



E-ISSN: 2706-9575
P-ISSN: 2706-9567
IJARM 2020; 2(1): 12-14
Received: 16-01-2020
Accepted: 18-02-2020

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Disfiguring and fatal evolution of a rare and aggressive T lymphoma: Nasal NK

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DOI: <https://doi.org/10.22271/27069567.2020.v2.i1a.31>

Abstract

Lymphoma T nasal NK is a rare type of non-Hodgkin's lymphoma of the ENT region. The diagnosis is based on the clinic and immunohistopathology. This type of lymphoma has a poor prognosis even under treatment at advanced stages, requiring multidisciplinary management. We report a case in a young patient whose evolution was fatal with a rapidly evolving and disfiguring character.

Keywords: NK nasal T lymphoma - rare - Disfiguring evolution

Introduction

NK/T nasal lymphoma, formerly known as central-facial malignant granuloma, is a malignant lymphoma most often found in the ENT region, hence its qualifier "nasal type" [1]. It is a rare lymphoma, its diagnosis is based on the clinic and immunohistopathology. The prognosis for this condition is poor even when treated with chemotherapy and radiotherapy. We report a case of underdiagnosed T/NK lymphoma in a young patient whose course was fatal.

Case report

A young 32-year-old adult, chronic smoking for 15 years weaned 01 month ago without any other significant pathological history, initially consulting for a red, painful and indurated swelling in the wing of the left nose [Figure 1] without other signs associated in particular no nasal obstruction nor rhinorrhea the whole evolving in a context of febrile sensation and unencrypted weight loss, having benefited from an emergency scanner objectifying an infiltration of the lower palpebral soft parts, jugale and left nasolabial extended to the pit homolateral nasal cavity without bone collection or lysis with filling of the left maxillary sinus and some ethmoid cells, the patient was treated with antibiotic therapy for suspected cellulitis. The evolution was marked by a worsening 15 days later, with the installation of an ulcer of irregular contours at the level of the wing of the left nose [Figure 2 (A)] gradually increasing in size [Figure 2 (B)] until destruction of the nasal wall after one month [Figure 2 (C)]. There were no cervical, axillary or inguinal lymphadenopathies. The somatic examination revealed no other signs of appeal. A skin biopsy with immunohistochemical study was performed objectifying a malignant tumor process compatible with nasal T lymphoma (NK), with expression by tumor cells of CD2, CD3, CD56 and some isolated neoplastic cells are marked by anti EBV. A biological assessment was carried out showing a normal LDH at 233 and a microglobulin BETA2 increased to 6.47.

As part of the extension assessment, a cranio facial +cervico thoraco abdomino pelvic CT scan was performed showing a tissue process centered on the left nasal fossa enhanced after contrast measuring approximately 38x28x68 mm invading the anterior and posterior ethmoid cells below. homolateral and extending to the sub chin tissues bilaterally as well as into the maxillary sinus responsible for its total obstruction, extending upwards in the left frontal sinus infiltrating laterally to the left the bilateral soft tissue, periorbital and homolateral jugals responsible for a thickening of the latter and presents intimate contact with the eyeball without border of separation respecting the contralateral nasal fossa on the right side without any sign of secondary location at a distance.

The patient was referred to internal medicine for additional treatment. The clinical course was marked by the extension of the mid-facial ulceration surmounted by fibrin [Figure 3 (A)] arriving at the left lateral wall of the nose and the upper lip [Figure 3 (B)] then arriving at the lip inferior and the left orbital region [Figure 4 (A)] progressing towards a loss of necrotic center substance [Figure 4 (B)], gradually destroying the nasal, and buccal region, extending towards the frontal region at the top, the orbital, genial and malar regions bilaterally, exposing the maxillary bone which appears necrotic after two months [Figure 5]. The patient benefited initially from a polychemotherapy (4 cures according to the CHOEP protocol: Adriamycine, Vincristine, Cyclophosphamide, Etoposide and Prednisone) with a non-response at 24% on the re-evaluation scanner, hence his placing on 4 cures of methotrexate +L asparaginase. But in front of the aggravation of the losses of facial substance and the progression of the tumor process in the cavum with extension to the nasopharynx and to the oropharynx and appearance of secondary pulmonary and ganglion mediastinal localizations objectified with the control scanner, the decision was to put it under ICE (Ifosfamide, Carboplatin and Etoposide) having received two cures. Spontaneous evolution was fatal, the patient died soon after.

Discussion

Central facial malignant granuloma was first described in 1933 and recognized by WHO in 2001 as nasal T/NK lymphoma [2]. It mainly affects adults (5th decade) and male subjects [3], when our patient was only 32 years old. Present clinically by plaques and/or tumors affecting the extremities or the trunk, but also and especially the mid-facial line in the destructive nasal form as in the case of our patient, dominated by local signs such as unilateral nasal obstruction, purulent and/or streaked rhinorrhea, recurrent epistaxis and a chronic sinusitis pattern [4] as well as general signs such as asthenia, fever, weight loss, and macrophagic activation syndrome can be demonstrated [5]. It is characterized by its clinical polymorphism which can explain the diagnostic problems and the delay in therapeutic treatment often observed. Indeed, the mode of revelation of this pathology is not very specific which can even confuse the clinician. The diagnosis is based on histology and immunohistochemistry showing a proliferation of atypical lymphocytes with angiotropism and angiodestruction leading to the presence of necrotic phenomena and ulcers characteristic of this type of lymphoma with immunohistochemical confirmation by revealing an expression of CD2, CD56 and cytotoxicity markers (T1a, Granzyme B, perforin). The T markers, in particular CD5, CD4, membrane CD3, CD8 are negative [2]. Epstein Barr virus (EBV) is present in almost all tumor cells in 100% of patients [6]. Computed tomography of the sinuses of the face is used to orient the diagnosis and MRI has an interest in the assessment of extension to adjacent structures. The treatment is based on a combination of radiotherapy and polychemotherapy, but the prognosis remains grim with a rapidly unfavorable course. The median spontaneous survival of these patients has recently been evaluated at 7 to 8 months [7].

Conclusion

This lymphoma is rare, and its prognosis is poor including under treatment in advanced stages, multidisciplinary management would allow early diagnosis and management. The case we reported shows a decaying rapid course and the severity of the prognosis for this type of lymphoma.



Fig 1: Painful and hard red swelling on the wing of the left nose.



Fig 2: Ulceration of irregular contours of the wing of the left nose (A) gradually increasing in size (B) with extension until destruction of the nasal wall (C)



Fig 3: Vast mid-facial ulceration (A) reaching the left lateral wall of the nose and upper lip (B)



A

B

Fig 4: Extension of the ulceration above the left orbital region and below reaching the lower lip (A) progressing to extensive loss of substance with necrotic center (B).



Fig 5: Extension of the loss of substance occupying the nasal and buccal region, extending towards the frontal region at the top, the orbital, genial and bilateral malar regions exposing the maxillary bone

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