Malignant Non-Hodgkin's Lymphomas of the Nose and Paranasal Sinuses

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Summary: Malignant non-Hodgkin's Lymphoma of the nose and paranasal sinuses are rare. The clinical and histopathological findings in seven cases that presented to the Dept. of Otolaryngology, UKM are reviewed.

Introduction

Malignant lymphomas of the nose and paranasal sinuses are rare.[1] Involvement of this site only constitutes 1.7% of all lymphomas[2] and less than 6% of lymphomas affecting the head and neck.[3]

They mimic carcinomas in this site in their clinical and radiological presentation and may pose problems in diagnosis even on histological examination.[4]

In a three year period from September 1983 to July 1986, seven cases of malignant non-Hodgkin's lumphomas of the nose and paranasal sinuses were diagnosed and treated in the Department of Otolaryngology, UKM. In this report we present the clinical and pathological features of this uncommon disease.

Material and Methods

The clinical and pathological findings of seven consecutive cases of malignant non-Hodgkin's lumphomas of the nose and paranasal sinuses that presented to the Dept. of Otolaryngology, University Kebangsaan Malaysia over a three year period from Sept. 1983 to July 1986 were reviewed.

Clinical data including age, sex, symptomatology, physical findings, radiological findings and treatment were obtained from the records. The histopathological slides were reviewed and the lymphomas were classified according to the Rappaport Classification.

After an otolaryngologic examination, the patients were investigated to determine the extent of the disease. These included full blood count, a search for hepatosplenomegaly, bone marrow examination, plain radiography and computerised tomography (CT) of the paranasal sinuses. Examination of the nose including the nasopharynx and biopsy of the nasal tumours under general anaesthesia were performed for all cases.

Symptoms	No. of Cases
Nasal Obstruction	7 (100%)
Epistaxis	6 (85.7%)
Rhinorrhoea	1 (14.3%)
Diplopia	4 (57.1%)
Epiphora	1 (14.3%)
Facial / Cheek swelling	5 (71.4%)
Frontal Headache	1 (14.3%)
Neck Swelling	1 (14.3%)

Table 1:
Presenting Symptoms.

Clinical Signs	No. of Cases
Mass in Nose	7 (100%)
Mass in Post-Natal Space	4 (57.1%)
Diplopia	4 (57.1%)
Proptosis	3 (42.9%)
Loss of Vision	1 (14.3%)
Cervical Lymphadenopathy	2 (28.6%)

Table 2:
Clinical Features

Results

Clinical Findings

There were four male and three female patients and their ages ranged from 16 to 67 years. The average age was 38.7 years. Four (57%) patients were in the 20 to 40 years range. There were six Malays and one Orang Asli.

Tables 1 and 2 list the presenting symptoms and significant clinical features. The duration of symptoms was from one to three months with an average of 1.75 months. The predominant symptom was unilateral nasal obstruction with recurrent intermittent epistaxis. Unilateral facial swelling was a complaint in five (71.4%) patients. Four (57%) patients complained of diplopia. Constitutional symptoms, in particular fever and night sweats, were not encountered at all.

Unilateral facial swelling involving the nose and adjacent malar region was found in five (71.4%) patients. Proptosis was found in three (42.9%) patients and all three were initially seen in the Dept. of Ophthalmology. In one, the chemosis was very severe (Figure 1) while in another there was diminished vision in the affected eye (Figure 2). All three had varying degrees of diplopia because of the restriction of eye movements.

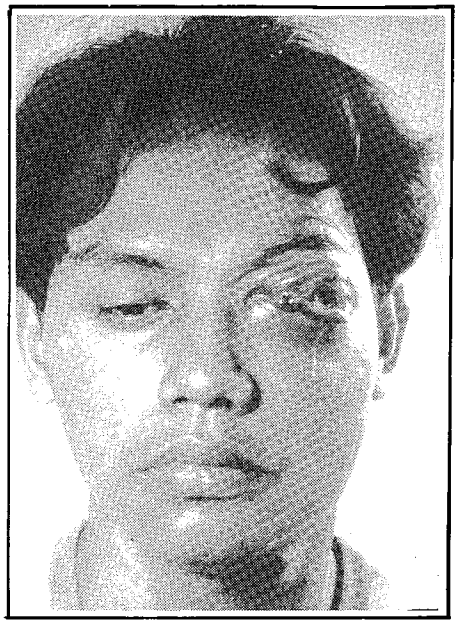

Figure 1:
Patient with severe proptosis and chemosis of the left eye as well as swelling of the left cheek.

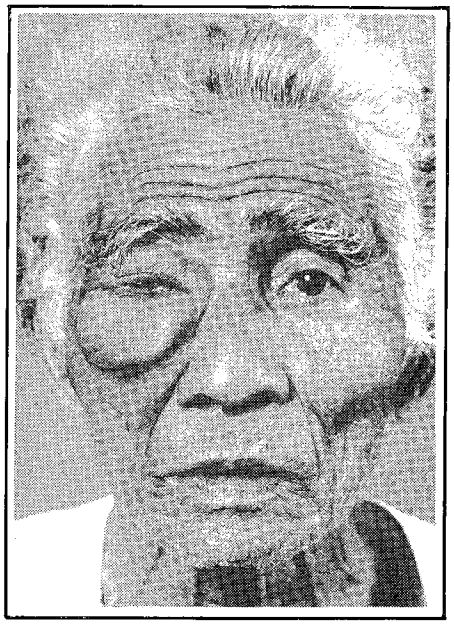

Figure 2:
Patient with peri-orbital swelling and loss of vision of the right eye.

A nasal mass was a finding in all patients. In all of them, the nasal passages on the affected side were occluded by the tumour. The gross appearance of the tumours varied considerably. Most of them were fleshy and varied in colour between red and grey. In four patients the tumours extended to the nasopharynx.

Cervical lymphadenopathy was found in two patients. In one the lymph nodes in both posterior triangles of the neck were enlarged. He also had enlarged nodes in the right axilla and both inguinal regions. In the other the lymph nodes of the deep cervical chain on one side of the neck was enlarged.

Radiological Features

The chest radiographs were all normal. Plain radiographs of the paranasal sinuses were available in four patients. Opacification of the nose and the ipsilateral maxillary sinues was the feature in all four cases. Destruction of the medial wall of the maxillary sinus was found in two cases and opacification of the ethmoidal sinuses was found in one patient.

CT scans were performed in the four patients seen in 1985 and 1986. There was tumour involvement of the nose and maxillary sinuses together with destruction of the medial wall of the maxillary sinus in all patients (Figure 3). There was destruction of the floor of the nose in one patient and erosion of the nasal septum in another. The tumour extended to the ethmoidal sinuses in three patients (Figure 4), while the frontal sinuses were opacified in two others, probably as a result of mucosal reaction. The floor of the orbit was eroded in two patients and the medical wall in one.

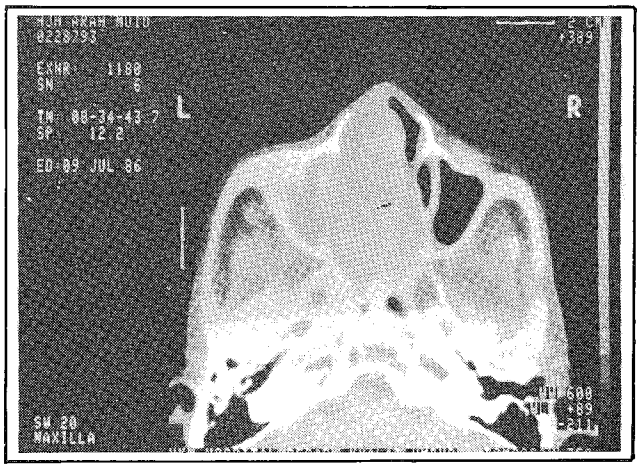

Figure 3:
Computerized tomography of the nose and para-nasal sinuses showing tumour filling the left nasal passage and extending to the nasopharynx. The nasal septum is eroded and the right nasal passage is also obstructed by the tumour. The medical wall of the left maxillary antrum is eroded and the antrum is filled with tumour.

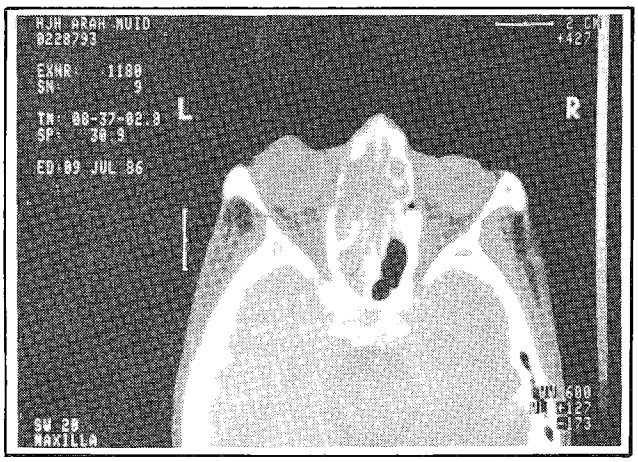

Figure 4:
Computerized tomography of same patient is in Figure 3. The ethmoidal sinuses are filled with tumour and there is proptosis of the left eye.

Carotid angiograms were performed in two cases which were diagnosed clinically as angiofibromas. In one patient, the nasopharyngeal mass was seen to be receiving blood supply from both maxillary arteries. In the other the carotid angiogram was normal.

Pathological Features

All seven patients had non-Hodgkin's lymphomas with a diffuse pattern. Three patients had 'histiocytic' (large cell) lymphomas and two had mixed lymphocytic 'histiocytic (small and large cell) lesions. One patient had a poorly differentiated lymphocytic lymphoma. The biopsy received from the last patient was too minute to accurately subtype the lesion.

Treatment

All the patients received 4000-4500 rads of radiotherapy. In six patients, the response was dramatic with relief of local symptoms within two to three weeks of commencement of treatment. In one patient, chemotherapy was employed when there was no response to radiotherapy. Cyclophosphamide, Vincristine and Prednisolone was the regime employed. She unfortunately defaulted after only one course of treatment.

The duration of follow-up was short, ranging from one to three months only. There was no recurrence of local symptoms during this period of review. However, long term follow-up proved impossible as the patients defaulted.

Discussion

Patients with malignant lymphoma generally present with primary nodal involvement.[5] Extranodal disease generally represents early contiguous extension from the lymphatic site or disseminated spread to the extra lymphatic tissue or organ. In 25% of patients, however, the primary site may be the extranodal lymphatic tissue of Waldeyer's ring or gastrointestinal tract, as well as nonlymphatic organs or tissue.[6,7]

The changing nomenclature and histologic classifications of malignant non-Hodgkin's lymphoma are confusing. The commonly used Rappaport's classification is based on the cytology and the pattern of proliferation of the malignant cells affecting the involved lymph nodes.[8] The large cells are described as histiocystic and the small ones lymphocytic. A more favourable prognosis is associated with the nodular pattern and lymphocytic cell type. New classifications using immunological techniques have been proposed[9] but these were not available to us. The histopathological features in our patients reflect the nature of non-Hodgkin's in this country in which almost all such lymphomas are of diffuse pattern and of intermediate or high grade malignancy.

The clinical and radiological findings of patients with lymphomas in the nose and paranasal sinuses in the present study are comparable to those in other reports[5,10,11] with the exception that the patients in this study are younger than those in the other reports. The peak incidence in the other reports were in the 6th and 7th decades. In this study, most of the patients (50%) were in the 3rd and 4th decades of life. Except for one Orang Asli patient, all the rest (85.75%) were Malay patients. This apparent racial predisposition may be significant and warrants further study and confirmation.

The maxillary sinus is the most frequent site of involvement, often with bone destruction and contiguous spread to adjacent tissues. The symptoms are those of a rapidly enlarging mass in the nasal cavity and/or maxillary sinus. These symptoms and physical findings are similar to those of carcinoma in this site and are difficult to differentiate even on histopathological examination. Monoclonal antibodies to cell markers have proved very useful in this respect and can often differentiate carcinoma from lymphoma.

Evaluation of the extent of regional disease is of considerable importance as it affects the planning of treatment. The nose and paranasal sinuses are difficult to examine clinically. The introduction of computerised tomography (CT) has made the task of determining the extent of disease easier. CT scanning should be a routine investigation if available. When cervical lymphadenopathy is present, disease in other sites should be suspected and investigated for CT which is also useful for this purpose although lymphangiography and laparotomy may have to be carried out.

The primary modality of treatment of this disease is radiotherapy. Surgery is limited to the obtaining of material for histological diagnosis, evaluating the extent of disease and providing adequate drainage for the maxillary sinus during the stage of tumour necrosis. Radiotherapy is usually effective in controlling head and neck lymphoma in the primary site when doses of at least 4000 rads are administered.[1 2]

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