ORIGINAL RESEARCH PAPER

Oncology

KEY WORDS: Non Hodgkin Lymphoma, Edema, Ulceration, Lower limb

A RARE CASE OF PRIMARY CUTANEOUS DIFFUSE LARGE B-CELL LYMPHOMA, LEG TYPE: A CASE REPORT

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ABSTRACT

Edema with ulceration is one of the common presentation encountered in the clinical setting. The main aim is to identify the cause of edema with ulceration and treat the underlying cause. The edema and ulceration can be unilateral or bilateral. The main cause of unilateral leg edema with ulceration is mainly due to venous insufficiency (deep vein thrombosis) and rarely because of tumor. Presence of unilateral limb edema with ulceration but without inguinal lymphadenopathy is extremely rare. In this paper, we are reporting a very rare case of Non Hodgkin Lymphoma (NHL) of lower limb with ulceration but no lymphadenopathy 1, 2

BACKGROUND

Lower limb edema with ulceration is one of frequent symptom to be encountered in primary care setting[3]. The common differential diagnosis of unilateral limb edema with ulceration is deep vein thrombosis (DVT)[4]. Some of the differentials are trauma, unilateral limb surgery[5], radiation exposure and/or rarely tumor. Lower limb edema with ulceration can be classified depending upon their duration of onset, acute or chronic. Such a presentation is very rare in tumor. Thus, we are reporting a very rare case of unilateral lower limb edema with multiple ulcerations.

CASE PRESENTATION

A 65 years old female presented to our hospital with chief complaint of swelling in her lower limb for 8 months and multiple ulcerations in lower limb for 6 months. She was apparently well 8 months back when she developed swelling in her lower limb, which was insidious in onset, unilateral, non-progressive, not relived with any medication, any postural changes and not aggravated with any sort of movement.

She also complains of ulcer in her lower limb for 6 months which was insidious in onset, progressive, non-bleeding and non-tender.

She also complains of pain for past 3 months in her right lower limb which was insidious in onset, sharp, radiating to back and no aggreavating or relieving factors.

She had no history of fever, night sweats, fatigue, dyspnoea, exercise intolerance orthopnoea. She had no history of any trauma, exposure to radiation.

She is non-alcoholic, non-smoker and no history of any other substance abuse.

She denied any similar family history.

PHYSICAL EXAMINATION

She had well-built and her general condition was fair. Her vitals were stable. Her abdomen was soft. Her cardiovascular examination were within limit. Her respiratory examination were within limits.

LOCAL EXAMINATION

Inspection –
Mild right sided leg edema, non-pitting. There was around 5*5 cm multiple ulceroproliferative growth on right medial side of her knee joint, non-bleeding and no discharge was present.

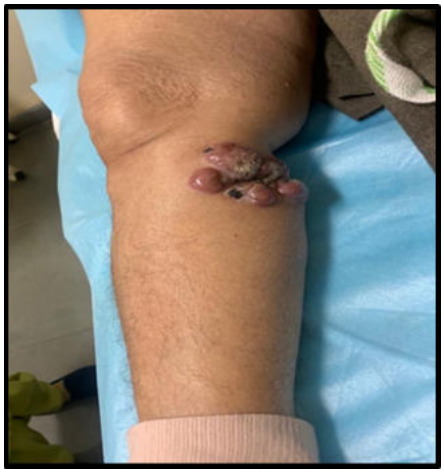

Figure 1. *Showing Mild Leg Edema With Multiple Small Cauliflower Like Ulcerations In Medial Aspect Of Right Thigh.*

Palpation –
Mild right sided leg edema, non-pitting, non-tender and warm on touch. Around 5*5 cm multiple cauliflower like ulceroproliferative growth on medial side of right leg, non-bleeding, non-pus discharging and non-tender.

INVESTIGATIONS

She was advised routine blood test like (complete blood count, liver function test, renal function test, random blood sugar level), chest x-ray PA view, ultrasound right lower limb to rule out DVT.

Her compression venous ultrasound showed no sign of any venous insufficiency.

Later PET scan was advised. PET CT showed –
1. Multiple FDG avid (SUV 16.2), nodular cutaneous lesions are seen involving the skin and subcutaneous soft tissue along the medial aspect of right knee measuring 6.6*2.4*5.8 cm suggestive of malignant cutaneous involvement.
2. No other hypermetabolic active lesion seen elsewhere in body.

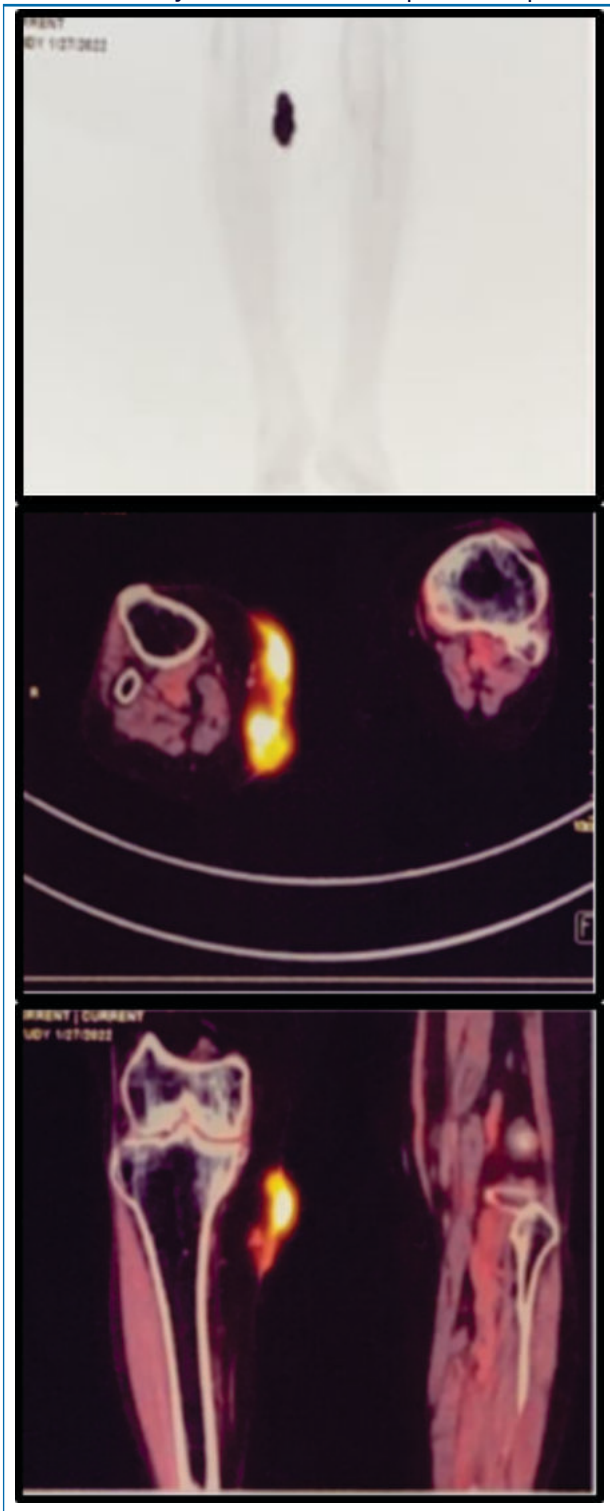

Figure 2,3,4. *Showing hypermetabolic lesion in medial aspect of right knee joint.*

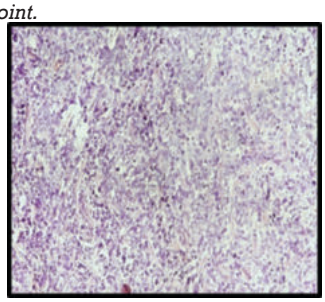

Figure 5. *Interspersed among the tumor cells are reactive lymphoid cells and small blood vessels.*

Histopathology Examination-

Excisional biopsy from cutaneous lesions was done and it was found to be Non Hodgkin Lymphoma – DLBCL type.

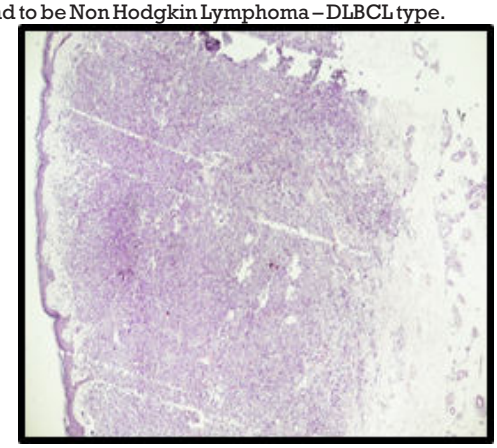

Figure 6. *Epidermis Lined Tissue Showing In Dermis Arranged In Diffuse Sheets. The Tumor Is Reaching Upto The* Subcutis

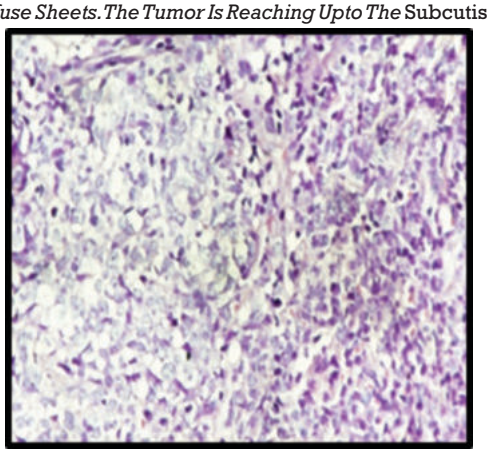

Figure 7. *The tumor cells are large cells with moderate to marked nuclear pleomorphism having coarse nuclear chromatin, conspicuous nucleoli and scant cytoplasm. Interspersed among the tumor cells are reactive lymphoid cells. Brisk mitosis also noted.*

Immunohistochemistry –

CD 20 – Negative

Differential Diagnosis–

Differential diagnosis in this case involving local and systemic cause of unilateral lower limb edema with ulcerations are DVT and some other rare differentials are – trauma, exposure to radiation, hypoalbuminemia and/or tumor.

Treatment

The patient was planned for 6 cycles of CHOP chemotherapy (injection Cyclophosphamide, Adriamycin, Oncovirin, Prednisolone. As, immunohistochemistry showed CD 20 negative so, there is no role of Rituximab.

DISCUSSION

Edema can be classified into acute or chronic depending on duration of onset. Acute edema is one which occur within 72 hrs of onset and chronic edema is more than 72 hrs.

Unilateral edema with ulcerations is mainly due to DVT. The other rare etiology including tumor, trauma or exposure to some radiation.

The history and physical examination play a very crucial role in diagnostic evaluation of unilateral limb edema with leg ulcers.

The key questions in history are –duration of edema, painless or painful, pitting or non-pitting, any history of any systemic disease including any trauma or radiation exposure. For ulceration the key points include whether acute or chronic, bleeding or non-bleeding, any exposure to trauma, travel history or long standing history.

For local examination the key points to rule out edema is whether pitting or non-pitting, to look for varicose vein, any skin changes, any other ulcers.

It is important to look for any lymphadenopathy like poplliteal, inguinal and tibial lymph nodes.

A review of literature, however reveals that unilateral lower limb edema with ulcerations is one of manifestation of occult malignancy.

Hawkins et al[6] found 10 cases of limb edema which is secondary to lymphoma. He found that among 10 cases, 7 cases were presented with unilateral leg edema as only presenting symptom. It was found that these patients were not responded to treatment of DVT like leg elevation, compression stocking etc as these were misdiagnosed. Later, it was found that lymphedema with leg ulceration is rare finding of lymphoma.

Another report of 35 cases of secondary lymphedema secondary to lymphoma[7].

So unilateral or bilateral leg edema with ulceration is one of diagnostic challenge.

So, proper history of presenting ulcer, history of B-symptom, proper physical examination like examination like examination of lymphadenopathy – inguinal, popliteal and tibial lymph nodes. PET scan is considered for disease assessment. Excisional biopsy with immunohistochemistry is standard of diagnosis.

CONCLUSION

The conclusion of the case is lymphedema with leg ulcerations could be secondary to occult malignancy. So, adequate history, physical examination and radiological investigations is must. The Primary cutaneous diffuse large B-cell lymphoma, leg type is extremely rare in incidence and aggressive in nature[8,9].

In such findings, it is important to rule out venous insufficiency.

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