A RARE CASE OF NASAL B CELL LYMPHOMA
A CASE REPORT

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ABSTRACT

Lymphomas arising in nasal cavity are relatively uncommon. Their rarity makes them dismissible compared to benign inflammatory lesions. Therefore while evaluating a nasal mass, clinician should keep nasal lymphoma in mind as it influences management and prognosis. A male patient presented with right nasal obstruction, hyposmia and one episode of epistaxis. Examination revealed a solitary reddish mass filling the right nasal cavity. Computed Tomography scan and Histopathological examination were both inconclusive in a diagnosis. Immunohistochemistry revealed Non Hodgkin’s lymphoma- B cell neoplasm [Extraosseous Plasmacytoma]

Keywords: Nasal mass, Non Hodgkin’s lymphoma, Immunohistochemistry.

INTRODUCTION:

Primary Non-Hodgkin Lymphomas of the nasal cavity or paranasal sinuses are defined as lymphoid cell neoplasms in which the bulk of the disease occurs in these anatomic sites. Extramedullary plasmacytoma is a mass forming lesion of monoclonal plasma cells that occurs outside the bone and bone marrow. Carcinomas of nasal cavity and paranasal sinuses account for 0.2-0.8 % of all malignant neoplasms. Malignant lymphomas account for 14% of all cancers in the nose and paranasal sinuses. They account for 2.6 to 6.7 % of all lymphomas. Non-Hodgkin lymphomas broadly comprise of NK/T or T cell lymphomas and B cell lymphomas. In Asians 29% of all primary nasal cavity lymphomas are of B cell type. Extramedullary plasmacytoma is a part of Non-Hodgkin’s Lymphoma (B cell type) and can be classified into well, moderately or poorly differentiated.

CASE REPORT:

A patient aged 31 years, a farmer by occupation presented with complaints of right sided nasal obstruction since 2 months which was insidious in onset and gradually progressive. He also complained of difficulty in breathing since 2 months which was gradually progressive and a reduced sense of smell since 2 months which was associated with watery nasal discharge. Episodes of headache, heaviness of head were present on and off since 2 months. He also gives history of 1 episode of bleeding from the nose 1 month back.

Clinical Examination showed a solitary mass filling the right nasal cavity and partially obscuring the view of inferior turbinate, reddish and was bleeding on touch. The mass could be probed medially and along the floor and the roof of the nasal cavity. Cold spatula test showed decreased fogging on the right side. Olfaction was reduced on the right side. There were no palpable cervical lymph nodes.

Full blood count, Peripheral smear, renal and liver function tests done were normal. Endoscopic examination of the nasal cavity was suggestive of a mass arising from the posterior end of the middle turbinate filling the nasal cavity. Computed Tomography scan of the Nose and Paranasal Sinus showed a mass in the right nasal cavity without any intracranial extension abutting the septum and middle turbinate suggestive of an exophytic papilloma. [Fig. 2]
Nasal Mass was excised under Endoscopic guidance and sent for histopathological examination and nasal cavity was packed to control post-operative epistaxis. Histopathological Examination showed metaplastic squamous epithelium overlying diffusely distributed single cells and nests of pleomorphic cells with nuclear infoldings, many binucleate and multinucleate cells. Diffuse sprinkling of plasma cells and lymphocytes were also seen. Few bits showed columnar epithelium with seromucinous glands in subepithelium. These features suggested possibility of a poorly differentiated carcinoma (lymphoepithelial type)/ Non Hodgkin Lymphoma.

These photomicrographs depict a diffusely arranged group of pleomorphic cells with enlarged nucleus and moderate amount of cytoplasm. Some cells exhibit prominent nucleolus. Few binucleate forms are seen. Nasal mucosa is also noted. [Fig 3][Fig 4] Immunohistochemistry staining showed cells positive for CD 138 [Fig 5] and CD 45 and negative for CD 20 and Cytokeratin. There was patchy positivity for CD 19 in the tumor cells.

Immunohistochemistry confirmed a diagnosis of B cell Non Hodgkin Lymphoma- Extra Osseous Plasmacytoma involving the nasal cavity.

Patient had been relieved of nasal obstruction and his other symptoms. The post operative period was uneventful. Patient has been planned for further chemo and radiotherapy as per the guidelines stated by national cancer institute.
DISCUSSION:

Non Hodgkin Lymphomas of the nasal cavity or paranasal sinuses are haematolymphoid tumours with the bulk of the disease in these sites. Non Hodgkins Lymphoma of the sinonasal tract are uncommon malignancies representing 3 to 5% of all malignancies and accounts for 60% of all lymphomas.

Extramedullary plasmacytomas (EMPs) are a mass forming lesion of monoclonal plasma cells that occurs outside the bone and bone marrow. They are seen in the age group of 34-78 years with a male predominance of 4:1. In the Asian population 90% of sinonasal lymphomas are T cell in origin unlike in this case. The nasal cavity is the most common site of occurrence for head and neck Extramedullary Plasmacytomas (28%) as was seen in this patient. In Asian patients the nasal cavity is more common as a primary site for lymphomas compared to the paranasal sinus unlike in the Western population. Presenting features are soft tissue mass (80%), airway obstruction (35%) as in this case, epistaxis (35%), local pain (20%), proptosis (15%), nasal discharge (10%), regional lymphadenopathy (10%)1.

The investigations of choice are usually Computed Tomography scan of nose and Paranasal Sinus to check extent of lesion and bony erosions. Biopsy of lesion and Immunohistochemistry can confirm the diagnosis of Lymphoma.

Radiographically these tumors mimic carcinomas and other neoplastic conditions which are more prevalent, therefore the surgeon will have a low suspicion of lymphoma. Even the gross appearance is misleading. They can be smooth, lobulated or nodular with a fleshy or rubbery consistency2.

Superficial sampling of lesion may not diagnose lymphoma as they are subepithelial and more deeper biopsies are required. Angiocentricty and angioinvasion are features which can be detected in deeper biopsies2. Sometimes due to ischaemia and necrosis initial biopsy may be negative requiring immunophenotyping4.

Histopathology shows a diffuse infiltrate of neoplastic plasma cells in the subepithelial tissue, accompanied by a scant vascularized stroma. The tumour can be well, moderately or poorly differentiated. Atypical plasma cells are usually seen which can be recognized by their eccentrically placed nuclei, coarsely clumped “clock-face” chromatin in some nuclei, and amphophillic cytoplasm with a paranuclear (Golgi zone)1. In this case pleomorphic cells along with variably sized nuclei were seen along with a diffuse sprinkling of plasma cells.

Moderately and Poorly differentiated EMPs cause significant difficulties in differentiating from large cell lymphoma, carcinoma, melanoma and olfactory neuroblastoma. These reasons initially produced the diagnosis of a lymphoepithelial carcinoma (poorly differentiated) in this case. Immunohistochemistry staining becomes essential to come to a definitive diagnosis. Poorly differentiated EMPs comprise large cells that are often barely recognizable as being plasma cells.

Immunohistochemistry of extramedullary plasmacytoma shows CD 20 negative and CD 138 positive as in this case. There is also usually expression of CD 38, VS38 but they are not specific for plasma cells. PAX-5 is negative, while Oct-2 and Bob.1 are frequently positive. Plasma cell neoplasms show positivity in CD 45 staining. These are proteins detected by immunohistochemical staining. Unlike in this case Cytokeratin positivity may be present which can lead to a misdiagnosis of carcinoma.

In terms of differential diagnosis Epstein Barr virus (EBV) studies can be very helpful. Nasal lymphomas of B cell origin have only a weak association with Epstein-Barr virus. Lymphomatoid granulomatosis is EBV positive, it is a B cell lymphoproliferative disease5.

After confirming the diagnosis by immunohistochemistry the treatment of choice is Chemoradiotherapy sometimes with multiple chemotherapeutic agents or Radiotherapy alone depending on the stage of lesion [1][6]. Majority of the patients with Nasal Lymphoma (Non Hodgkin’s type) present at an early stage with localized disease without any metastasis. Approximately 20% of patients with primary Extramedullary Plasmacytoma will develop Multiple Myeloma but it is not possible to predict which cases will progress1.

Unlike NK/T cell lymphoma or Diffuse large B-cell lymphoma (DLBCL), extramedullary plasmacytoma has a better prognosis. Unlike DLBCL which has a poor prognosis when associated with lymphatic metastasis7, this tumor of B cell origin responds well to treatment.
Several other differential diagnosis should be kept in mind while evaluating a nasal mass such as Carcinoma-squamous cell carcinoma, lymphoepithelial type, Melanoma, Papilloma, Olfactory neuroblastoma, Rhinoscleroma and Wegener’s granulomatosis.

CONCLUSION:

Lymphomas arising in the nasal cavity are relatively uncommon. Incidence of cancer of the nasal cavity and Paranasal sinuses is low (<1.5/100,000) of which 14% are lymphomas. This rarity causes clinicians to dismiss them as a benign inflammatory infiltrate. Furthermore routine blood investigations may be normal and radiographic images may be misleading. The importance of immunohistochemistry in the diagnosis of lymphomas cannot be overstated especially owing to its rarity. Therefore, while evaluating a nasal mass, a clinician should keep nasal lymphoma in mind as it influences management and prognosis.

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