



Case Report

Primary Extranodal Extralymphatic Hodgkins Lymphoma: A Rare Case Report

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ABSTRACT

Introduction: Extranodal extra lymphatic Hodgkin's lymphoma is an extremely rare condition. Under rare circumstances, these tumors arise from tissue other than lymph nodes.

Case presentation: 22 year old male patient presented with swelling in the left temporal region since 3 months and in the left side of neck since 15 days. CT scan showed no intra cranial extension. Total excision of left temporal swelling with left cervical lymph node biopsy was done. Histopathology report is suggestive of primary extranodal soft tissue Hodgkin's lymphoma with involvement of left cervical lymph node. Immunohistochemistry showed polymorphous background amidst which are large, nucleolated Reed – Sternberg cells (R-S cells) which expresses CD- 30 and are immune negative for CD- 15, LCA, CD- 20 and CD- 3. Post operative period was uneventful. Patient is receiving chemotherapy with Adriamycin, Bleomycin, Vincristine and Decarbazine.

Discussion: Lymphomas are second most frequent after carcinomas affecting head and neck. From this 80 -90 % are Non Hodgkin's lymphoma and almost 4 % represent Hodgkin's lymphoma. Most of the HL involves lymph nodes. Extranodal extra lymphatic lymphoma have been used to describe the uncommon form of lymphoid malignancy in which there is neoplastic proliferation at sites other than expected native lymph nodes and lymphoid tissue. In this case lymphoma was in the left temporal swelling.

Conclusion: The aim of presenting this case is it's rarity. Biopsy of the lesion and immunohistochemistry are valuable to establish diagnosis. The need for awareness is stressed.

Key words: Extranodal extra lymphatic Hodgkin's lymphoma. Immunohistochemistry; rarity

INTRODUCTION

Hodgkin disease is usually almost entirely confined to the lymph nodes. Extranodal involvement is much less common in Hodgkin disease than in non-Hodgkin lymphoma. Extranodal involvement (except in the spleen and

thymus) indicates stage IV Hodgkin disease. Initial staging is crucial for demonstrating the presence of extranodal involvement, which will affect therapeutic decisions. Here we report a case of Hodgkins lymphoma in a 32 year male presented with temporal

swelling in view of its rarity along with favorable prognosis.

CASE REPORT

32 year old male patient presented with a swelling in the left temporal region since 3 months and in the left side of the neck since 15 days. Clinical examination was unremarkable. Local examination showed a swelling in the left temporal region, soft to firm in consistency, non reducible, non compressible approximately 7 x 5 cms. Skin over the swelling normal, also swelling on the left side of neck was noted approximately 3 x 1 cms, firm in consistency. Laboratory investigations were within normal limits. CT scan showed no intracranial extensions. CT abdomen

showed multiple enlarged paraaortic and paracaval lymph nodes with well defined heterogeneous retroperitoneal lesion on right side along the medial border of psoas muscle causing loss of fat planes with multiple small diffusely spread lesions in the splenic parenchyma suggestive of splenic infiltration.

Question mark (?) incision was taken on left fronto-temporal region, scalp flap elevated, temporalis muscle was elevated, subperiostially greyish white moderately vascular, granular mass noted involving inner surface of temporalis muscle, eroding the periosteum but not bone, extending from the inferior temporal line upto the zygoma on left side (Figure 1).

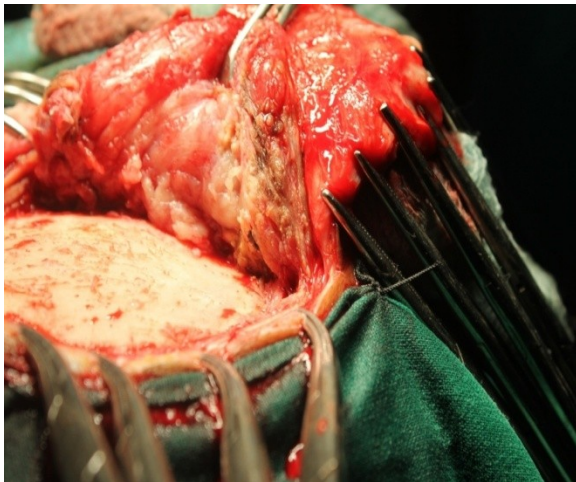


Figure 1 : Greyish white moderately vascular, granular mass noted involving inner surface of temporalis muscle.

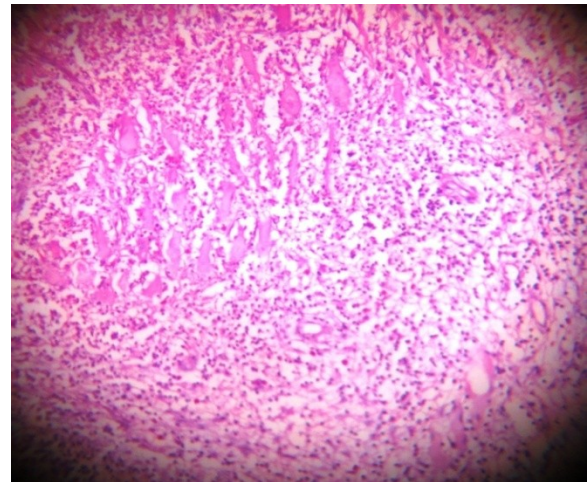


Figure 2 : Lymphoid background amidst which are large, nucleolated R-S cells.

Total excision of left temporal swelling with left cervical lymph node biopsy was done. Histopathology report is suggestive of primary extranodal soft tissue Hodgkin's lymphoma with involvement of left cervical lymph node (Figure 2 and 3).

Immunohistochemistry showed polymorphous lymphoid background amidst which are large, nucleolated R-S cells which expresses CD - 30 and are immunonegative

for CD - 15, LCA, CD - 20 and CD - 3. Post-operative patient received chemotherapy with Adriamycin, Bleomycin, Vincristine and Decarbazine (ABVD regime) Post-operative period was uneventful. Patient was followed on outpatient department without any complications post-operatively.

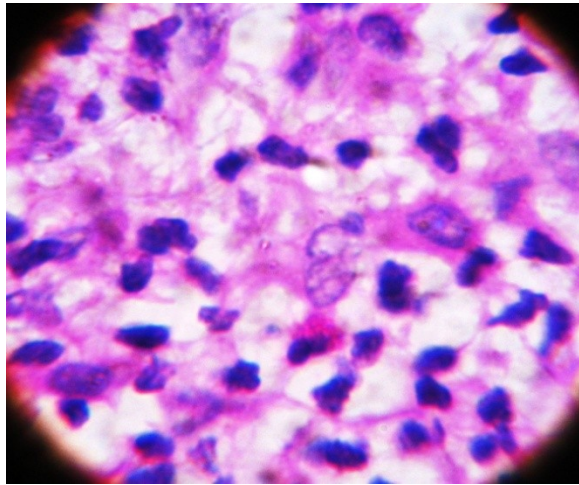


Figure 3 : Nucleolated R-S cells.

DISCUSSION

From the subset of malignancies affecting the head and neck region, lymphomas are the second most frequent after carcinomas. From those lymphomas, 80% to 90% are Non-Hodgkin lymphomas and about 4% represent Hodgkin lymphoma (HL).^(1,2) The incidence of HL is estimated to be 7400 new cases per year in the United States, accounting for approximately 30% of all lymphomas.⁽³⁾ As well known, most of the HL involves the lymph nodes. However, under some circumstances, those tumors arise from tissues other than the lymph nodes. Therefore, the term extranodal, extralymphatic lymphoma has been used to describe the uncommon form of lymphoid malignancy, in which there is neoplastic proliferation at sites other than the expected native lymph nodes and lymphoid tissue, respectively. Due to the difficulty in case definition, the frequency of this type of variation is not well established, nevertheless when sites rich in primary lymphoid tissue such as Waldeyer's ring and spleen are considered extranodal, extranodal lymphomas would represent 25–50% of all non-Hodgkin lymphomas and only 2–5% of classical Hodgkin lymphomas.⁽⁴⁾ In the case herein reported, the lymphoma was localized

in the temporal region eroding the underlying periosteum.

Hodgkin's disease is predominantly seen in male patients. Five subtypes of Hodgkin lymphoma are recognized: (1) nodular sclerosis, (2) mixed cellularity, (3) lymphocyte predominance, (4) lymphocyte rich, and (5) lymphocyte depletion. and mixed cellularity is its most frequent histological subtype.⁽⁵⁾

Hodgkin lymphoma encompasses a distinctive group of neoplasms that arise almost invariably in a single lymph node or chain of lymph nodes and spread characteristically in a stepwise fashion to the anatomically contiguous nodes. Clinical staging of Hodgkins Lymphoma is shown in Table 1.

Table 1: Clinical Staging of Hodgkin Lymphoma (Ann Arbor Classification)*.

Stage	Distribution of Disease
I	Involvement of a single lymph node region (I) or involvement of a single extralymphatic organ or tissue (I _E)
II	Involvement of two or more lymph node regions on the same side of the diaphragm alone (II) or with involvement of limited contiguous extralymphatic organs or tissue (II _E)
III	Involvement of lymph node regions on both sides of the diaphragm (III), which may include the spleen (III _S), limited contiguous extralymphatic organ or site (III _E), or both (III _{ES})
IV	Multiple or disseminated foci of involvement of one or more extralymphatic organs or tissues with or without lymphatic involvement

All stages are further divided on the basis of the absence (A) or presence (B) of the following systemic symptoms: significant fever, night sweats, unexplained loss of more than 10% of normal body weight.

Contiguous (E-stage) disease, which requires local radiation therapy, must be distinguished from stage IV disease, which is treated with chemotherapy alone or combined with general radiation therapy. Also, the extent of extranodal involvement

must be evaluated because it is considered prognostic.

The outlook after aggressive radiotherapy and chemotherapy for patients with this disease, including those with disseminated disease, is generally very good. With current modalities of therapy, the clinical stage is the most important prognostic indicator. The 5-year survival rate of patients with stage I-A or II-A disease is close to 100%. Even with advanced disease (stage IV-A or IV-B), the overall 5-year disease-free survival rate is around 60 to 70%.

With recent advances in the treatment of Hodgkin disease, the aim is now to cure affected patients and to limit long-term therapeutic toxicity. Consequently, therapeutic strategies are tailored according to initial prognostic factors. All prognostic scores established for Hodgkin disease take into account either the presence of stage IV disease or the number of extranodal sites.⁽⁶⁾ Therapeutic groups are determined according to biologic factors (eg, anemia, lymphopenia) and the extent of disease (eg, number of lymph nodes or presence of bulky tumor, extranodal involvement, stage IV disease). Stage IV disease is usually less responsive to radiation therapy and can be treated with chemotherapy if the number of cycles is sufficient.⁽⁷⁾ In contrast, patients with E-stage disease can be treated with less extensive chemotherapy combined with radiation therapy.⁽⁸⁾

CONCLUSION

The aim of presenting this case is its rarity. Biopsy of the lesion and immunohistochemistry are valuable to establish diagnosis. The need for awareness of Extranodal Extralymphatic Hodgkins Lymphoma is stressed.

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