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Case Report

An Usual Presentation of Extranodal Natural Killer (NK) T-Cell Lymphoma

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Abstract

Background: Lymphoma is a spectrum of blood diseases affecting lymphocyte cells in the body and ultimately the immune system. Many classifications has been identified to further divide the disease into sub types according to the main cells affected. Extranodal NK T-Cell Lymphoma is a rare and aggressive form of lymphoma. **Case Presentation:** We report a patient who presented mainly complaining of a painful thigh mass limiting his daily activity for the past three months. He was admitted and investigated and was found to have orbital, preseptal, scrotal and thigh masses with maculpaplur rash and scattered lesions all over his body. Biopsy from Orbital and thigh mass showed NK T-cell lymphoma. **Conclusion:** Lymphoma has many forms with wide range of clinical presentations amongst patients that need to be recognized by physicians early, especially rare forms that are not encountered as frequently.

Keywords: Lymphoma; T-Cell; Extranodal; Mass; Rash

Case Presentation

This is an 80 Years old male, Saudi and Living in Riyadh. A chronic smoker of 30 cigarettes per day for 50 years. He is known with Diabetes mellitus and hypertension on medications with good compliance. Also with Ischemic heart disease with RBBB and history of unstable angina (Inferior Myocardial Infarction) on Aspirin and Diltiazem. The patient gave a history of Brucellosis infection that was treated 6 years ago. He also has Benign Prostatic Hyperplasia (BPH) on Omnic following with urology outpatient department since 2009. He had a past surgical history of Haemorrhoidectomy only. The patient presented to the emergency department complaining mainly of a painful ulcerating thigh mass. The mass first appeared three months ago in the left thigh as a subcutaneous lesion and increased in size with time with skin ulcerating and pain in the last week. He was also complaining of maculopapular rash over his eyes, chest, abdomen, upper limbs and lower limbs. This was his second presentation to our hospital complaining of the rash and scattered lesions. At that time, he was assessed in emergency department and sent to dermatology clinic as possible infected mosquito bite lesions. Also With two episodes of documented fever at home, decreased appetite, unintentional weight loss of 15 Kgs in three months and recent change of voice. On examination, the patient was conscious, alert, well oriented to time, place and person. He was not in pain or distress. BMI was 25. Bilateral eyelid swelling with maculopapular lesions (Figure 1). Scattered maculapapular rash with different sized subcutaneous nodules over bilateral upper limbs, lower limbs, chest and abdomen (Figures 2 to 7). Abdomen was soft and lax, no tenderness and no organomegaly with enlarged inguinal lymph nodes bilaterally, more on the right side. There was 6 x 10 cm fungating mass at the posterior lateral aspect of the left thigh with eroding edges and covered with slough tissue that bleeds easily to light touch. (Figures 8 and 9). There was no fouly smell or discharge from the mass itself and no similar swellings seen elsewhere in the body. Blood investigations including liver profile, renal profile and electrolytes were within normal. Other blood work up during his hospital stay are summarized in Table 1.

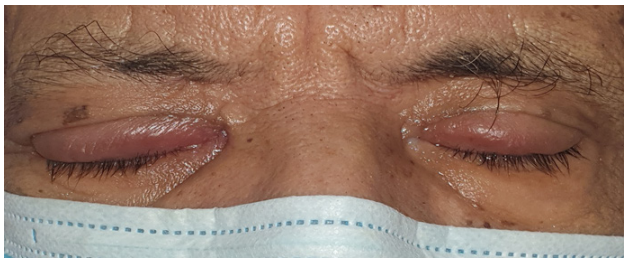
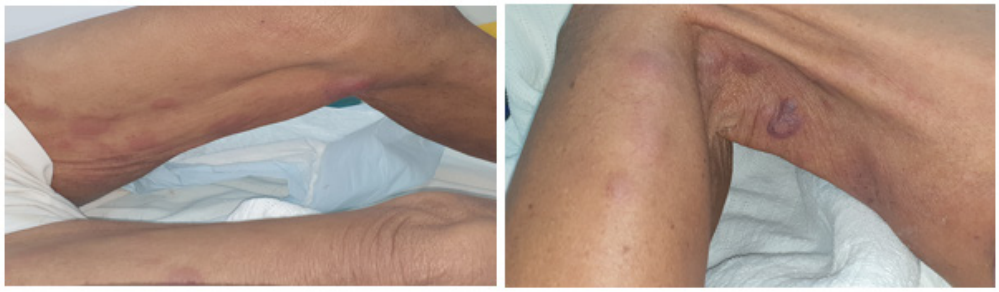
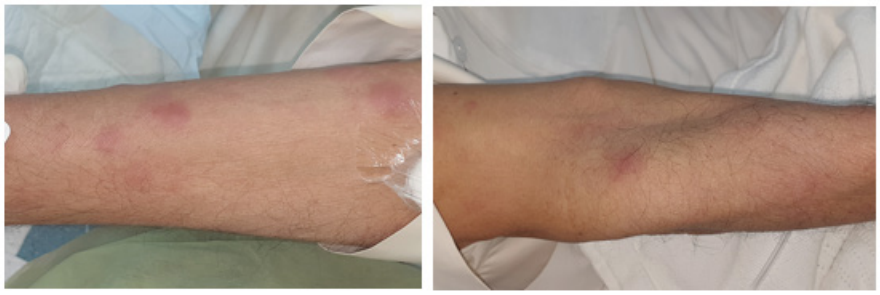


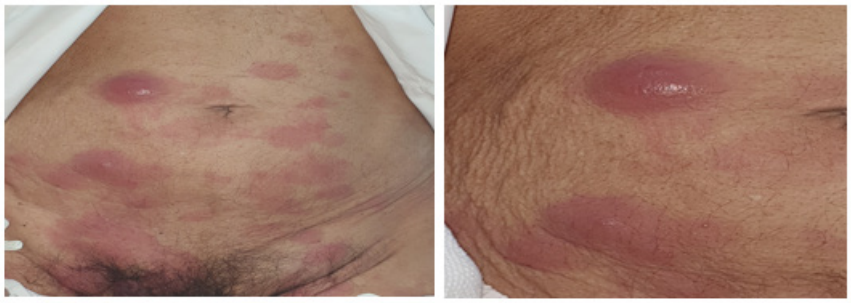
Figure 1: Bilateral eyelid swelling.



Figures 2 and 3: Right and Left Lower Limb Lesions.



Figures 4 and 5: Right and Left Upper limbs Lesions.



Figures 6 and 7: Abdominal and Inguinal Lesions.

Figures 8 and 9: Left Thigh Mass.

	Result	Reference Range Unit
WBC	3	4.0 - 11.0 10^9/l
Lymph Absolute	1	1.5 - 4.0 10^9/l
Neutrophils Absolute	1.3	1.8 - 7.5 10^9/l
HGB	13.3	12.5 - 18.0 g/dl
HCT	0.4	0.380 - 0.540 l/l
MCV	93	75 - 95 fl
PLT	220	150 - 450 10^9/l
INR	1.1	0.9 - 1.3
Sodium	137	136 - 145 mmol/L
Potassium	4.4	3.5 - 5.1 mmol/L
Albumin	37	35 - 52 g/L
NT-proBNP	469 pg/mL	
Troponin T	0.017 f ng/mL	
ESR	7	0 - 15 mm/hr
C-Reactive Protein	10	0 - 6 mg/L
AFP	2.1	0.0 - 5.8 kIU/L
CEA	1.4	0.0 - 3.4 ug/L
CA-125	8	0 - 35 U/ml
CA 19-9	9	0 - 34 U/ml
CA 15-3	15	0 - 25 U/ml
PSA	0.401	< 4.400 ug/L

HIV1/2 Ab Scr	Negative
HBsAg Result	Negative
HCV Antibody	Negative
Brucella Serology	Brucella Abs Negative

Table 1: Blood work up during his hospital stay are summarized.

Diagnostic radiological investigations conducted during his hospital stay included computerized tomography of head and neck (Illustration 1 and 2) which showed a 2.7 x 2 cm soft tissue mass in right vocal cord extending superiorly till the base of epiglottis causing significant airway narrowing and obliteration of right pyriform fossa. No associated bony lesions. Another 2.1 x 1.6 cm soft tissue mass seen along the right pre-septal region showing intraortibal extension through medial canthus causing lateral deviation of right globe. There were Small subcutaneous nodule in left lower cheek and a small 1.2 x 0.8 cm right level Ib lymph node. Computerized tomography of the chest showed multiple axillary lymph nodes, the largest on the right was 1.2 cm. Small sub centimetre supraclavicular lymph nodes. Findings suggestive of aspiration in both lower lung lobes. Computerized tomography of the abdomen and pelvis showed normal liver, gallbladder, spleen, pancreas, adrenal glands, small bowel and large bowel. Bilateral simple renal cysts with normal prostate and seminal vesicles. There was a 4.5x5.7 cm subcutaneous mass lesion in left inguinal region with multiple enlarged left inguinal and external iliac lymph nodes (the largest 1.4x1.5 cm). Prominent right inguinal lymph nodes (Illustration 3 and 4). MRI thigh (Illustration 5-9) showed an exophytic soft tissue mass lesion measuring 5.4x9.2x8.5 cm in AP, transverse and CC dimensions, arising from the posterolateral aspect of the proximal left thigh. It was showing intermediate signal intensity in T1 and T2 with heterogeneous enhancement in post gadolinium sequences. Another soft tissue lesion seen at bilateral scrotal cavities measuring 5.3x8.2x7.3cm in right scrotum/testicle with hydroceles. Another subcutaneous lesion measuring 3.6x5.6x5.6 cm anterior to the left inguinal region with almost similar signal characteristics. There were multiple variable sizes bone lesions seen at bilateral femora, biggest in left femur, 3.8 cm long, which show low signal intensity in T1 and high signal intensity in T2 with intense enhancement in post gadolinium sequence. Modified Barium Swallow showed mild pharyngeal dysphagia. And aspiration with thin liquid only.

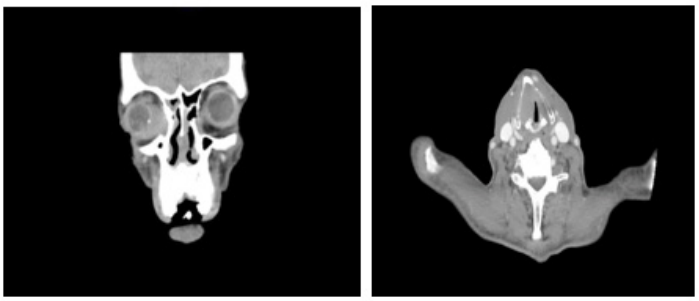


Illustration 1 and 2: CT head and Neck showing Right orbital and Right Vocal cord mass.

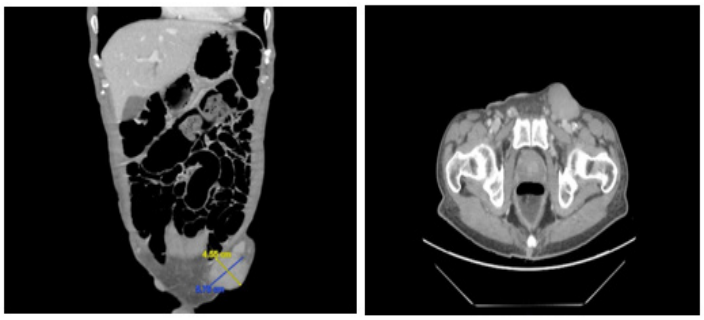
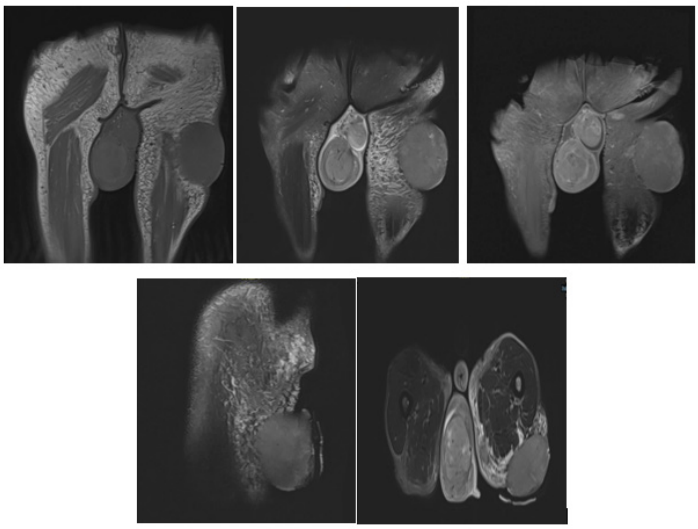


Illustration 3 and 4: CT Abdominal showing Left Inguinal mass.



Illustrations 5-9: Thigh and Scrotal Mass.

Punch biopsy was taken from the right thigh mass and the right forearm lesion, which confirmed the diagnosis of T-Cell Lymphoma. Sections showed infiltration of the dermis by atypical lymphocytes showing high mitotic rate. The cells were positive for CD3 and negative for CD20. Ki67 is positive in 80% of the cells. Additional immunohistochemical study results showed the tumor cells are positive for CD3, CD7, CD56 (weak), and C-myc (focal), while negative for CD5, CD8, CD30, Alk-1, EMA, CD23, CD38, MUM1, PAX-5, CD10, Bcl-2, cyclin D1, and Bcl-6. The T-cell receptor alpha/delta locus at chromosome 14q11.2 rearrangements was not detected

Thigh tissue culture	Serratia Marcescens Isolated after enrichment culture Only Sensitive to Ciprofloxacin
Thigh Tissue Fungal culture	No growth after 8 weeks
Thigh biopsy ZN Stain	No acid-fast bacilli seen on direct smear
AFB CULTURE	Quantiferone Result Negative cps/ mL

Right Orbital Lesion Biopsy also showed T-cell lymphoma with histological sections showing respiratory-type and squamous epithelium with underlying diffuse infiltration of monomorphic medium to large lymphoid tumor cells with irregular pleomorphic vesicular chromatin and increased mitotic figures. The tumor cells were positive for CD3, CD7, CD56, CD43, and negative for CD20, CD5, and CD8. Ki67 proliferation index is positive in about 85% of tumor cells. The tumor cells were positive for GATA3, granzyme B and perforin (weak). Representing extranodal NK T-cell Lymphoma.

The patient was referred to ENT surgery for the change of voice and the findings of vocal cord mass. There was no intervention from their side and he was started on Dexamethasone. He was assessed by Swallowing team and was kept on puree diet. There was surgical intervention from urology regarding the scrotal mass and Hematology took over the care of the patient as a case of Extranodal T-Cell Lymphoma most probably originating from the thigh mass with secondary metastasis.

Discussion

Lymphocytes are white blood cells present in our blood that help fight infections. T-Lymphocyte represent 80% of circulating lymphocytes. They are made in the bone marrow but they mature fully in the thymus and are involved in Cell-mediated immunity. B-Lymphocytes are manufactured in bone marrow and mature there before but they spread in the lymphatic system. These cells account for 20% of circulating lymphocytes in the blood and are involved in humoral or antibody-mediated immunity [1]. Lymphoma is a type of cancer that affects lymphocytes causing these cells to grow out of control at an abnormal rate affecting innate and adaptive immunity. It can be classified according to morphology, anatomical site affected, age at presentation or type of cell affected [2]. There are two main types of lymphoma according to the type of involved cell. Hodgkins Lymphoma (HL) represent 10% of cases and is a B-Cell Lymphoma affecting Reed-Sternberg cells. Non-Hodgkin Lymphoma (NHL) represent 90% of cases and have almost 30 subtype classified according to morphology and immunophenotype features. T -Cell lymphoma account for 7% of all NHL and don't have a known genetic alteration [2,3]. Nodal lymphoma is lymphoma primarily originating in lymph nodes but can spread to extra nodal site. Primary Extranodal lymphoma definition in literature is controversial but the most acceptable definition is lymphoma with no or minor nodal involvement associated with a clinically dominant extra nodal component [4]. If the patient presented with different masses as different locations, it might be different to identify where the lymphoma primary originated and thus proper staging which follows the Ann Arbor classification of Lymphoma [3,5]. Primary Extranodal lymphoma account for 33% of NHL and is very rare in HL [3]. Almost 50% of all extra nodal NHL were histologically of Large B-Cell Lymphoma while Mature T and Natural Kill Lymphoma account for 15% of NHL [6].

Extranodal Lymphoma it can occur at any organ but from literature review, the most common site was found to be the gastrointestinal track followed by skin, bone and brain [4,6,7]. Orbital lymphoma account for 50% of primary orbital malignancy in adults, 2% of all adult lymphoma and almost 15% of all extra nodal lymphoma [8,9]. It is mostly extraconal in location and 97% are B-Cell NHL with the most common subtype being Extra nodal Marginal Zone B-cell Lymphoma (EMZL) at 59%. NHL in known to be the most common testicular malignancy in adult patients. Primary testicular NHL account for 9% of testicular neoplasms and 2% of all NHL [10]. Primary Bone Lymphomas (PBL) account for 7% of all adult bone cancer and 5% of all primary extra nodal NHLs and the femur is the most common site of involvement. Cutaneous T-Cell Lymphoma (CTCL) account for 2% of all NHLs and 11% of T-Cell lymphoma. It is usually confined to the skin and soft tissue appearing as skin patches, plaques, rash or most commonly a tumour. These lesions and rash are often misdiagnosed as eczema or dermatitis and sometime insects bites [11]. Prognosis and treatment options differs according to the patient presentation, histopathological tumour differentiation and staging.

Conclusion

Extranodal T Cell/ NK Lymphoma is an aggressive form of lymphoma which could present late or be misdiagnosed in the early stages thus affecting prognosis. There is no designated referenced classification in the literature due to its overlapping and difficulty in identifying primary origin.

Consent for Publication

A written informed consent was obtained from the patient for the use of pictures attached in this review.

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