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An unusual case of primary extranodal non-Hodgkin's lymphoma in the muscles of facial expression

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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 common. Presentation in the muscles of the face has only rarely been described. We present a case of primary extra-nodal NHL in a non-immunocompromised patient involving only the muscles of facial expression and not extending to the oral cavity or sinonasal tract. The patient was subsequently treated with chemotherapy and at the time of writing remained in remission. We suggest that lymphoma should be considered among the causes for malignant infiltration of the muscles of the face.

Key words: Facial Muscles; Lymphoma, Non-Hodgkin; Antineoplastic Agents

Introduction

Non-Hodgkin's lymphoma (NHL) presents as an extra-nodal disease in 25–40 per cent of patients.[1,2] Primary involvement of the skeletal muscles is rare and usually affects the extremities such as the gluteal and pelvic areas.[3–8] Only a few cases of malignant infiltration by lymphoma in the muscles of the head and neck area have been published and these have involved the muscles of mastication.[9–12] We describe a case of a non-immunocompromised patient who presented with a non-Hodgkin's lymphoma in the muscles of facial expression. There was no extension into the sinonasal tract or the oral cavity and the cranial nerves and overlying skin were not affected. We have undertaken a PubMed and Medline search and to our knowledge there has been no other similar case reported in the literature.

Case presentation

A 64-year-old woman presented with a three-month history of painless swelling over her left cheek. She was otherwise fit and was aware of the swelling only when wearing her glasses. There were no associated nasal or neurological symptoms. She denied any history of weight loss, night sweats or fever. On examination, a firm swelling was present to the left of the nasal bridge occupying the nasolabial fold, which was approximately 2 cm at its greatest dimension. Examination of the nose, oral cavity and oropharynx was normal. There was no lymphadenopathy and the rest of the head and neck examination was unremarkable.

A computerized tomogram (CT) scan of the face and the paranasal sinuses revealed a soft tissue swelling with well defined edges extending from the left nasolabial fold as far as the infra-orbital foramen and involving the adjacent muscles; there was no bone involvement. The left maxillary and ethmoid sinuses were completely clear and there was no extension to the oral cavity (Figures 1 and 2).

In view of the findings, surgical exploration was undertaken to establish a diagnosis. Biopsies were taken via a sublabial approach and the tumour was found to be intimately attached to the infra-orbital nerve.

Macroscopically, the lesion was a circumscribed, cream-coloured nodule, 20 mm in its maximum dimension. Histological examination showed it to consist of a lymphoid infiltrate encasing nerve fascicles and also extending into the muscle fibres and adipose tissue. The lymphoid infiltrate consisted of centrocyte-like cells of B cell lineage (CD 20 and CD 79 positive on immunohistochemistry) with many reactive lymphoid germinal centres. Immunohistochemistry also demonstrated kappa light chain restriction. The features were typical of extra-nodal marginal zone B cell lymphoma (NHL) as stated by the Revised European American lymphoma (REAL)/World Health Organization classification (Figure 3).

The patient was subsequently referred for a full haematological investigation and staging. A complete systematic workup (including: CT scans of the chest, abdomen and pelvis; a bone marrow biopsy; and a bone scan) did not reveal any other sites of disease. The results of laboratory examination (full blood count, serum electrolytes, lactate dehydrogenase level, liver function tests and serum immunoglobulins) were all negative and the patient was evaluated as stage IE according to the Ann Arbor system (Table I).

The patient was treated with single-agent chemotherapy (chlorambucil) and at the time of writing was in remission (12 months following presentation).

Discussion

Infiltration of the skeletal muscles by lymphomatous tissue is considered extremely rare. In a series of 2147 cases of malignant lymphoma from 1976 to 1978 Komatsuda *et al.* found involvement of muscle in only 1.4 per cent.[3] In HIV-positive patients NHL is approximately 60 times more frequent than in the general population and

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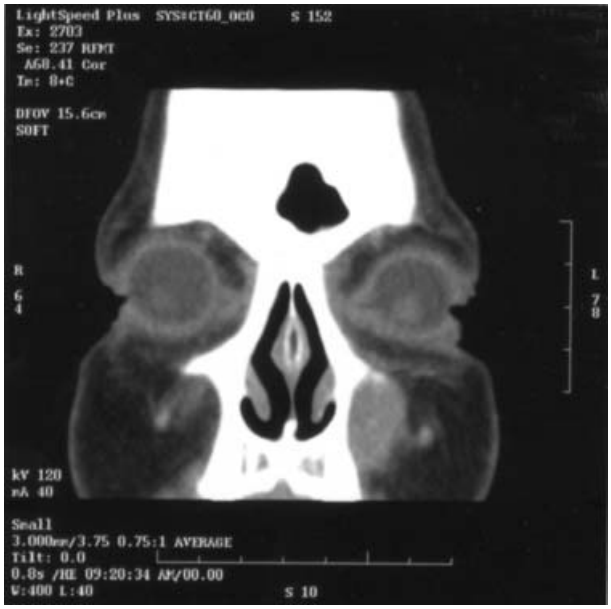

FIG. 1

Coronal computed tomogram showing the soft tissue swelling in the left nasolabial fold with ill-defined edges attached to the surrounding muscles.

lymphomatous involvement of the muscles can be seen in 8.8 per cent.[13] Infiltration of muscles in lymphoma can occur via metastatic spread, direct extension from adjacent affected tissues or as a primary extra-nodal lesion.[4,5] In a recent study by Samuel *et al.*[7] all patients with primary muscle NHL registered with the Scotland and Newcastle lymphoma group over a 15-year period were reviewed. The database included over 6000 lymphoma patients. The authors retrieved all the cases recorded to involve lymphomatous infiltration of muscle and subsequently excluded patients from the analysis if the muscle

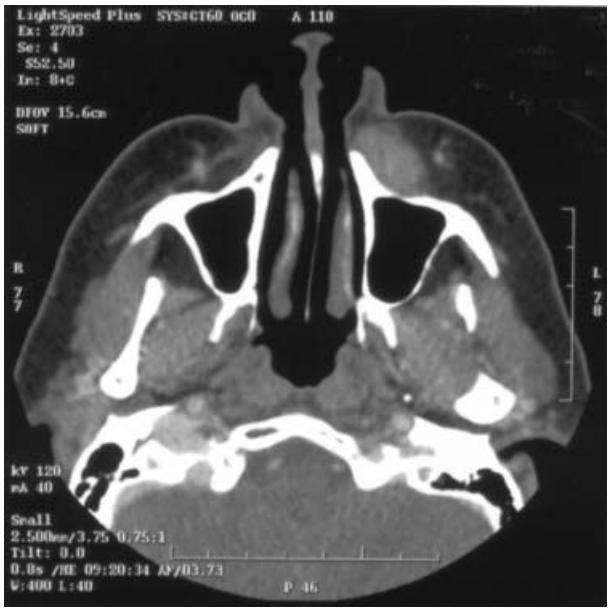

FIG. 2

Axial computed tomogram showing the mass in the soft tissues with no extension into the left maxillary sinus or the left nasal fossa.

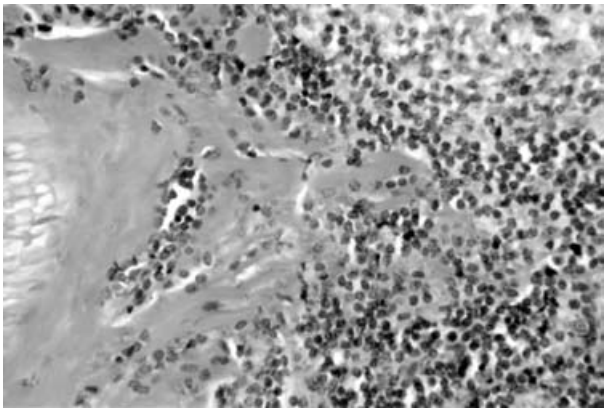

FIG. 3

High-power photomicrograph showing neoplastic marginal zone lymphoid cells invading normal structures. A nerve is identifiable on the left with residual non-neoplastic lymphoid follicle in the upper right corner (H & E; original magnification ×3).

infiltration was simply an extracapsular spread from adjacent lymph nodes or if there was soft tissue involvement only. Using strict criteria only eight patients with primary NHLs of muscle were identified. A review of the English literature in 1999 revealed only 31 cases of primary skeletal muscle lymphoma.[8] The muscles most commonly affected are those of the extremities as well as the glutei, paraspinal and pelvic muscles.[7,8]

In the head and neck area only six cases of primary NHL arising in the muscles have been previously reported.[9–12,14] Five of these cases involved the muscles of mastication.[9–12] One of these patients was also infected with HIV.[9] Amo *et al.* reported a 75-year-old patient with NHL affecting the facial muscles.[14] However the patient also had skin lesions which were positive for lymphomatous infiltration. The trigeminal and facial nerves were also affected and the patient presented with facial hemiplegia and paraesthesia. Reichman *et al.* described an NHL presenting as facial swelling in a paediatric patient.[15] Histopathology proved that the disease affected the soft tissues overlying the maxillary antrum and not the muscle. The disease also extended to the sinonasal tract, obstructing the maxillary and ethmoid sinuses.

TABLE I

ANN ARBOR STAGING SYSTEM FOR LYMPHOMA

Stage	Definition
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 extra-lymphatic 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

Each stage can be classified into A (no systemic symptoms) or B (fever, sweats and/or weight loss >10 per cent of body weight).

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In our case the disease was restricted to the muscles of the face from the nasolabial fold to the infra-orbital area. There were no associated skin lesions and no extension to the oral cavity or sinonasal tract. Our patient also did not have any cranial nerve impairment. However, as with previously reported and similar cases our patient did not present with typical symptoms such as fever, weight loss and night sweats and she was also immunocompetent.

Other, similar cases have been treated either with combined chemotherapy,[9,12] radiotherapy plus chemotherapy[14] or with radiotherapy alone.[9] As with the patient described by Set *et al*.[10] our patient received only single-agent chemotherapy and responded very well. At the time of writing she was in remission 12 months following diagnosis.

Conclusion

Non-Hodgkin's lymphoma is a rare entity in the muscles of the face. A high index of suspicion and close co-operation between clinician, radiologist and pathologist is essential for appropriate clinical and pathological diagnosis and staging, as a multidisciplinary approach is crucial in the appropriate management of these patients.

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