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Case Report

Sinonasal T-Cell Lymphoma: A Case Report

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Abstract

Peripheral T and NK lymphomas of the nasal cavity are a heterogeneous group of neoplasia of lymphocytic origin. Nasal T/NK lymphoma is a rare and severe disorder in Africa and Europe. It is an aggressive form of non-Hodgkin's lymphoma with specific clinicopathological features. Diagnosis is based on immunohistochemical study. Treatment includes chemotherapy and radiotherapy. This type of lymphoma has a poor prognosis overall, even with treatment. We report a case of T / NK lymphoma simulating an orbito-jugal cellulitis. This is the case of a 49-year-old patient admitted with an extended left facial cellulitis. The CT scan of the facial mass showed a left unilateral pansinusitis with orbital infiltration. The diagnosis was made after an immunohistochemical study of the nasal biopsies. Histological examination revealed nasal-type T / NK lymphoma. The treatment consisted of radio-chemotherapy. T / NK lymphomas are aggressive; they mainly affect the nasal sinus cavities. They are responsible for angio-destruction and necrosis which make the symptoms nonspecific and the biopsies often negative, causing a problem of differential diagnosis. The treatment is based on radiotherapy and chemotherapy and the prognosis remains poor. The patient was treated with CHOEP chemotherapy followed by radiotherapy.

Keywords: Extranodal NK/T-cell Sinonasal lymphoma - Immunohistochemistry.

INTRODUCTION

Nasal-type extra-ganglionic T / NK (natural killer) lymphoma (LTNK) is a rare form of malignant non-Hodgkin lymphoma, accounting for 1.4% of all lymphomas [1]. It corresponds to a clinical entity carrying out a necrosis which begins preferentially in the nasal cavities and the nasopharynx to extend to the centrofacial bone structures, whose spontaneous evolution remains fatal. Its pathological diagnosis is not always obvious. Modern immunophenotyping and molecular biology tools have made it possible to isolate this type of lymphoma. Treatment is based on radiotherapy and chemotherapy. We report a case and discuss the particularity of its localization, the diagnostic difficulty and the therapeutic modalities.

CASE REPORT

This is a 49-year-old patient with no particular pathological history admitted for left orbital-cheek cellulitis. The symptomatology goes back to several years by a permanent unilateral left progressive nasal obstruction, associated since 3 months with an unilateral purulent rhinorrhea and epistaxis, and headache resistant to oral treatment with antibiotics. It was evolving in a context of fever and deterioration of the General condition, complicated by a swelling of the left hemiface and left eyelid with slight exophthalmos and odynophagia and closed rhinolalia.

The otolaryngologic examination revealed a swelling of the left hemiface centered on the lateral wall of the left nasal pyramid extended to the internal cantus and the lower eyelid, erasing the nasolabial fold with left nasal obstruction, and thick purulent rhinorrhea filling both nasal cavities more marked on left side.

Endoscopic examination revealed a very swollen inflammatory nasal mucosa bleeding on contact, preventing the progression of the endoscope, with the presence of thick purulent secretion associated to false membranes.

Examination of the oral cavity showed cryptic pultaceous tonsils associated to a swelling of the soft palate and ulceration of the hard palate.

Ophtalmic examination shows left ocular swelling without any collection nor ophthalmoplegia. The rest of the clinical examination was normal, especially that of the lymph node areas.

Cytobacteriology examination of the nasal swab showed a gram-negative bacilli and the culture revealed Acinetobacter baumanii sensitive to antibiotics administered by IV, associated with antifungal treatment, but without much clinical improvement.

The biopsy samples for the Xpert gene test were negative for tuberculosis. Nasosinus CT-scan revealed left pansinusitis and right sphenoidal and ethmoidal sinusitis of chronic appearance, left pre-septal cellulitis associated with ipsilateral dacryocystitis. (Figures 2 and 3)

Angio-MRI: revealed left hemi-pansinusitis and right maxillary sinusitis, orbital cellulitis with left medial extra-conical abscess. Left nasopharyngeal inflammation, with absence of cerebral venous thrombosis (Figure 4).

The histopathological study of biopsy samples from the left nasal cavity taken under local anesthesia found friable tissue with necrotic areas, suggestive of infectious or tumoral pathology, without being able to decide between the two. Immunohistochemistry diagnosed sinonasal NK/T-cell lymphoma NOS: the lymphoid elements were positive for CD7, CD30 and anti garenzyme B. (Figures 5-6).

The patient then was addressed to the hematologic unit, and was treated with CHOEP chemotherapy followed by radiotherapy.

Fig-1: Swelling of the left hemiface centered on the lateral wall of the left nasal pyramid extended to the internal cantus and the lower eyelid/ ulceration of the hard palate

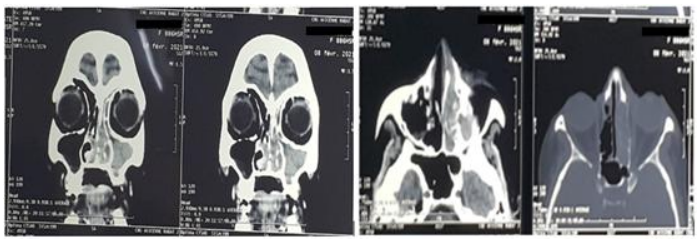

Fig-2-3: coronal CT- scan: left pansinusitis and right sphenoidal and ethmoidal sinusitis of chronic appearance, left pre-septal cellulitis associated with ipsilateral dacryocystitis.

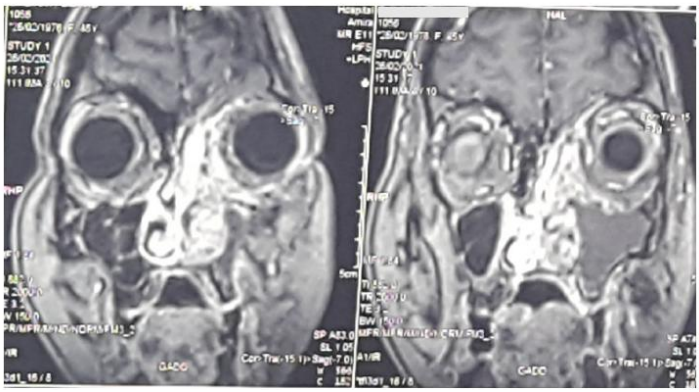

Fig-4 Angio-MRI: left hemi-pansinusitis and right maxillary sinusitis, orbital cellulitis with left medial extra-conical abscess. Left nasopharyngeal inflammation, with absence of cerebral venous thrombosis

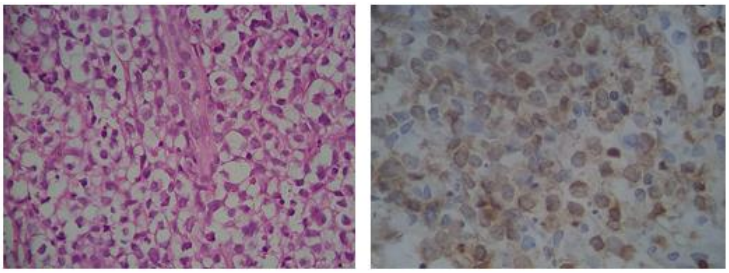

Fig-5-6: photomicrograph showing a broadly neurotic tumor proliferation made up of diffuse webs of medium-sized monomorphic cells HE at Gx20. Tumor cells have vesicular chromatin anisocayotic nuclei with HE angiotropism at Gx40. Immunostaining tumor cells express CD7 and CD30

DISCUSSION

Sinonasal lymphomas were confused with various infections, autoimmune and inflammatory diseases. Now, they are considered to represent peripheral T cell lymphomas [1]. Nasal T cell lymphomas usually originate from the nasal cavity and invade the adjacent structures that like the sinonasal B cell lymphomas usually arise from the paranasal sinuses and is more common in the western population [2]. The usual symptomatology of sinonasal lymphomas are nasal obstruction, discharge, epistaxis, unilateral facial and cheek swelling, headache and symptoms secondary to tumour extension.

The diagnosis of destructive diseases of the sinonasal region depends on clinical and pathological findings as imaging in these lesions may not confirm specific diagnosis. Nasal obstruction and nasal discharge which are commonly seen in sinonasal lymphoma are also seen in rhino sinusitis as well [3].

The diagnosis of sinonasal lymphoma is difficult because of the presence of ischemic type of necrosis, inflammatory cells, and polymorphous composition of neoplastic cells. The main differential diagnosis for sinonasal lymphoma is Wegener's granulomatosis. At times, it may be difficult to distinguish these diseases histilogically [4]. Adequate amount of biopsy material and immunohistochemistry helps in such situations. Sinonasal lymphoma cases express T cell markers such as CD 2, CD 45RO and CD 43 and often express CD 56. CD 20 is universal B cell marker. The diagnosis of Wegener's granulomatosis is based on histopathological features like the presence of non caseating multinucleated giant cell granulomas and necrotizing vasculitis. The diagnosis maybe completed biologically by elevated titers of anti-neutrophil cytoplasmic antibodies [3, 4] wich was not the case of our patient.

In addition, all other chronic inflammatory and granulomatous diseases of the nose will have to be kept in mind as differential diagnosis. Also, trauma and cocaine abuse can be considered as differential diagnosis as these entities are important causes of septal perforation which is a common feature in sinonasal lymphoma [3], though not seen in our case.

Nasal T cell lymphomas respond well to multidrug chemotherapy followed by field radiotherapy [5]. In spite of this, death from this disease occurs in 50% of patients as a result of distant extra nodal spread or relapse [6, 7]. Patients with sinonasal lymphomas have a better prognosis compared to those with nodal lymphomas of similar grades.

Nasal NK/T cell lymphoma has a highly aggressive clinical manifestation with poor outcome and short survival times. Patients with stage I/II disease are treated with radiotherapy. Multiagent chemotherapy (CHOP regimen) with or without involved field radiation appeared to be another effective treatment. When compared to other type of lymphoma of head and neck, the response rate seems lower and the local relapse rates were high in NK/T cell type (21.4%). A complete response is estimated in 56% of cases. Median survival reported as 12.5 months and of the present of lymphoma cell dissemination, the survival rate is less than 6 months [6, 7].

CONCLUSION

Diagnosis of sinonasal lymphoma is often difficult. The purpose of publication of this article is for its rarity, the difficulty in diagnosis, the need for adequate biopsy material which may require an incisional biopsy and the need for immunohistochemical study. A careful examination and adequate biopsy avoid an unwarranted delay in the diagnosis. The prognosis remains poor.

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