Report on an atypical case of non-Hodgkin's lymphoma type B of the subcutaneous soft tissues of the cheek region

Case Report

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ABSTRACT

Primary extra nodal non-Hodgkin's lymphoma (NHL) of the skeletal muscles is a well-recognized entity, although such occurrences are not very frequent. Even rarer is its presentation in the facial muscles.
This report details a case of a primitive extra nodal type B non-Hodgkin's lymphoma in a non-immunosuppressed patient involving only subcutaneous tissues and muscles of the cheek region and not extending to the oral cavity or sino-nasal passages. The patient was subsequently treated with chemotherapy and, at the time of writing, was in remission.
This case highlights the diagnostic challenges posed by atypical symptoms. Given the unusual presentation, it emphasizes the need to consider lymphoma as a potential cause of malignant infiltration in facial muscles. The rarity of such cases often leads to delays in diagnosis and treatment, underscoring the importance of awareness among clinicians. The patient's treatment response underscores the efficacy of chemotherapy in managing this rare manifestation of NHL.

Key Words: Lymphoma, Non-Hodgkin, Face

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INTRODUCTION

After squamous cell carcinoma, lymphoma is the most common malignant tumor of the cervical cap. However, non-Hodgkin's lymphoma (NHL) remains a rare tumor of the subcutaneous tissues of the face. Only a few cases of malignant lymphoma infiltration of head and neck muscles have been published, and these mainly concerned the muscles of mastication.

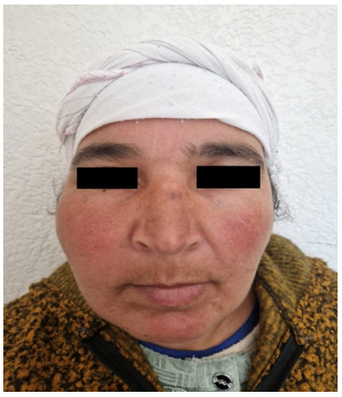
We report here the case of an immunocompetent patient presenting with a primary type B non-Hodgkin's lymphoma localized in the subcutaneous tissues of the cheek region, which illustrates the diagnostic difficulties encountered in the face of an unambiguous symptomatology.

A review of the literature in the PubMed and Medline databases identified only one similar case, described by Liapi et al. (1).

CASE REPORT

A 45-year-old female with no notable medical or surgical history presented with a right cheek swelling that had persisted for four months and appeared inflammatory. Clinical examination identified a solid, immobile lump measuring 5 cm in length, situated in the right cheek (Figure 1). There was no lymph node involvement, and the rest of the physical examination was normal.

Figure 1 : Clinical image showing swelling of the right cheek.



A CT examination of the face revealed a tumor-like tissue mass in the right cheek subcutaneous soft tissue. The mass was poorly defined, lobulated in outline, and measured 48 x 19 mm in axial diameter. It was centered on the right zygomatic bone and caused a grass-fire periosteal reaction at this level (Figures 2 and 3). These clinical and radiological findings were consistent with a tumoral process.

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Figure 2 : CT axial section showing a tissue process over the right zygomatic bone.

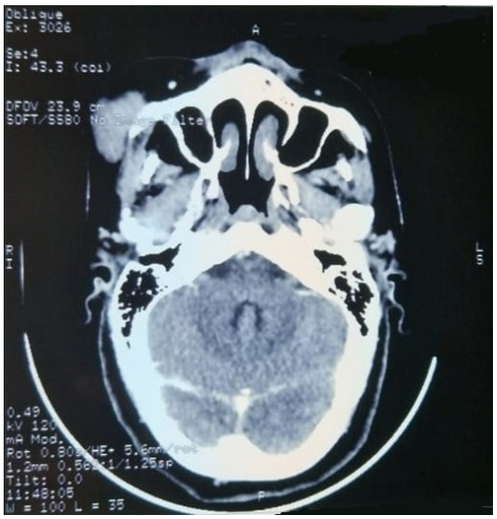
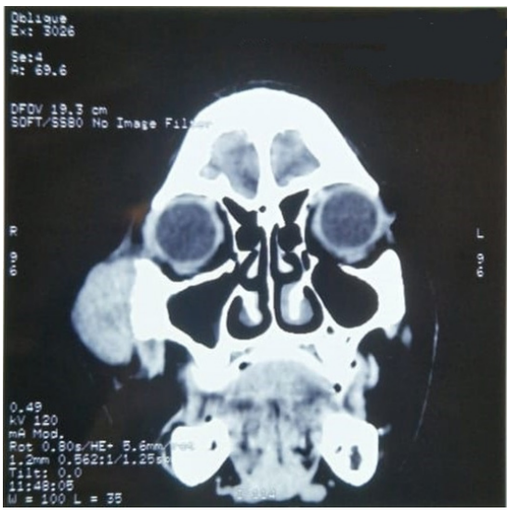


Figure 3 : Coronal CT section showing a tissue process opposite the right zygomatic bone.



In light of these findings, surgical exploration was conducted to establish a diagnosis. Biopsies were taken using the Caldwell Luc incision, revealing the tumor to be both friable and strongly adherent to the zygomatic bone.Histological examination revealed densely cellular tumor proliferation arranged in a diffuse, artefactual sheet, characterized by blue tumor cells and hyperchromatic. Immunohistochemical analysis was performed, revealing the following:

• The cells diffusely expressed CD20, and their nuclei were strongly marked by PAX5.

• Numerous T cells at the periphery expressed CD3 and were strongly labeled with Ki67.

All these immunohistochemical features are characteristic of a high-grade malignant non-Hodgkin's lymphoma (NHL) with a diffuse large B-cell phenotype. An extension workup, including a chest X-ray, cervical and abdominal ultrasound, and thoraco-abdominal CT scan, showed no dissemination or other localizations.
The lymphoma was therefore concluded to be high-grade, diffuse large B-cell type, primary, localized in the soft tissues of the cheek region, and classified as stage IE according to the Ann Arbor classification (Table 1).
Therapeutic management consisted of six courses of R-CHOP chemotherapy. The response to treatment was very favorable, with complete remission. At the time of writing, the patient is being followed up every three months and shows no signs of therapeutic failure.

Table 1: Ann Arbor staging system for lymphoma

I	Involvement of a single lymph node region (I) or a single extra-lymphatic organ or site (IE)
II	Involvement of 2 or more lymph node regions on the same side of the diaphragm (II) or localized involvement of extra-lymphatic organ or site and 1 or more lymph node regions on the same side of the diaphragm (IIE)
III	Involvement of lymph node regions on both sides of the diaphragm (III), which may also be accompanied by localized involvement of extralymphatic organ or site (IIIE), involvement of the spleen (IIIS), or both (IIISE)
IV	Diffuse or disseminated involvement of 1 or more extra-lymphatic organs or tissue with or without associated lymph node enlargement

DISCUSSION

Lymphomas encompass a diverse group of malignant neoplasms arising from lymphocytes or their precursor cells. They are broadly categorized into two main types: Hodgkin lymphomas (HL) and non-Hodgkin lymphomas (NHL), distinguished by their histological characteristics and clinical behavior. Hodgkin lymphomas constitute approximately 14% of all lymphomas, while non-Hodgkin lymphomas account for the remaining 86% [2].
Non-Hodgkin's lymphomas (NHL) have a significant tendency to spread to extra-ganglionic sites, which occurs in almost 25% of cases. In the majority of cases, lymph node involvement is also observed. In the cervicofacial region, Waldeyer's lymphatic ring is the most frequent site of NHL, followed by the main salivary glands, notably the parotid and submandibular glands [3].
Infiltration of skeletal muscle by lymphomatous tissue is extremely rare. Komatsuda et al. found muscle involvement in only 1% of the 2147 cases of malignant lymphoma studied between 1976 and 1978 [4]. In HIV-positive patients, NHL is 60 times more frequent than in the general population, with muscle involvement in 8.8% of cases [5].

Muscle infiltration can occur through metastatic spread, direct extension to adjacent tissues, or as a primary extranodal lesion [6, 7]. Muscles most frequently affected include those in the extremities, gluteal region, paraspinal area, and pelvis [8].
In the head and neck region, cases of primary muscular NHL are exceptionally rare, mostly involving the muscles of mastication. Liapi et al. reported a single case in a 64-year-old immunocompetent patient with NHL of the facial muscles, particularly the skin muscles, without nasosinus or cranial nerve involvement [1].Imaging tools are pivotal in diagnosing skeletal muscle lymphoma. CT scans are essential for assessing bone invasion and estimating lesion size within the TNM classification framework. Lymphomatous lesions typically appear parenchymal and isodense, with potential invasion into adjacent cavities. Generally, bone destruction is less pronounced in NHL compared to squamous cell carcinoma. Moreover, reactive bone sclerosis due to malignant lymphoid infiltration is often evident, influenced by the tumor's growth rate [9, 10].

MRI is considered the most effective radiological modality for assessing skeletal muscle lymphoma, despite its cost and limited accessibility for some patients. On MRI, NHLs involving skeletal muscle typically appear hyper intense or isointense compared to normal muscle on T1-weighted sequences, and hyper intense on T2-weighted and fat suppression sequences [11].
Radiological features and discrepancies between imaging findings and clinical manifestations can suggest a potential diagnosis of a malignant tumor lesion. However, the definitive diagnosis relies on precise histological analysis of tumor proliferation and characterization of its phenotype through immunohistochemical studies. These immunohistochemical criteria are crucial in determining the treatment plan, as they, along with disease staging, are key prognostic factors.
The most commonly used classification system for staging non-Hodgkin's lymphomas (NHL) is the Ann Arbor system, which considers the number of tumor sites (lymph node and extra nodal), their location, and the presence or absence of B symptoms. Treatment of NHL typically involves multi-agent chemotherapy as the mainstay, either alone or combined with external radiotherapy (30 to 50 Grays), particularly in more advanced cases (stages T3, T4). Localized clinical forms, such as stage IA E, generally have a favorable prognosis, with 5-year survival rates ranging from 70% to 100% [12].

CONCLUSION:

Non-Hodgkin's lymphoma rarely affects facial muscles. Achieving an accurate clinical and pathological diagnosis, as well as precise staging, requires a high level of suspicion and close collaboration among clinicians, radiologists, and pathologists. A multidisciplinary approach is crucial for ensuring optimal management of patients with this rare presentation.

Competing interests
The authors declare no competing interests.

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Informed Consent:
Patient's consent was taken before presenting the case report and intraoral pictures.

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