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NON HODGKIN LYMPHOMA PRESENTING AS UNILATERAL NASAL MASS: A CASE REPORT
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ABSTRACT: Non-Hodgkin’s Lymphoma (NHL) is a group of neoplasms that originate from the cells of the lymphoreticular system. Forty percent of Non-Hodgkin's lymphoma arises from extranodal sites. Common primary extranodal sites of lymphomas include stomach, liver, soft tissue, dura, bone, intestine and bone marrow. Primary non-Hodgkin's lymphoma of the nose and paranasal sinus is rare. We report a case of a young 22 years aged man who came to us with symptoms of nasal obstruction and swelling over right side of face with subsequent development of neck swelling. We diagnosed the lymphoma by biopsy and FNAC of the submandibular lymph node. This case highlights the importance of making a full visual inspection of the involved sinus in order to avoid missing an unexpected, albeit a rarely encountered, pathology.

KEYWORDS: Non-Hodgkin, Lymphoma, Nasal polyp, B-Cell.

INTRODUCTION: Lymphomas are malignant neoplasm of lymphoreticular cells. Hematologic malignancies are very often seen in immune compromised patients. Large B-cell lymphoma (LBCL) is the most common non-Hodgkin’s lymphoma. Large B-cell lymphoma is a fast growing malignancy that may arise inside or outside of the lymphatic system. Skin, abdomen, lung, central nervous system, and oral cavity are common locations. Nasal Large B-cell lymphoma is a rare presentation.

The etiology is unknown but it is thought to be due to virus and immunosuppression. Primary non-Hodgkin lymphomas (NHLs) of the sinonasal tract comprise a rare entity that constitutes 1.5% of all NHLs and 2.2% of extranodal lymphomas. Clinical diagnosis may be very challenging because of the variety and low specificity of the presenting symptoms and signs. In this article, we report a case of lymphoma that was diagnosed after we used an endoscopic procedure.

CASE REPORT: A 22-year man came to the otolaryngology department with a one month history of right nasal cavity obstruction and right sided face swelling, which had spread across the right maxillary region and right neck region in last seven days. He also complained of headaches, and intermittent but brisk epistaxis. He had noticed a gradual diminution of his sense of smell. Initially there was no lymphadenopathy later patient developed right submandibular lymphadenopathy.

There was no limitation of eye movements or visual acuity. He was afebrile. Further systemic examination was unremarkable and his Hb was 11.8 gm%, total count of 12500 cells/cumm, platelet count 180000 cells/cumm, ESR 40 mm/hr, serum creatinine 1.3 mg/dl, blood urea 43 mg/dl was HIV negative. His USG abdomen and bilateral inguinal region shows mild splenomegaly, mild hepatomegaly, both kidneys were grossly enlarged with lymphomatous invasion (right kidney-15.8 cm, left kidney-15.3 cm).No inguinal lymphadenopathy was seen. His FNAC from submandibular lymph node were suggestive of non-Hodgkin’s lymphoma (diffuse large cell type). CT scan face, neck with thorax shows large soft tissue density space occupying mass lesion in right side of face and para
nasal sinuses with extension into cutaneous and subcutaneous zones in nasal and paranasal regions, dehiscence and erosions of bony outlines and extensions into right medial orbit. Right carotid space deposits at level 1 and 2 are seen with normal thoracic study.

H.P.E study was suggestive of round cell tumor (non Hodgkin’s lymphoma). We made the final diagnosis on the basis of histopathological examination and FNAC. Before starting chemotherapy bone marrow examination of the patient was done which revealed marrow infiltration by atypical lymphoid cells 80%), following which patient was send for chemotherapy. Patient was started on inj vincristine, inj adriamycin, inj cyclophosphamide, tab prednisolone. Four cycles of these have been given to patient till now. Over the next 3 months, the swelling resolved and the patient’s symptoms improved.

**DISCUSSION:** NHLs of the sinonasal tract are uncommon malignancies representing 3% to 5% of all malignancies. Most frequent histological type was B-cell lymphoma. Various studies says, predominance of the histological type is unclear, with some finding B-cell lesions more common, whereas others noted a greater frequency of T-cell lesions. On the contrary, according to Hatta et al, the most common histological type, in Japan, is angiocentric lymphoma (35.9%), followed by B-cell lymphoma (22.6%), peripheral T-cell lymphoma types (15.1%), and other lymphomas and non-specific types.

B-cell lymphoma is predominant in paranasal sinuses. Tumor cells with positive T-cell markers (angiocentric lymphoma and peripheral T-cell lymphoma) are predominant in nasal cavities. Children present with a higher proportion of extra-nodal NHL of the head and neck, and an early diagnosis of NHL of nasal cavity and paranasal sinuses is necessary for adequate and successful management.

In Western populations, lymphomas of the maxillary sinus are more common than in the nasal cavity. On the contrary, in Asian patients the nasal cavity is more common as a primary site than the maxillary sinus.

Lymphomas are usually sub-mucosal and on gross appearance differ from squamous cell carcinomas, which are usually ulcerative. Early diagnosis of primary NHL of the nasal cavity and paranasal sinuses was difficult in our patients, because this lesion develops in an anatomic space and expands toward the sinus, nasal cavity or nasopharynx, not usually causing symptoms in the early stages. Only after reaching a considerable size and involvement do the symptoms appear, and these may simulate other nasal or head and neck diseases. The most common presenting symptoms of sinonasal lymphomas are nasal obstruction, epistaxis, headache, and unilateral facial, cheek, or nasal swelling. Other infrequent symptoms are diplopia or blurred vision, and nasal or cheek pain.

This latter group of patients has a younger median age (i.e., 37 years) and a female predominance (66%). Subtypes of diffuse large B cell lymphoma, including those with an immunoblastic subtype and tumors with extensive fibrosis, are recognized by pathologists but do not appear to have important independent prognostic significance.

Diffuse large B cell lymphoma can present as either primary lymph node disease or at extranodal sites. Primary non-Hodgkin lymphomas (NHLs) of the sinonasal tract comprise a rare entity that constitutes 1.5% of all NHLs and 2.2% of extranodal lymphomas. Essentially any organ can be involved, making a diagnostic biopsy imperative.

The clinical presentation of these lesions can be confused with those of infectious, granulomatous, and nonlymphomatous neoplastic processes. The limited clinical experience with this
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lesion has led to controversy regarding its pathologic classification, natural history, and optimal management.

Most lesions are locally advanced and involve multiple anatomic structures. This makes the assignment of a primary site somewhat arbitrary in cases of large tumors.

Our patient's clinical picture suggested obstruction of nasal cavity with episodes of epistaxis and facial and neck swelling suggestive of nasal mass extending to paranasal sinuses involving orbit with regional lymphadenopathy which was later on confirmed on nasal examination which revealed a mass in the right nasal cavity extending to right paranasal sinuses with involvement of orbit and regional submandibular lymph nodes involvement. FNAC from submandibular lymph node, CT of face, neck and thorax and biopsy of nasal mass was done.

CT scan face neck with thorax shows large soft tissue density space occupying mass lesion in right side of face and para nasal sinuses with extension into cutaneous and subcutaneous zones in nasal and paranasal regions, dehiscence and erosions of bony outlines and extensions into right medial orbit. Right carotid space deposits at level 1 and 2 are seen with normal thoracic study. HPE study was suggestive of round cell tumor (? non Hodgkin’s lymphoma/alveolar rhabdomyosarcoma/olfactory neuroblastoma).

The initial treatment of all patients with diffuse large B cell lymphoma should be with a combination chemotherapy regimen. The most popular regimen in the is CHOP plus rituximab.

CONCLUSION: Non hodgkin’s lymphoma (diffuse large cell) nose and paranasal sinus is a rare entity and more than 50% of patients will have site of extranodal involvement as gastrointestinal tract and bone marrow. It has a younger median age (i.e., 37 years) and a female predominance (66%) but in over case male is involved with an age of 22 which is another rare entity.

Treatment of all patients with diffuse large B cell lymphoma should be with a combination chemotherapy regimen. The most popular regimen is CHOP plus rituximab but in over case we used vincristine, Adriamycin, Cyclophosphamide and prednisolone as alternate regimen with equal efficacy.

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