

Non Hodgkin's lymphoma of the scalp - A case report

Dr. Rakul Nambiar K¹, Dr. Rona Joseph P², Dr. Unnikrishnan P³, Dr. Geetha N⁴

^{1,2,3}Senior Resident, Department of Medical Oncology, Regional Cancer Centre, Trivandrum 695011, India.

⁴Professor & Head, Department of Medical Oncology, Regional Cancer Centre, Trivandrum 695011, India.

*Corresponding author

Dr. Geetha N

Email: geenarayanan@yahoo.com

Abstract: Lymphoma is the second most common primary malignancy occurring in the head and neck with Diffuse large B cell lymphoma(DLBCL) being the most frequent type. Primary Non-Hodgkin's Lymphoma of the scalp is extremely rare. We are describing a case of the same in a 42 year old lady who presented with a diffuse swelling in the right forehead since 3 months of duration. Imaging showed soft tissue lesion arising from the scalp with diffuse hyperostosis of right parietal bone. Further investigations with CT scan chest, abdomen, and pelvis did not reveal any other evidence of systemic disease. Biopsy of the scalp mass was diagnostic of DLBCL scalp. She was treated with chemotherapy followed by radiation.

Keywords: Diffuse large B cell lymphoma, Non Hodgkin's lymphoma, scalp.

INTRODUCTION

Extranodal lymphomas constitute 20% to 30% of all non Hodgkin's lymphomas (NHL)[1,2]. The common sites involved are skin, stomach, brain, small intestine[3,4] and the most common histology is diffuse large B cell lymphoma (DLBCL). NHL arising from the scalp is rarely reported. We report the case of NHL of the scalp in a 42 year old woman.

CASE REPORT

A 42 year old lady presented with swelling on the right forehead of 3 months duration. She gave history of a similar swelling at this site 2 years ago for which an excision was done. Her ECOG performance score was 1. On examination, there was a 3x3 cm soft tissue swelling on right forehead. The swelling was soft, non-tender and non-fluctuant. There was no pallor, lymphadenopathy or organomegaly. Computed Tomography (CT) scan of the head revealed a soft tissue lesion arising from the scalp with diffuse hyperostosis of right parietal bone (Figure-1). An incisional biopsy of the lesion was done. Histopathological examination showed monotonous population of large mononuclear cells in sheets with scant cytoplasm, large irregular vesicular nucleus with clumped chromatin (Figure-2). On immunohistochemistry, the cells were positive for CD20, CD10 and bcl6 with MIB labeling index around 40%. The picture was diagnostic of DLBCL of scalp. Her hemoglobin was 12.5 g/dl, platelets were 4.5 lakhs/microl and total leucocyte con was 8800 / microl. LDH was 568 U/L. Her serum chemistries, CSF and bone marrow studies were normal. A CT scan of neck, thorax, abdomen and pelvis did not show any evidence of disease and she was staged as DLBCL of scalp -

stage 1AE. She was started on combination chemotherapy with Rituximab, cyclophosphamide, doxorubicin, vincristine and prednisolone (R-CHOP regimen). There was complete resolution of the tumor clinically after 2 cycles of chemotherapy. Radiological remission was documented after 6 cycles of chemotherapy. She was then consolidated with radiation.



Fig-1: CT brain - Diffuse hyperostosis of the right parietal bone with enhancing soft tissue component involving the scalp

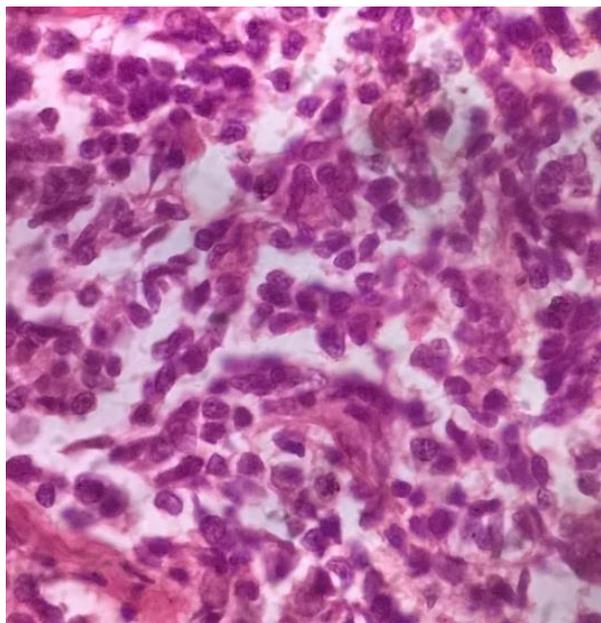


Fig-2: H&E stain(x100) showing monotonous population of large mononuclear cells in sheets with scant cytoplasm, large irregular vesicular nucleus

DISCUSSION

Lymphomas which originate outside the lymph nodes, spleen, and thymus are referred to as primary extranodal lymphomas. The common extranodal sites are the gastrointestinal tract, the head and neck, followed by the lung. In the head and neck region, sites involved are salivary glands, orbit, paranasal sinuses and thyroid gland. However, primary ENHL of the scalp is rare. Only few cases have been reported in the literature[5,6]. Most of cases are reported in middle-aged and elderly women.

NHL of the scalp usually runs an indolent course, presenting as a lesion on scalp, but eventually may progress to involve the underlying skull and dura[5,6]. Long standing lesions can present with focal neurological deficits. Histologically, all reported lymphomas of the scalp are of B-cell origin and the most common histological type is DLBCL. DLBCL characteristically express pan B-cell markers including CD 19, CD 20, CD 22, CD 79a and PAX 5. IHC was confirmatory of DLBCL in our patient.

Primary scalp lymphoma should be differentiated from lymphoma of cranial vault. Cranial vault lymphomas involve the pericranium, meninges, and subcutaneous tissues. Primary cranial vault lymphomas have intracranial extradural extension in addition to the scalp mass, however more than half of the patients report with a scalp mass rather than any neurological sign. The other clinical differences between the two include a shorter duration of symptomatology, early onset of focal neurological deficits, and extensive osteolytic lesions in primary NHL of the cranial vault[7,8].

Treatment for extranodal scalp NHL includes anthracycline based chemotherapy CHOP, and radiation. The prognosis is relatively good and overall survival is 12–14 years[9]. Any involvement of the cerebral structures by direct invasion or by leptomeningeal seeding indicates a less favorable prognosis.

In conclusion, it has become essential for physician to consider primary NHL in the scalp in the differentials for scalp swellings as early diagnosis and treatment can contribute to an increased life expectancy of these patients. A proper clinical evaluation, histology as well as immunohistochemical evaluation of biopsy specimen and radiological investigations may aid in early diagnosis and effective management.

REFERENCES

1. Zucca E, Roggero E, Bertoni F, Cavalli F; Primary extranodal non-Hodgkin's lymphomas. Part 1: Gastrointestinal, cutaneous and genitourinary lymphomas. *skin*. 1997;19:20.
2. Zucca E, Roggero E, Bertoni F, Cavalli F; Primary extranodal non-Hodgkin's lymphomas. Part 2: Head and neck, central nervous system and other less common sites. *Ann Oncol*, 1999; 10: 1023–1033.
3. Gospodarowicz MK, Ferry JA, Cavalli F; Unique aspects of primary extranodal lymphomas. In Mauch PM, Armitage JO, Harris NL et al.: *Non-Hodgkin's Lymphomas*. Philadelphia, PA: Lippincott Williams & Wilkins 2003; 685–707.
4. Thurnher MM, Rieger A, Kleibl-Poppy C, Schindler E; Malignant lymphoma of the cranial vault in an HIV-positive patient: imaging findings. *EurRadiol*. 2001; 11:1506–1509.
5. Madan R, Narula MK, Anand R, Gurtoo A; Primary cutaneous non-Hodgkin lymphoma of the scalp: A case report. *Indian J Radiol Imaging*. 2004;14:385–7
6. Martin J, Ramesh A, Kamaludeen M, Udhaya, Ganesh K, Martin JJ; Primary non-Hodgkin's lymphoma of the scalp and cranial vault. *Case Rep Neurol Med*. 2012; 2012: 616813.
7. Mongia S, Shukla D, Devi BI, Reddy TV; Primary cranial vault non-Hodgkin's lymphoma. *Neurol India*. 2003; 51:293–4.
8. Burg G, Dummer R, Kerl H; Classification of cutaneous lymphomas. *Dermatol Clin*. 1994; 12:213–7.
9. Abrey LE, Yahalom J, DeAngelis LM; Treatment for primary CNS lymphoma: the next step. *J Clin Oncol*. 2000; 18:3144–50