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Case Report

Non Hodgkin Lymphoma Presenting as Unilateral Nasal Mass: A Diagnosing Challange

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Abstract

Background: NHL is comprised in a group of tumors arising from the lymphoreticular cell system. The causative factors for this condition are infective etiologies like EBV, HT cell leukemia virus *h. pylori*, HHV, hepatitis c and HIV and other causes being the chemoradiation therapy and certain chemical exposure like- herbicides etc. certain genetic disorders are also known to show association with NHL like SLE, klinefelter syndrome, ataxia etc. NHL in nose and PNS as primary lesion is a very rare entity.

Objective: The purpose of our study is to throw light on the significance of complete history, examination and significance of immunohistochemistry over CT and histopathology in patients presenting with nasal mass to diagnose such a rare entity which is easily missed.

Method: We report a case of 60 year male with right facial pain and right nasal obstruction who was diagnosed by immunohistochemistry.

Result: Computed tomography and histopathology revealed inflammatory pathology and polyp. But immunohistochemistry was consistent with non Hodgkin lymphoma- diffuse large B cell lymphoma.

Conclusion: With this study we emphasise the role of IHC as a routine investigation in all sinonasal masses for early diagnosing and managing such rare entity.

Keywords: Non-Hodgkin; Nasal Polyposis; B-Cell; Human T Cell; Paranasal Sinuses; Ebstein Bar Virus

Introduction

Hematologic malignancies are very often seen in immune compromised patients [1]. The second most common malignancy in Head and neck region is known to be Lymphomas. Among these NHL is a entity known to have its origin from the lymphoreticular cells. Of all the non hodgkins lymphoma the proportion which arises from the extranodal sites is 40%. Common primary extranodal sites of lymphomas include stomach, liver, soft tissue, dura, bone,

intestine and bone marrow. The nasal cavities and paranasal sinuses are rarely affected by primary NHL [2].

Malignant lymphoma is a neoplastic process of the lymphopoietic portion of the reticulo-endothelial system. Most of them originate from B-lymphocytes making large B cell lymphoma the most common one. Diffuse large B cell lymphoma (DLBCL) is the most common lymphoma, accounting for about 25% to 30% of all the non-Hodgkin lymphomas. It typically presents as a rapidly growing mass or enlarging lymph nodes in a nodal or extranodal site. The common locations of the same are skin, CNS, lungs, liver, intestine, dura, bone marrow, and oral cavity. Though aggressive, they do respond well to six cycles of rituximab along with cyclophosphamide, doxorubicin, vincristine, and prednisone (R-CHOP) [3].

The subtype and frequency of NHL of sinonasal tract depend on the geographical factors. One such differentiating feature between the B cell and T cell lymphoma appears to be the population in which it arises like Asian peoples are shown to suffer predominantly from Tell lymphoma whereas western population ahs B cell predominance [4]. With the help of this article, we present a case of nasal mass which appeared to be polyp but was diagnosed as lymphoma on immunohistochemistry.

Case Report

A 60 year old male patient presented to ENT opd with complaints of right sided facial pain and nasal obstruction since 1 month with no history of recurrent upper respiratory tract infection or nasal bleeding. He was on oral hypoglycemic agents for past 6 years for type 2 diabetes mellitus.

On examination left sided deviated septum with right sided polypoidal mass was visualized. On computed tomography it showed sinonasal polyposis on right side with left inferior turbinate hypertrophy and left maxillary sinusitis.

Patient was planned for functional endoscopic sinus surgery (minimally invasive surgical technique). Intraoperatively the nasal mass on right side was suspicious of being polyp and was bleeding profusely. In view of which nasal mass debulking was performed and tissue was sent for histopathology.

Figure 1: Coronal ct showing mass in right maxillary sinus and nasal cavity.

Figure 2: Axial ct showing mass in right maxillary sinus and nasal cavity.

Histopathology reports were suggestive of inflammatory sinonasal polyp with underlying lymphoproliferative disorder? Neoplastic and was advised for immunohistochemistry.

Further on IHC the mass came positive for LCA, CD20, and Bcl-2 and negative for CD 3, CD 56, cyclin D1, c-myc, panCK. Ki 67 proliferation index was 90%. Findings came to be consistent with high grade non Hodgkin lymphoma - diffuse large B cell lymphoma-non germinal centre B cell phenotype.

Post diagnosis patient was initiated on chemotherapy with 4 cycles of R-CHOP regimen. Following this treatment patient showed symptomatic and clinical improvement.

Discussion

Of all the malignancies NHL of nose and paranasal sinus contribute to 3-5%. of which the most common histoligical variant was B cell lymphoma. Multiple studies states that the histological type is not clear, with some showing B cell lesions common [5], whereas others documented higher frequency of T-cell lesions [6,7]. whereas Hatta., *et al*. [8] mentioned in his study that in Japan the most common variant was angiocentric lymphoma (35.9%) next being the B cell Lymphoma (22.6%), T cell lymphoma (15.1%) followed by other lymphomas and the non specific types [8].

Parnasala sinuses predominantly shows B cell Lymphoma. Whereas nasal cavity shows tumor cells predominantly with positive T-cell markers. Children are most commonly diagnosed with extra nodal NHL thus an early diagnosis of sinonasal masses is mandatory for prompt management [9,10].

The maxillar sinus was seen to more commonly involved site that nasal cavity in western population where as opposite is seen in asial patients with nasal cavity being the common site of origin [11,12].

Macroscopically the differentiating feature between lymphomas and squamous cell carcinoma being that the former are submucosal and later ulcerative. In our case patient just presented with right sided nasal mass with complain of facial pain and it came as inflammatory polyp or sinusitis in computed tomography and histopathology. The lesion was diagnosed on immunohistochemistry.

Patient suffering from sinonasal lymphomas are shown to have symptoms like nasal blockage, nasal bleeding, headache, swelling over face, cheek and nose [13,14]. Other less frequent symptoms include blurred or double vision, pain over cheek and nose.

The B cell lymphomas result from the malignant proliferation of B cells during their various stages of development. Depending on the morphology, genetics, and immunophenotype of the neoplastic cells, a cell of origin (COO) is proposed. The development of B cells can be categorized into 3 stages- pre-germinal, germinal, and post-germinal center. Most of the B-cell lymphomas are derived from the germinal center.

DLBCL commonly presents with enlarged lymph nodes or rapidly growing mass along with B symptoms, which include fever, night sweats, and weight loss. The B symptoms can be seen in 30% of the patients. Bone marrow involvement is more common in indolent disease and can be seen in up to 50% of the cases [15].

About 50% of the patients have extranodal involvement. The involvement of the stomach or gastrointestinal tract is the most common primary extranodal site, followed by the involvement of the skin. Renal involvement can be secondary to bulky disease or lymphadenopathy, causing a ureteral obstruction or due to tumor lysis syndrome. Extralymphatic involvement is seen in aggressive NHL [16].

Differential diagnosis for this include infectious and granulomatous diseases. Thus a elaborate examination and proper investigation is required for prompt diagnosis and early management.

The treatment of B cell lymphomas depends not only on their staging but also on the type of the disease (indolent or aggressive) and the molecular subtype. Though DLBCL is aggressive, with appropriate chemotherapy, the survival can be long, but with a limited cure rate. Patients with GCB DLBCL respond well to 6 cycles of rituximab along with cyclophosphamide, doxorubicin, vincristine, and prednisone (R-CHOP) regimen given every 21 days.

Conclusion

Due to the symptom overlap between many nasal pathologies and sinonasal Non Hodgkin lymphoma and because it is a rare condition it becomes difficult and challenge in diagnosing this condition. Though it is difficult to diagnose and aggressive in nature but once diagnosed localized diseases are responsive to combination chemotherapy. We emphasise the role of immunohistochemistry as a routine as even HPE can be misdiagnosing.

Further such studies are required to conclude that inclusion of immunohistochemistry in all cases of sinonasal masses aid in diagnosing the case early.

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