Cutaneous, Lacrimal and Testicular Lymphomas; Case Series

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ABSTRACT

Background: One-fifth of the patients suffering from lymphoma had an extra-nodal presentation.

Objectives: We aimed to provide our experience with three cases of cutaneous, lacrimal, and testicular lymphomas.

Patients and methods: We provided three cases with extra-nodal lymphoma arising from the lacrimal gland, the subcutaneous tissue, and testis.

Results: The first case was a 42-year-old lady who presented with relapsed diffuse large B-cell lymphoma (DLBCL) in the form of subcutaneous nodules within both thighs. The second case was 72-year-old lady who presented with an orbital mass. The mass was proven to be NHL arising from the orbit with regional lymphadenopathy. The third case had advanced NHL with testicular and renal involvement. Graphical illustrations of their PET-CT scans were included.

Conclusion: Extra-nodal presentation of NHL is a rare but serious condition. We presented three cases with lacrimal, subcutaneous, and testicular presentation.

Keywords: Lymphoma, Non-Hodgkin's lymphoma, Hodgkin's lymphoma, Extra nodal lymphoma.

INTRODUCTION

Lymphoma is a complex haematological malignancy that arises from lymph nodes. These diseases are usually present as nodal diseases with supra and infradiaphragmatic presentation. In this piece of art, we present our experience with three cases of extra-nodal presentation originating from the orbit, testes, and subcutaneous tissue ^(1,2).

PATIENTS AND METHODS

Study design and patients: This is a case series of cases with a rare form of extra-nodallymphoma.

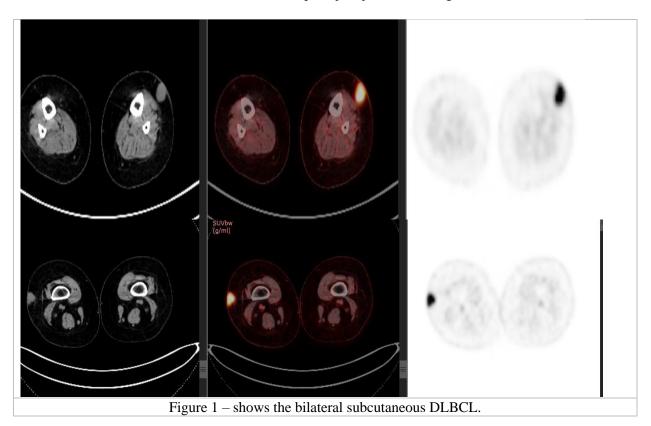
Ethical consent: The Academic and Ethical Committee, Sohag University approved the study. Every patient signed an informed written consent for acceptance of using his data in the study. This work has been carried out by the Code of Ethics of the World Medical Association (Declaration of Helsinki) for studies involving humans.

CASE 1

A 42-year-old female patient with NHL, DLBCL, was diagnosed in 2017. The patient received chemotherapy in 2018 and was under follow-up. In 2020 she developed subcutaneous nodules at the right thigh. An excisional biopsy was done, revealing NHL. 8 mci of Fluorine 18 FDG was given IV, and glucose level was ensured to be less than 90 mg/dl. Multi-detector tomography examination was done without injection of IV contrast.

Images of CT and corresponding functional PET images are displayed in axial, coronal and sagittal planes. Reference liver SUV was 3. Reference mediastinal blood pool SUV was 2.4. The scan showed low-grade FDG avid sub-centimetric cervical, mediastinal, and pelvic lymph nodes. There was an increased FDG uptake in the subcutaneous soft tissue lesion at the right thigh and the left leg with SUV max 7.5, 7.4 respectively, and Deauville score was 4 (Figure 1).

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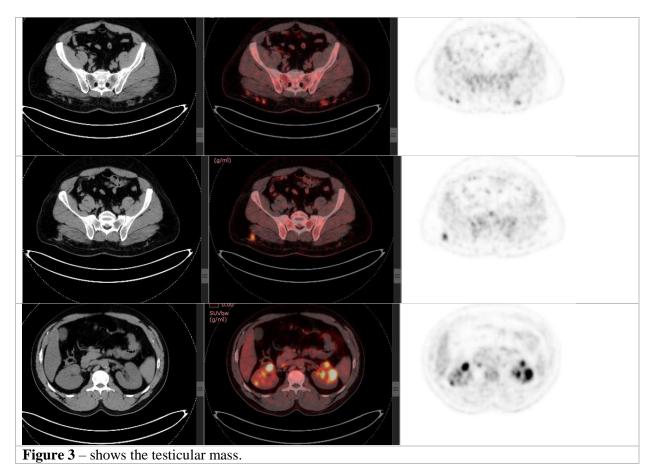
CASE 2

A 72-year-old female patient presented with left orbital swelling and submandibular swelling. CT head and neck revealed a left lacrimal gland mass displaying the eye globe. The mass measured 2.3x3 cm and it encroached on the left superior and lateral rectus muscle. There was also a circumferential thickening of the nasopharynx and proximal oropharynx with bilateral submandibular, submental, intra-parotid, and deep cervical lymph nodes; the largest of them was 22x14 mm. Tru-cut biopsy revealed NHL, B-cell type. She received chemotherapy then PET/CT was requested for assessment of treatment response. 8 mci of Fluorine 18 FDG was given IV, glucose level was ensured to be less than 90 mg/dl. Multi-detector tomography examination was done without injection of IV contrast. Reference liver SUV was 2. PET/CT revealed a left lacrimal gland soft tissue mass lesion of 3x2 cm with SUV max 10.6 involving the left lateral rectus muscle—nasopharyngeal thickening of 1.5 cm with SUV max 8.3. Bilateral tonsillar FDG uptake was more at the left side with SUV max 7.3. Bilateral intraparotid lymph nodes were with SUV max 4.7. Bilateral cervical lymph nodes were 1.2x 0.7 cm with SUV max 2.9. Mediastinal lymph nodes and left hilar lymph nodes were with SUV max 2.7 (Figure 2).



CASE 3

The third case was a middle-aged male with non-Hodgkin lymphoma. He received induction chemotherapy then found to have a progressive disease within the scrotum. There was a scrotal enlargement. Scrotal ultrasound and Doppler scan showed multifocal infiltration by lymphomatous tissue. PET-CT was done and it showed diffuse lymphomatous infiltration of the nodal chain above and below the diaphragm. Also, there was fluorine 18 avidity in the left testis, and both renal cortices as shown in Figure 3.



DISCUSSION

Classically, the non-Hodgkin lymphoma comes in the form of nodal disease. The most affected nodal groups are the cervical and the mediastinal nodes. However, it is not uncommon to see extra-nodalpresentation of the disease. Roughly one-third of the advanced non-Hodgkin lymphoma had some sort of extra-nodal presentation. The stomach, skin and lungs were the most affected organs. Diagnosis of extra-nodal lymphoma follows the same principle as nodal disease. Excisional biopsy is necessary to achieve proper diagnosis and disease classification ^(1,2).

Positron emission tomography/computed tomography depends on two imaging techniques: PET and CT. PET relies on the use of positron emitters radiopharmaceuticals such as FDG. The annihilation process of the positions results in the release of two highenergy photons that can be detected by specialized detectors ⁽³⁾. Using a CT scan could help in the localization process of the annihilation event. A commonly used

positron emitter is FDG. It could differentiate between normal and malignant tissue depending on the differential uptake ^(3,4).

Multidetector PET-CT was provided to be better and more accurate in the detection of extra-nodallesions. It allows for accurate FDG localization without compromising the time of acquisition ⁽⁵⁾.

Some reports pointed out that diffuse large B cell lymphoma was more prone to express extra-nodal infiltration compared to other types of lymphomas. The most involved site was the bone marrow at a rate of 28.7% ⁽⁶⁻⁸⁾. The sensitivity of PET-CT to detect extra-nodal lymphoma differs according to the site of the extra-nodal involvement. The sensitivity and specificity for the detection of splenic involvement were 98% and 94% respectively. However, these numbers drop to 93.7% and 96% respectively for bone marrow involvement. Furthermore, the sensitivity and specificity of positron emission tomography for non-splenic and non-bone marrow extra-nodal organs were 94% and 96.2% respectively ⁽⁷⁾.

It is well known that extra-nodal involvement is usually part of advanced disease. Roughly two-thirds of extra-nodal lymphoma are present in stage IV disease. Unfortunately, extra-nodal involvement is a worse prognosis and is linked to poor survival ^(1,2,9). In our study, all the cases were advanced stages of the disease, and the extra-nodal presentation was part of the heavy disease burden. Also, all of our cases were relapsed rather than de-novo cases.

In our study, we presented three cases with extranodal lymphoma. The first case was a lady with relapsed diffuse large B-cell lymphoma. The patient was treated and enjoyed a disease-free survival of 3 years before it relapsed. The relapse of the disease comes in the form of subcutaneous infiltration of both her thighs. The diagnosis was confirmed by a biopsy of such lesions. The second case was a lady who had a presentation of orbital mass on the left side. PET-CT showed diffuse infiltration of the orbital muscles along with cervical lymphadenopathy. Biopsy from the lesion proved the diagnosis as non-Hodgkin lymphoma. The third was a relapsed DLBCL with testicular and renal involvement. Our experience supports the already published data by some of the Egyptian colleagues; Alnouby et al. and El-Haddad et al. (10,11) in 2018 and 2015, respectively.

CONCLUSION

Extra-nodal presentation of non-Hodgkin lymphoma is not an uncommon presentation of the disease. We reported our experience with three cases of extra-nodal lymphoma arising from the skin, the orbit, and the testis.

- **Limitation:** This case series was limited to drawing any definite conclusion.
- **Role of funding:** The study was fully funded by the Sohag University Hospital and the Sohag Faculty of Medicine.
- **Conflict of Interest:** The authors had nothing to declare.

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