Original Research Article

Unusual presentation of non hodgkin's lymphoma in forearm

G M Vignesh Krishna[1*], K Aiswarya Chowdary[2], Praveen Kumar Vasanthraj[3], P M Venkata Sai[4]

[1,2]Junior Resident, [3]Assistant Professor, [4]HOD, Department of Radiology, Sri Ramachandra Medical College and Research Institute, Chennai, Tamil Nadu, INDIA.
Email: vickykrish09@yahoo.com, kodaliaiswarya@gmail.com, doctorvpk@gmail.com, hodvenkatasai@gmail.com

Abstract

Extra-nodal, extra-axial presentation of Non-Hodgkin's lymphoma is rare. We present an uncommon case of Non-Hodgkin's lymphoma in an elderly woman who presented with a swelling in the right forearm. Initially considered as a lipoma, patient returned after 3 months with increase in pain and swelling. Further imaging done revealed an aggressive infiltrative lesion involving proximal half of ulna and head of radius. The patient also had another scalp swelling which on ultrasound screening showed cortical irregularity. The patient was worked up and HPE revealed Non Hodgkin's lymphoma. **Conclusion:** Extra nodal non Hodgkin`s lymphoma are rare and early identification and prompt referral to oncologist would result in timely management of the patient.

Key Words: Extranodal Non Hodgkin's Lymphoma; Forearm swelling; NHL; Non Hodgkin's Lymphoma.

[*]**Address for Correspondence:**
Dr. G. M. Vignesh Krishna, Junior Resident, Department of Radiology, Sri Ramachandra Medical College and Research Institute, Chennai, Tamil Nadu, INDIA.
Email: vickykrish09@yahoo.com

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INTRODUCTION

Extra-nodal Non-Hodgkin's lymphoma outside the axial skeleton is rare. We came across an elderly woman who presented with swellings in the right forearm and scalp. After undergoing a series of imaging and histopathological correlation, we have confirmed it to be diffuse B cell non-Hodgkin's lymphoma.

CASE REPORT

A 63 year old postmenopausal woman with no associated co-morbidities presented to our hospital with complaints of painful swelling over right forearm for past 3 months. History and clinical examination revealed that she also had a non traumatic painless forehead swelling for past 6 months. The forearm swelling initially was painless and X-ray, at that time, revealed no significant bony abnormality (Figure 1). Based on examination findings, a provisional diagnosis of lipoma was made and patient was reassured with an advice on short term follow up. Patient returned to the surgery department after 3 months, with increase in size of the swelling and new onset of pain, which warranted further evaluation. In the mean time the forehead swelling also increased in size. Clinical examination of the right forearm revealed a diffuse oval shaped swelling in the ulnar aspect of right forearm which was soft in consistency. Skin over the swelling appears normal. Tenderness over the swelling was present. No obvious muscle wasting noted. Range of motion is free at elbow and wrist. Sensations were normal. Clinical examination of the forehead swelling revealed a 5 x 2 cm soft swelling predominantly over the right side of the forehead. Skin over the swelling appears normal. No palpable cervical nodes or any other nodes were noticed. At this juncture, the possibilities of primary or secondary neoplastic aetiology was considered and patient was subjected for further investigations. Routine blood investigations, urine analysis, and chest radiograph were done and found to be normal. Thyroid function test revealed Hypothyroidism. Serology for HIV was

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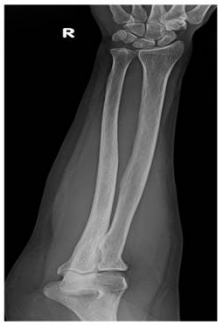
negative. Repeat X-ray of the right forearm (Figure 2) revealed linear displaced fracture in the shaft of the ulna with underlying bony erosions and moth eaten appearance of matrix. The soft tissue was also prominent at this region. Since the imaging features were more in favour of neoplastic aetiology, further evaluation with MRI was done.

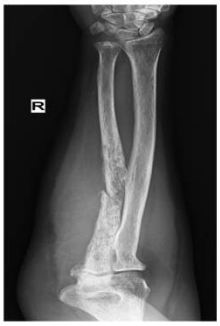
Figure 1



Initial X ray

Figure 1:

Figure 2

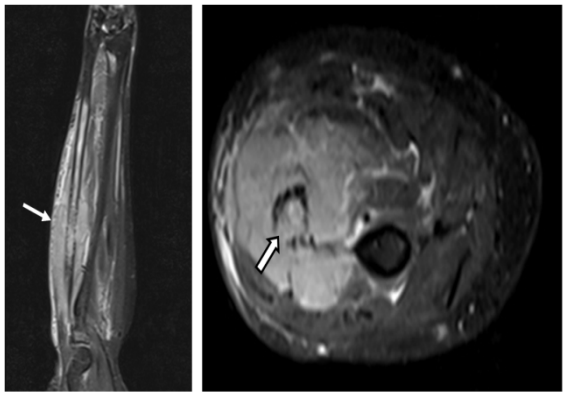


X-ray after 3 months

Figure 2:

MRI of right forearm revealed aggressive infiltrative lesion involving proximal and mid ulna with soft tissue extension encircling the bone (Figure 3). Similar signal intensities also noted in the head of radius. This soft tissue lesion is seen to extend in to the subcutaneous aspect on medial side and involving both flexor and extensor compartment, laterally up to the proximal radius with involvement of interosseous compartment. No extension seen into the joint cavity. On contrast administration there was heterogeneous enhancement in the abnormal areas. The lesion showed restriction on DWI. The ulnar artery was seen to be involved by this lesion. The proximal aspect of interosseous artery is seen to be abutted by this lesion. Radial artery was normal. The ulnar nerve and median nerve were not involved. Based on MR imaging, The possibility of Primary malignant or metastatic cause was considered.

Figure 3:



Coronal Axial

Figure 3:

In view to look at the nature of scalp swelling, Ultrasound was done which revealed a large soft tissue lesion with internal vascularity and underlying bony erosions. CT scan performed for the same confirmed pericranial soft tissue lesion in the frontal region predominantly on the right side with underlying bony infiltration, sclerotic changes and periosteal reactions (Figure 4). With a high probability of expansile metastatic lesions in differential diagnosis, a search for primary was done with USG neck, Sonomammogram and USG abdomen. Ultrasound of the neck revealed few nodules in both lobes of thyroid and isthmus. Single nodule noted in left breast during sonomammogram. USG abdomen was normal. Whole body PET CT was done to look for spread of the disease and to look for possible breast lesion as primary, which revealed significant uptake of tracer in the right forearm, skull, both thyroid, left parotid and left gluteal region. The confusion of primary thyroid neoplasm or a primary breast neoplasm could not be resolved at this point. After multidisciplinary team meeting, Fine needle aspiration cytology from both Thyroid and Breast nodules were done, which interestingly revealed a Lymphoproliferative lesion at both sites.

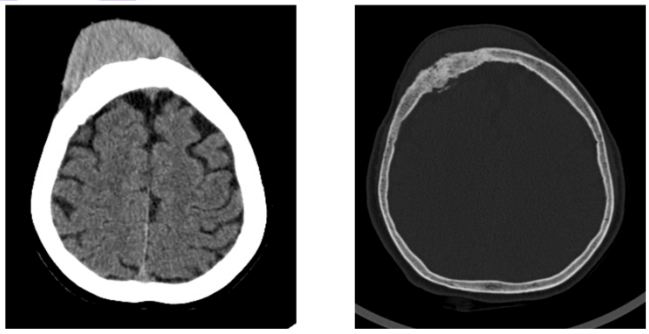
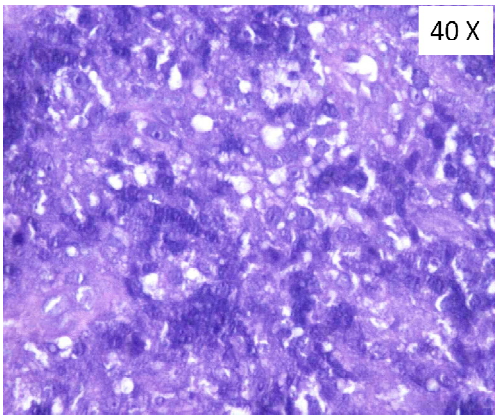


Figure 4

Figure 4:

An excision biopsy of the lesion in the right forearm was performed under local anesthesia and was subjected to histopathological examination. Microscopic features showed Bony trabeculae admixed with fragments of tissue showing crush artifact and tumour cells arranged in diffuse pattern. The small round blue cells are interspersed with occasional mitotic figures (2/10 HPF). Adjacent callus formation and reactive bone is also noted. By immunohistochemistry tumour cells are positive for Vimentin, CD45 and CD20. The background lymphocytes are positive for CD3. Tumour cells are negative for HMB45, CD117, CK and CD34.The histopathological features were suggestive of malignant lymphoproliferative lesion - Non-Hodgkin's lymphoma B-cell type (Figure 5). The patient was referred for further management where chemotherapy was started. Patient is ongoing treatment and so far has responded well.



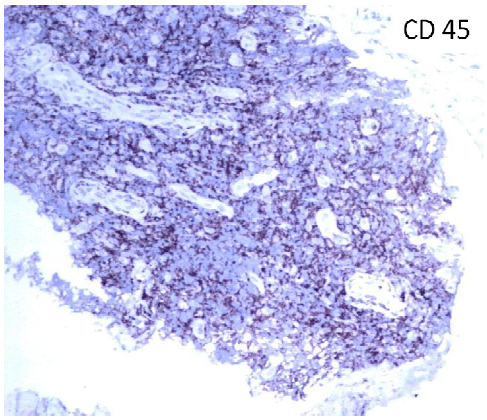


Figure 5:

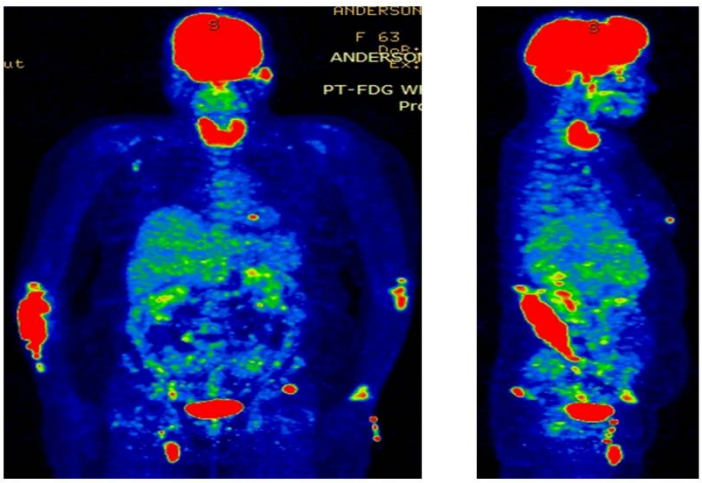


Figure 6

Figure 6:

DISCUSSION

Lymphomas arise from the mutation of lymphocyte progenitor cells. They are classified into two broad groups: Hodgkin's and non-Hodgkin's lymphoma. The incidence of Non-Hodgkin's lymphoma has steadily been on the rise whereas the incidence of Hodgkin's lymphoma has been relatively steady. The incidence rates in India for non-Hodgkin's lymphoma in men and women are 2.9/100,000 and 1.5/100,000, respectively[1]. The incidence within India is several times higher in urban areas compared to rural areas and the overall highest incidence of NHL in the world are reported from countries having a very high human development index suggesting that urban lifestyles and economic progress may be the cause of increased incidence. The median age of patients in India is 54yrs. Slight male predilection noted. Subtypes of non-hodgkin's lymphoma Figure 7

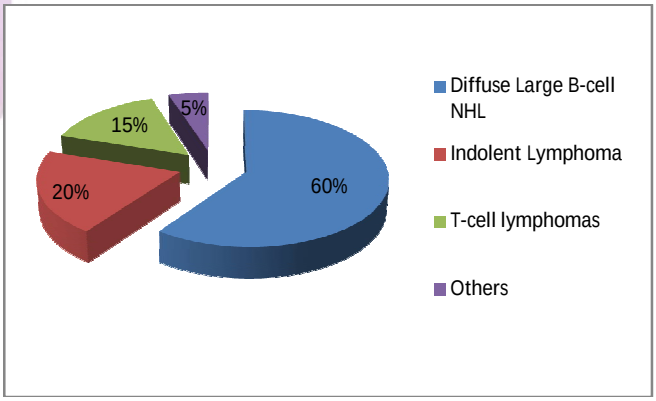


Figure 7: Percentage of nhl

80– 85% of all Non-Hodgkin's lymphoma in India are B-cell Non-Hodgkin's lymphoma. The most common subtype (60%) is the Diffuse large B-cell NHL followed by Indolent Lymphoma which contributes about 20%. 10–15% of cases are contributed by T-cell lymphomas. The three common subtypes of T-cell NHL seen in the Indian subcontinent are Peripheral T-cell lymphoma,

lymphoblastic lymphoma and anaplastic large-cell lymphoma.[2,3,4] About two-thirds of the NHL cases occurs in the lymph nodes and the remaining one third has been reported in the extranodal sites, including gastro intestinal tract, bone marrow, abdominal viscera and extranodal locations in head and neck[5].

The etiology of Non-Hodgkin's lymphoma is unknown, but the incidence of NHL is higher in immune-compromised conditions. B-cell lymphoma in immune-deficient patients is Epstein-Barr virus (EBV) positive when compared to sporadic type of lymphoma. Primary immunodeficiency syndromes are associated with an increased risk of developing lymphoma. Non-Hodgkin's lymphoma is the second most common neoplasm in HIV patients affecting 4% - 10% of the individuals with HIV. The risk of getting Non-Hodgkin's lymphoma is 60 folds greater in HIV-positive patients than in otherwise healthy persons[6]. Serological test for HIV disease was negative in our patient. The histological subtype in our patient was B-cell lymphoma. Prognosis of NHL depends on the extent of the disease, staging, histopathological subtype, and presence or absence of HIV disease. For example, diffuse large B-cell lymphoma and Burkitt's lymphoma among the B-cell lymphomas have an aggressive clinical course, whereas small lymphocytic lymphoma and follicular lymphoma have an indolent course among the B-cell lymphomas. Anaplastic T-cell lymphoma shows an aggressive behavior while mycosis fungoides have an indolent behavior among the T-cell lymphomas. Rapid progression of the lesion and increased incidence of opportunistic infections causes poor prognosis of Non-Hodgkin's lymphoma in HIV patients. The median survival time for HIV positive patients is severely compromised.

CONCLUSION

Extra nodal Non-Hodgkin's lymphomas are rare and early identification and prompt referral to oncologist would result in timely management of the patient.

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