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Nasal Extranodal NK/T Cell Lymphoma: A Case Report

Medical Science

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ABSTRACT *Extranodal NK/T-cell lymphoma, nasal type (ENKL) is a rare lymphoid neoplasm, characterized by a destructive process of the upper respiratory tract that has an unusual and rapid evolution. Despite the malignant clinical course, histological diagnosis can be difficult because of extensive tissue necrosis and multiple biopsies that are often required. We report a case of Nasal Extranodal Nk/T Cell Lymphoma, also known as angiocentric T-cell lymphoma.*

INTRODUCTION

Lymphomas account for 3-5% of all malignant tumors; Non-Hodgkin's lymphomas (NHL) account for 60% of all lymphomas. Involvement of the nasal cavity and paranasal sinuses by these tumors is uncommon.[1]

Extranodal NK / T-cell lymphomas, nasal type (ENKL) are aggressive, locally destructive midfacial necrotizing lesions characterized by extranodal involvement, particularly the nasal/paranasal area and represent about 75% of all nasal lymphomas, the rest being B-cell lymphomas.[2] The lesion typically causes local destruction of cartilage, bone and soft tissues. Lesions may arise de novo at the site or represent a localized progression through stages.[3]

Extranodal NK/T-cell lymphoma is a distinct clinicopathologic entity which is often associated with Epstein Barr virus (EBV).

CASE REPORT

A 47-year-old male patient presented with a chief complaint of nasal blockage since 3 months. Examination revealed mass in left nasal cavity. CT Scan report suggested possibility of inverted papilloma with superadded fungal infection. Functional Endoscopic Sinus Surgery (FESS) was done and tissue was sent for histopathological examination.

HISTOPATHOLOGICAL EXAMINATION:

GROSS features: specimen consisted of multiple brownish white soft tissue portions measuring 3x3 cm in aggregate.

MICROSCOPIC features: Hematoxylin and Eosin (H & E) stained sections revealed polypoid lesion lined focally by respiratory epithelium. Subepithelial tissue showed polymorphic population of small lymphocytes, plasma cells and neutrophils. Scattered atypical lymphoid cells with enlarged hyperchromatic nuclei and mitosis were seen. Perinuclear clearing was seen in some of the cells. Dilated blood vessels with angiocentricity by atypical lymphoid cells & focal areas of vascular destruction were observed. Extensive necrotic areas were seen. No granulomatous lesions or giant cells were found. Periodic Acid Schiff (PAS) stain did not reveal presence of fungal elements. From routine H & E stained section, possibility of nasal NK/T-cell lymphoma was rendered.

On immunohistochemical analysis tumor cells were positive for CD 45, CD 3 (cytoplasmic) & bcl-2 while they were negative for CD20 and Pan CK. Ki-67 proliferation index was 50%.

Based on clinical findings of localized destruction of the soft and hard tissues confined to the nose and paranasal sinuses, histopathological features of angiocentric distribution of atypical lymphoid cells, angiodestruction by neoplastic cells, extensive areas of necrosis ,the presence of immunoreactivity with markers indicative of a neoplasm of lymphoreticular origin (CD45, CD3) , the diagnosis of "Extranodal T cell lymphoma" was established.

Before the patient could pursue further immunohistochemical evaluation for CD56, he died of clinical complications.

DISCUSSION:

Malignant lymphomas of the head and neck region that originate in the nasal cavity, paranasal sinuses and hard palate form an interesting and frequently diagnostically difficult group. Variety of pathologic terms have been applied to this lesion, including midline lethal granuloma, polymorphic reticulosis, midline malignant reticulosis, idiopathic midline destructive disease and lymphomatoid granulomatosis.[4]

These lymphomas are uncommon neoplasms in the United States, representing approximately 1.5% of all lymphomas. A higher incidence, however, has been reported in Asian and South American countries, especially Peru. In these areas, primary NHL accounts for approximately 6.7-8.0% of all lymphomas.[4]

The mechanism of tumorigenesis remains unclear for ENKL. EBV is found in most cases of NK-cell leukemia/lymphoma, suggesting an oncogenic role, but patients may have biclonal or polyclonal populations of malignant cells based on differential EBV genome incorporation.[2]

NK/T-cell lymphomas of the head and neck affect patients in a broad age range, with a peak incidence in the fifth decade.[5] The initial signs and symptoms are often localized to the nasal region and include nasal obstruction and chronic rhinorrhea. Systemic symptoms are not typically noted except in advanced cases. The clinical course varies with the clinical stage. Patients with limited stage disease (usually nasal disease) typically have an indolent course with tumor restriction to the original site, but others with advanced stage suffer rapid progression to systemic dissemination often accompanied by hemophagocytosis or disseminated intravascular coagulation.[2]

Computed tomography is the essential tool for precise staging of the lesion. MRI more reliably demonstrates soft tissue invasion and is

useful to evaluate the anatomical relations of the tumor with intracranial structures[6].

Cytogenetic studies demonstrated a variety of cytogenetic aberrations, such as mutation of the Fas and p53 tumour suppressor genes [7]. However, the most common cytogenetic abnormality is a deletion in the q21, q25 or p10 region of chromosome 6.

Histopathological examination of NK/T-cell lymphomas reveals polymorphic infiltrate of inflammatory cells including lymphocytes, plasma cells & neutrophils. Scattered atypical lymphoid cells are present which have enlarged, hyperchromatic, convoluted nuclei with scant basophilic cytoplasm. Frequently, they are surrounded by a zone of clearing, an artifact of formalin fixation that is helpful in spotting these cells at low magnification. They show typical angiocentric distribution with angiodestruction. However, the angiocentric quality of the lesion may be masked in biopsy specimens containing large number of inflammatory cells, foci of necrosis, and proliferating capillaries. Deep biopsies are usually necessary for diagnosis, as superficial biopsy specimens may contain predominantly granulation tissue with acute inflammation; which may hinder the neoplastic nature of lesion. [6]

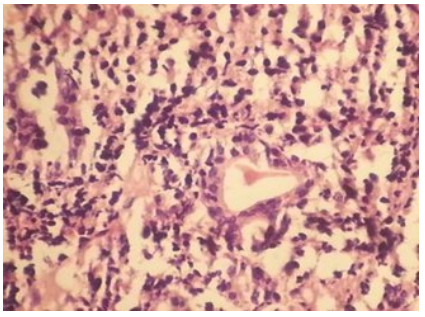

Figure 1: Photomicrograph shows angiocentric (around the blood vessels) distribution of atypical lymphoid cells with infiltration of the vessel wall (H and E Stain, 40X)

Immunohistochemistry provides useful tool to confirm the diagnosis. Neoplastic cells typically express CD45RO and CD43, as well as some T antigens including CD2 and CD3 (cytoplasmic). The neoplastic NK/T cells show CD56 positivity (71%) and CD57 negativity. Almost all NK/T-cell lymphomas contain large amounts of EBV RNA by in situ hybridization. The biological behavior of NK-positive and T-positive nasal extranodal lymphomas is similar; hence they are collectively termed NK/T-cell lymphomas.

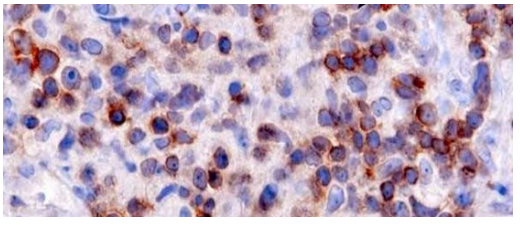

Figure 4 – Photomicrograph shows Cytoplasmic staining for polyclonal CD3

Despite progress in immunohistochemistry and molecular biology, extranodal NK/T-cell lymphoma, nasal type, still remains a diagnosis of exclusion due to the absence of any specific clinical and histopathological features. Many diseases can present in the form of nasal and mid-facial ulceration and destruction, similar to that caused by extranodal NK/T-cell lymphoma, such as Wegener's granulomatosis, syphilis, tuberculosis, other malignant tumours and cocaine abuse. [8]

This tumour has a poor prognosis, with a 5-year overall survival ranging between 10 to 45% depending on the series. [9]

CONCLUSION:

Extranodal NK/T-cell lymphoma, nasal type, is a rare disease, but the number of new cases reported each year has been continually increasing over recent years due to advent of ancillary techniques for the diagnosis. Extranodal NK/T-cell lymphoma often poses diagnostic difficulties and so, the management is delayed. Histopathological examination in conjunction with immunohistochemistry is essential to establish the diagnosis. The disease has poor prognosis with a high local recurrence rate.

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