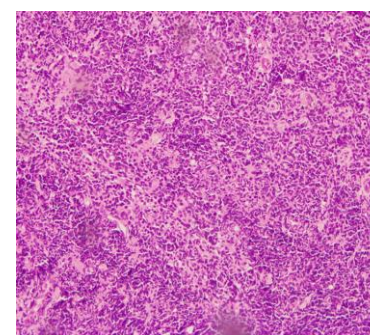


Fig. 6. H & E

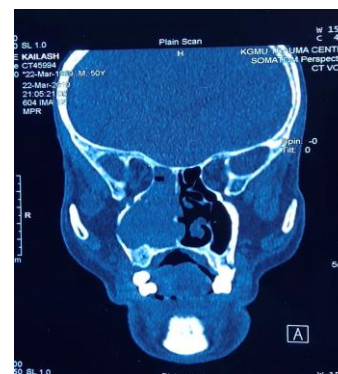


Fig. 7. Plain Scan

4. Conclusion

The clinical features of primary nasal lymphoma at presentation are nasal obstruction, unilateral and progressive epistaxis, hyposmia, and nasal swelling or mass [6]. As in this case, patients with nasal lymphoma may have experienced long-standing symptoms with initial examination failing to reveal the lymphoma. The clinical signs could be a diffuse erythematous swelling inside the nasal cavity covered with exudates and crust or in some advanced cases there may be extensive ulceration and necrosis [5].

Differential diagnosis of a sinonasal mass includes benign and malignant tumors. The common benign lesions are osteoma, chondroma, fibrous dysplasia, haemangioma, leiomyoma and schwannoma. The common malignant tumors are squamous cell carcinoma, adenocarcinoma, adenoicycstic carcinoma, esthesioneuroblastoma, sinonasal undifferentiated carcinoma, malignant melanoma, haemangiopericytoma and lymphoreticular neoplasms. Chronic granulomatous conditions like Wegner's granulomatosis, sarcoidosis, tuberculosis and

syphilis can also involve the nose and paranasal sinuses. Nasal T-cell lymphoma needs to be considered in the differential diagnosis of destructive midline sinonasal lesion.

References

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