NON HODGKIN'S LYMPHOMA OF RIGHT TONSIL: A RARE CASE STUDY

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ABSTRACT: Non-Hodgkin's lymphoma is not one disease but many. It embraces a spectrum from low grade; indolent disease with an untreated natural history which may be measured in decades through to aggressive types which, if untreated may prove fatal within days of presentation. NHL as a group is more common than Hodgkin's disease, but many of the subtypes are relatively rare. Usually, NHL is a disease of the lymphnodes, but extra nodal NHL is significantly more common than extra nodal Hodgkin's disease. NHL can present at virtually any age, but different types are more common at different ages. In this case study we discuss a relatively rare presentation of a Non-Hodgkin's lymphoma presenting as grade III tonsillar hypertrophy.

KEYWORDS: Non-Hodgkin's Lymphoma, lymph node.

INTRODUCTION: Non-Hodgkin's lymphoma is one of the commonest malignancies detected in the lymphoid tissues. Most of these patients present with undiagnosed cervical lymphadenopathy. These patients should be investigated as any other patient who presents with a lump in the neck. The history may give useful clues and the examination should include not only the neck but the draining cutaneous sites and the upper aero digestive tract, and other superficial lymph node sites and the abdomen. Fine needle aspiration cytology may point towards a lymphoma, but formal lymph node biopsy will usually be required to yield a definitive diagnosis. Routine blood tests and a chest X-ray to evaluate the mediastinum should be performed prior to biopsy, but CT scans, bone marrow and any other staging investigations may usually be deferred until a definitive histopathological diagnosis has been made. Other rare presentations other than lymph node enlargement are also seen ad in a nasopharyngeal tumour blocking the Eustachian tube in a child presenting with otalgia and deafness, or a rapidly enlarging goiter in an old woman may be due to the development of a high grade thyroid lymphoma, or nasal obstruction and discharge may prove to be due to plasmacytoma.

CASE STUDY: A 30 year old male patient presented to the OPD complaining of difficulty in deglutition since a year. No complaints of pain or constitutional symptoms were given. On examination a severe grade III hypertrophy of the right tonsil was seen. The only other significant presenting finding was diffuse axillary lymphadenopathy as well as inguinal lymphadenopathy. A tentative diagnosis of lymphoma was made and the patient was sent to the pathologist for the FNAC of the tonsil, the axillary and the inguinal lymph nodes. The reports were intentionally sent to two different pathologists to cross verify the findings. One of the reports revealed a suspicion of lymphoma but advised a tissue biopsy for confirming the same. So

a decision was made to do the Tonsillar resection of both the sides and the tissue sent again for histopathology with one specimen having the diseased tonsil and the other a normal appearing tonsil tissue. Routine investigations were also done including X-rays of the chest, Ultrasound abdomen, CT scan of the neck. The excisional biopsy report revealed the lesion as Non-Hodgkin's lymphoma and paraffin blocks were retrieved for immune histochemistry. The lesion was identified as a low grade B cell lymphoma and the patient was referred to the oncologist for further management.







DISCUSSION: Non-Hodgkin's lymphoma can present as a unilateral tonsillar enlargement. A strong element of suspicion along with proper investigations would help in the diagnosis of the pathology namely the Fine needle aspiration cytology of the involved group of lymph nodes or the excisional biopsy of the node along with Immunohistocytochemistry to further confirm the type and grade of the lymphoma. Ultrasound abdomen and CT scans also help in identifying the involved site. Once a patient with NHL has had their tumour characterized, classified and staged, it is possible to think about what treatment may be appropriate. As with any other patient with head and neck cancer, it is necessary also to take into account the patients age, performance status, comorbidity, lifestyle factors, social support and personal wishes, before deciding on treatment.

Despite the profusion of classification systems with counterintuitive nomenclature, NHL can for practical clinical purposes largely be divided in just two groups: high-grade NHL and low-grade NHL. The first clinically relevant pathological classification in modern times was the Rappaport classification (1966). This pre-dated immunocytochemistry and the recognition of the T-cell and B-cell lineages. In Britain and Europe the updated Kiel classification became more widely used (broadly classifies the tumour into B-cell lymphomas and T-cell lymphomas, which are further divided into low grade and high grade and rare varieties). The working formulation for clinical use (1982) represented an American attempt at a practical classification and was widely adopted in the US despite the flaws, that it ignored the T-cell and B-cell immunophenotypes (namely the B-cell neoplasms and T-cell and postulated natural killer (NK) cell neoplasms). More recently, attempts have been made to draw together the American (working formulation) and European(Kiel classification) systems of Nomenclature. The revised European-American

lymphoma classification (the REAL classification) was published in 1995 and it is hoped that it will become the new standard. Whichever classification is used by each pathology department, the simplistic division of NHL into high grade and low grade subsets remains for the most part entirely valid.

Staging of the NHL is done in the same way as the Hodgkin's disease, using the Ann Arbor classification (stages 1-4A/B). In children, extranodal disease is more common and an alternative staging classification, the Murphy staging system is used. This is a shybrid system which takes in account location, surgical treatment and spread into non-lymphoid structures.

Most cases diagnosed with NHL would further be referred to the oncologist for expert and Chemotherapy which includes Radiotherapy depending management immunohistochemistry report. Treatment options for low grade disease if the disease is extensive with bone marrow involvement must be considered incurable. The disease may not require specific treatment following diagnosis, unless or until symptoms require palliation. Simple chemotherapy with oral chlorambucil and prednisolone may be all that is required to make the patient symptom free and achieve la long remission. At relapse, if symptoms dictate, then the same treatment can be repeated. Low dose palliative radiotherapy is often effective at shrinking tumour masses and relieving symptoms. Eventually the remission duration may shorten, the disease may become refractory to treatment or it may transform into a high grade NHL. High grade NHL despite significant early mortality from progressive refractory disease or from treatment toxicity is often curable. The standard chemotherapy regime CHOP comprises of cyclophosphamide, doxorubicin, vincristine and prednisolone. For patients with localized disease especially in head and neck, this is often followed by consolidation radiotherapy. Radiotherapy can provide valuable palliation with minimal toxicity in patients who have relapsed or who are too frail for initial chemotherapy.

This case of unilateral hypertrophy of the tonsil in a relatively young individual who apparently had no significant complaints would often go unnoticed as a strong degree of suspicion is required and treatment options planned well in advance to manage it. On follow up after a month of treatment with oral chlorambucil and prednisolone, patient was tolerating the treatment well and showed signs of improvement as in resolution of the lymphadenopathy in the other sites as in axillary and inguinal to a certain degree.

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