

CASE REPORT

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# An extremely rare presentation of non-Hodgkin lymphoma in the head and neck: a case report

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## Abstract

**Background:** Non-Hodgkin lymphoma (NHL) frequently presents as extra-nodal involvement, although primary diffuse large B cell lymphoma of extra-nodal origin is a distinct entity. Different subtypes may have propensity for specific anatomic locations. In the head and neck region, diffuse large B cell lymphoma (DLBCL) is most commonly presented in the paranasal sinuses, jaws, and Waldeyer's ring. DLBCL involving fibroadipose tissue and presenting as an extra-nodal mass is extremely unusual, particularly in the head and neck region.

**Case presentation:** We will present a case of forehead mass as presenting features of a highly aggressive B cell lymphoma.

**Conclusion:** Although soft tissue DLBCL presenting as scalp mass is rare, it should be considered in differential diagnosis of every soft tissue lump.

**Keywords:** Non-Hodgkin lymphoma, Lymphoma, Paranasal sinuses, Mass, Forehead, B cell lymphoma

## Background

Hodgkin lymphoma (HL) and non-Hodgkin lymphoma (NHL) are main types of lymphoma, with various subtypes according to WHO classification system. NHL presents as extra-nodal lesion, more frequent than HL. Clinical features are helpful in determination of these two types, but just cellular examination accurately defines types and subtypes. Imaging modalities are helpful in assessment of tumor progression and bone destruction. Highly aggressive subtypes of NHL like Burkitt, diffuse large B cell lymphoma (DLBCL), and natural killer (NK)/T cell lymphoma show a destructive pattern [1]. The gastrointestinal tract is the most common site of extra nodal presentation of lymphoma; the head and neck region is the next. Almost 2.5% malignant lymphomas

present in oral and para-oral sites [2]. NHL frequently presents as extra nodal involvement (25–30%), although primary DLBCL of extra nodal origin is a distinct entity [3]. Different subtypes may have propensity for specific anatomic locations. DLBCL is most commonly presented in the paranasal sinuses, jaws, and Waldeyer's ring [1]. DLBCL involving fibroadipose tissue and presenting as an extra nodal mass is extremely unusual, particularly in the head and neck region. In this case, we will introduce a case of forehead mass as presenting feature of a highly aggressive B cell lymphoma, in order to emphasize the diversity of presentations of malignant lymphoma.

## Case presentation

A 56-year-old male was admitted to otolaryngology clinic of a referral hospital, with a chief complaint of forehead mass. He had noticed the left supraorbital mass with a gradual growth, after bitten on vertex scalp by a spider 2 weeks before visit (Fig. 1). He also mentioned postnasal discharge, mild frequent frontal headache, night sweating, and significant weight loss in recent

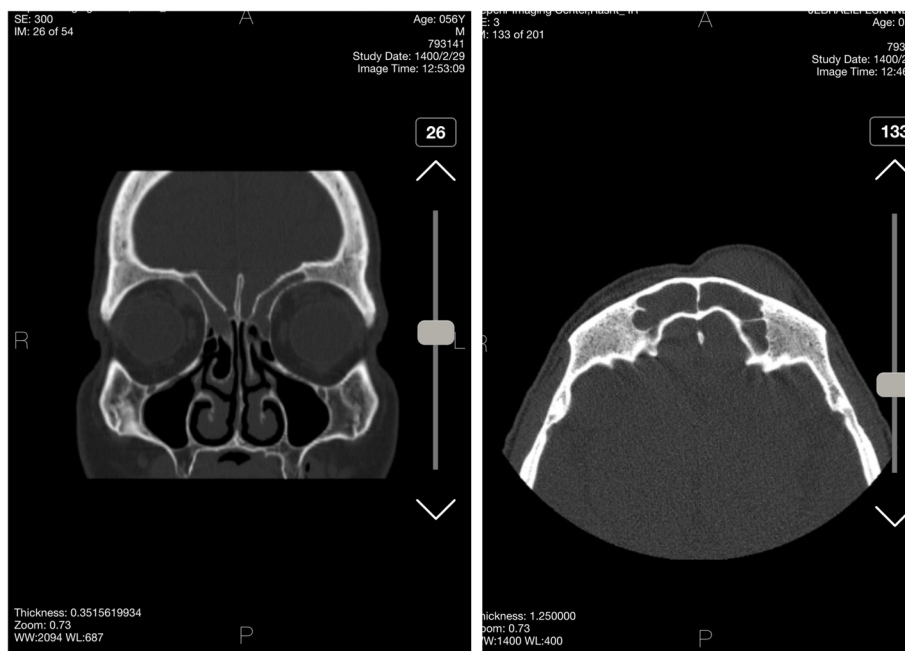
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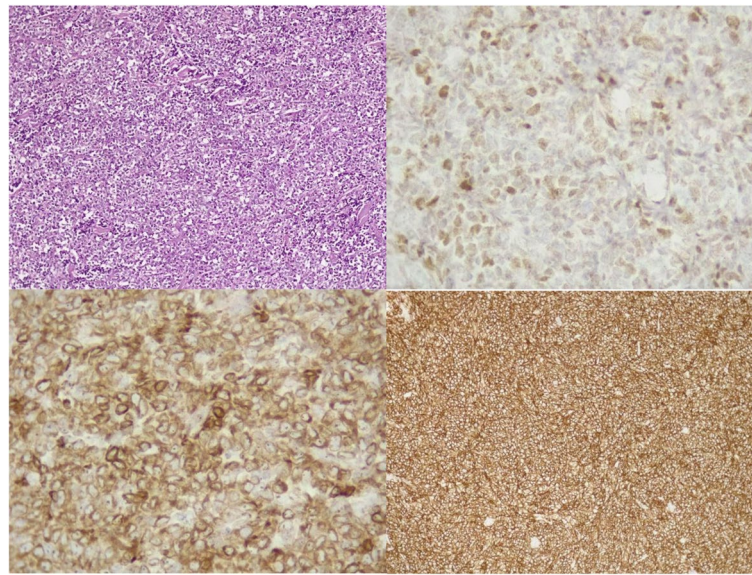


**Fig. 1** The patient presenting with a left-sided frontal bossing

2 months. Detailed history taking, physical examination, and screening lab data did not reveal significant findings. On CT scan, haziness and soft tissue density were observed on both frontal sinuses, contralateral sphenoid sinus and ethmoidal cells; right osteomeatal complex was obstructed (Fig. 2). The left frontal soft tissue mass, almost 3 cm × 3 cm in diameter, was subcutaneous; the cortical bone of the anterior table of the frontal bone was intact and not invaded. Fine needle aspiration of mass was undiagnostic, so an excisional biopsy was planned. Due to B symptoms, the possibility of malignant lesions was considered, and chest CT scan and liver function test were performed pre-operatively. There was not a significant finding. During endoscopic sinus assessment, we encountered polypoid tissue in paranasal sinuses, so samples were taken. Frontal boss was an un-encapsulated firm soft tissue mass, which was exposed through a direct brow lift incision performed just on the medial half of the brow. The overlying skin was intact; there was infiltration of subcutaneous tissue, but the pericranium was intact. Complete excisional biopsy was successfully conducted. On cellular examination, malignant lymphoproliferative disorder was reported for sinonasal specimen. Forehead mass was reported to be fibroadipose tissue involved by malignant lymphoproliferative disorder. Immunohistochemistry demonstrated diffuse high-grade B cell lymphoma (NOS type) with moderate positive reaction of Cmyc in about 55–60% of tumoral cells and mild to moderate positive reaction for cyclin-D1 in about 40–45% of



**Fig. 2** Coronal and axial views of paranasal sinus CT scan of the patient



**Fig. 3** Histopathologic examination; respectively from left to right: H & E staining\*10, immunohistochemistry study for Bcl6, Bcl2, and CD20 markers

tumoral cells in both samples (Fig. 3). Neoplastic cells revealed diffuse and strong positivity for CD-79 and CD-20. Markers of CD-10, BCL2, BCL6 (about 35–40%), and Ki-67 (75–80%) were positive. All other checked markers were negative. A subtype of diffuse large B cell lymphoma (DLBL) which is having Burkitt-like features (high grade B cell lymphoma with myc, BCL2 and or BCL6 rearrangement in WHO classification) was probable, which needed genetic and PCR analysis for confirmation. The patient was immediately referred. After thorough standard metastasis work-up including imaging studies by an oncologist colleague, the treatment regimen was initiated.

## Discussion

Despite the fact that bone involvement is not uncommon in some types of lymphoma, involvement of cranial vault is extremely rare. El Asri collected a systematic review of 36 articles including 38 patients with a chief complaint of subcutaneous scalp mass. There was a diversity in histologic subtype; however, DLBCL was the most frequent type [4].

Setta and coworkers reported an interesting case of primary cranial vault lymphoma, diagnosed accidentally in imaging studies following a minor head injury in a 75-year-old man. In magnetic resonance imaging, a subcutaneous frontal soft tissue mass was encountered at the level of head insult, in addition to brain parenchymal involvement without remarkable bone destruction. In tissue sampling, pathologic diagnosis was DLBCL. The

tumor invasion was hypothesized to be via long vessels [5].

It is rare to see DLBCL that involves soft tissue, especially skeletal muscles, and this presentation forms less than 1% of extra nodal NHL. Hatem reported a 79-year-old man with involvement of left triceps muscle, which is extremely unusual [6].

Derenzini and colleagues performed a monocentric analysis and also reviewed data in past 2 decades. They found 16 cases of NHLs presenting as soft tissue masses treated in their center during 1996–2011 and also 67 patients were selected from 5 series and 16 case reports through a systematic review of 1990–2011 time-frame. The most common subtype was reported to be DLBCL (50% cases in both groups). The researchers reported worse prognosis in soft tissue DLBCL as opposed to soft tissue indolent B cell lymphoma and also in comparison to historical data on DLBCL patients. Regarding inferior outcome in this subtype strategies of first-line therapy, intensification was proposed [7].

On 221 patients with head and neck lymphoma (193 cases of NHL and 28 HL), Stork reported that highly aggressive NHL and HL patients were significantly younger ( $p < 0.0001$ ). In this study group among NHL, 77 were indolent, 110 were aggressive, and 6 were highly aggressive. Different origins of NHL consisted of involvement of lymph nodes, tonsils, major salivary gland, sinonasal tract, and hypopharynx/larynx. Predictably, different presentations, especially in aggressive subtypes of NHL like Burkitt, and HL would make diagnostic challenges and it is recommended to

perform biopsies early in diagnostic measures whenever a head and neck mass is encountered [8].

Of 56 head and neck DLBCL patients reported by Kwak, in 52%, site of origin was Waldeyer's ring, 14% lymph nodes, 20% nasal cavities and paranasal sinuses, and 14% other sites as the submandibular gland, thyroid, and lacrimal sac [9].

A retrospective study which was conducted during 2004–2014 reported data on 110 nodal and extra nodal CD20+ DLBCL of head and neck area. Palatine tonsils were the most common extra nodal site (29.1%). Among cell origin markers (CD10, BCL6 and MUM1), MUM1 was strikingly correlated with extra nodal lesions ( $p=0.029$ ). According to the results, the authors believed the extra nodal DLBCL of head and neck area may have longer survival of solely nodal cases [10].

## Conclusion

Early diagnosis and prompt CHOP-based chemotherapy is crucial in this fast-growing tumors, so tissue sampling must be considered in any unusual soft tissue lumps in head and neck area even presenting as scalp mass lesions, although soft tissue masses are rare presentation of extra nodal DLBCL.

## Abbreviations

NHL: Non-Hodgkin lymphoma; DLBCL: Diffuse large B cell lymphoma; HL: Hodgkin lymphoma; WHO: World Health Organization; NK cell: Natural killer cell; NOS type: Not otherwise specified; CD: Cluster of differentiation; BCL: B cell lymphoma; MUM: Multiple myeloma oncogene; CHOP: Cyclophosphamide, doxorubicin hydrochloride (hydroxydaunorubicin), vincristine sulfate (Oncovin), and prednisone.

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## Authors' contributions

SN was the responsible attendant and surgeon and gathered the data on patient and was a major contributor in writing the manuscript. AM performed the histological examination of the specimen referred to pathology lab. FM was responsible resident and collected the required data. All authors read and approved the final manuscript.

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## Availability of data and materials

All data related to the patient are available to send at any time asked for.

## Declarations

## Ethics approval and consent to participate

Not applicable

## Consent for publication

Patient was informed about the broadcasting data and written informed consent was taken from the patient for publication including patient's anonymous details and images.

## Competing interests

The authors declare that they have no competing of interests.

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