

# Primary Extranodal Temporal Hodgkin Lymphoma: A Case Report and Review of the Literature

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## Abstract

Hodgkin's lymphoma (HL) is one of the few adult malignancies that most frequently presents with a progressive, painless enlargement of the peripheral lymph nodes. A primary extranodal presentation of HL, without lymph node involvement, is extremely rare.

The current study presents an unusual case on the extra nodal presentation of HL arising in a 63 year old woman, and reviews the relevant literature with particular emphasis on treatment.

The patient presented 13-months evolving mass in the temporal and frontal region of the face, the tumor has been diagnosed, based on radiologic findings, as a temporal sarcoma. It's the anatomopathological study of the operative specimen that revealed a Hodgkin lymphoma nodular sclerosis.

The patient received chemotherapy followed with local radiotherapy, and is currently in remission.

**Keywords:** Hodgkin; Temporal; Extranodal; Sarcoma

## Introduction

Classical HL is defined as a well established, proliferative neoplasm of the lymph nodes that is composed of mononuclear Hodgkin cells and multi nucleated Reed Sternberg (RS) cells in variable proportions, along with neutrophils, eosinophils, histiocytes, fibroblasts, collagen fibers, non neoplastic lymphocytes and plasma cells. The majority of patients commonly present with progressive painless enlargement of the lymph nodes, particularly around the cervical and supraclavicular lymph node regions [1,2].

Primitive extra nodal forms of HL are rare, accounting for <1% of all HL cases [1].

Diagnosis may fail to be suspected when the clinical and/or radiological tumoral syndrome mimics epithelial tumor or infection, but needs to be considered as treatment is specific [3].

The present study describes the case of a 63 years old female diagnosed with primary HL of the temporal muscle.

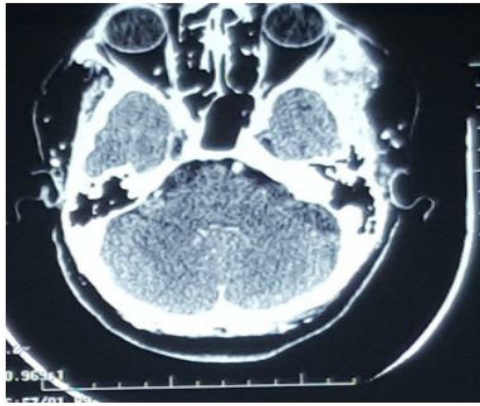
## Case report

A 63 years old women with no particular background consulted in our department for a 13 months' history of a left facial mass of the temporal region, chronic headaches weight loss and asthenia.

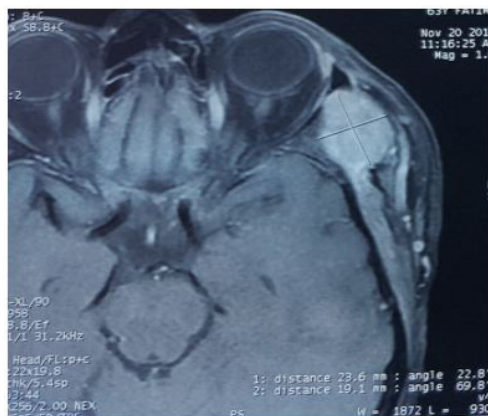
The physical examination showed a firm ovoid painless and non-pulsatile mass, measuring between 6 and 7 centimer of diameter fixed to the profound plan mobile to the superficial one with an intact overlying skin, localized in the frontal and temporal region of the face. No palpable lymph nodes or hepatosplenomegaly were noted.

Laboratory investigation didn't show any abnormalities.

A Computer Tomography of the face identified a malignant tumor infiltration of the left temporal muscle with irregular limits, and heterogeneous enhancement after the administration of intravenous gadolinium. It comes into contact with the zygomatic arch outside, the lateral wall of the orbit inside and the frontal bone, without bone lysis. A sarcomatous origin was the most suspected.



**Figures 1:** We completed the imaging exploration with a cerebral MRI which also showed a malignant tumor of the temporal region with a soft tissue sarcoma as a main suspected diagnosis



**Figures 2:** A complete excision of the mass was performed under general anesthesia. The mass was adherent to the temporal muscle. The temporal and frontal bone were intact



**Figures 3:** As per operative view of the temporal mass



**Figures 4:** The temporal bone after excision of the mass

The anatomopathological study of the operative specimen revealed a Hodgkin lymphoma nodular sclerosis: the pathological examination revealed that the tissue contained cells bearing the characteristic morphology of RS cells, with abundant cytoplasm and marked eosinophilic nucleoli, dispersed against a background of reactive inflammation. The immunohistochemistry analysis of the RS cell population showed positive staining for CD15 and CD30.

The patient was sent to the hematology department where she has been screened for serological tests (HIV, HBV, HCV, TPHA VDRL). A total body computed tomography (CT) scan did not detect other localizations.

The patient was prescribed four courses of Adriamycin, bleomycin, vinblastine and dacarbazine (ABVD) chemotherapy followed with a total body computed tomography scan evaluation. For socio economic reasons, the patient didn't receive a staging Pet scan.

The treatment was completed with local 30 Gy radiotherapy.

In the last physical examination (6 months after treatment), general condition of the patient was good and the patient was in remission, a CT scan was prescribed and revealed no signs of Hodgkin lymphoma.

## Discussion

Extranodal localizations as the primary site of Hodgkin lymphoma is unusual but has been reported in as many as 10%-15% of cases.

There are various types of extra nodal manifestations of HL. The gastrointestinal (GI) tract is the most frequent site of involvement by localized extranodal HD, with the stomach being the most common site followed by the small bowel, colon, and esophagus [4].

Other reported sites of primary extranodal HD include the lung, tongue, palate, chest wall, cheek, bone, thyroid, and central nervous system [1,5-11].

The case of a 22 years old female, described by Wei Luo et al revealed also an extranodal Hodgkin lymphoma with multifocal osseous localization [1].

In our study, we present a rare case of 63 years old female patient with a temporal mass mainly diagnosed, based on CT and MRI results, as a temporal sarcoma [Figure 1 and 2].

Misinterpretation of these malignancies may result in inappropriate treatment. In fact, although surgery plays a fundamental role in the management of benign and malignant lesions, HL treatment is based on immunopoly chemotherapy and/or radiotherapy [Figure 3 and 4].

Several studies suggested that the sensitivity and specificity of 18F-FDG PET-CT for the assessment of nodal and extranodal involvement were higher than those of standard contrast-enhanced CT (CE-CT) including Hodgkin and non-Hodgkin lymphoma patients. PET-CT has a significant advantage for the diagnosis of diffusely infiltrating organs without mass lesions or contrast enhancement compared to CE-CT [14].

Therapeutic groups are determined according to biologic factors (anemia, lymphopenia) and the extent of disease (number of lymph nodes or presence of bulky tumor, extranodal involvement, stage IV disease) [12].

Treatment is based on classifying patients as having limited (stages I and II, non-bulky) or advanced (stage III or IV) disease, with stage II bulky disease considered as limited or advanced disease based on histology and other prognostic factors (IPS or GHSG) [15].

The aim in the treatment of Hodgkin disease is to cure affected patients and to limit long-term therapeutic toxicity. In early stages of disease extended field irradiation is now replaced by short periods of chemotherapy followed by limited radiotherapy to decrease late sequelae [16].

The HD10 trial showed that in patients with early-stage Hodgkin's lymphoma and a favorable prognosis, treatment with two cycles of ABVD followed by 20 Gy of involved-field radiation therapy is as effective as, and less toxic than, four cycles of ABVD followed by 30 Gy of involved-field radiation therapy [17].

In our case, the patient presented a limited-stage disease with unfavorable risk factors, therefore she was prescribed four courses of Adriamycin, bleomycin, vinblastine and dacarbazine (ABVD) chemotherapy followed with local radiotherapy in the oncology and hematology department.

It is important to distinguish between extra nodal involvement representing a primary manifestation or dissemination of systemic disease, because the prognosis is much less favorable in systemic Hodgkin disease [9]. Our patient's case falls into the first category. In fact, she responded well to the treatment and showed no sign of recurrence.

## Conclusion

Extranodal Hodgkin lymphoma is a rare and an unusual entity. Its importance should be emphasized to avoid misdiagnosis that can change the prognosis of the disease.

When Hodgkin disease is diagnosed, extensive staging must be performed to determine whether extranodal involvement represents a primary manifestation or dissemination of systemic disease.

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