An intriguing case of cutaneous noduloulcerative lesion in the vicinity of genitalia

Sir,

A 41-year-old married man presented with an asymptomatic ulcerated mass over the right inner aspect of the thigh for 3 weeks. It started as papular lesions that rapidly enlarged to form coalescing nodules with surface ulceration within 2 weeks. It was associated with swelling of the right leg. He was diagnosed with donovanosis at another center and received oral doxycycline for a week without any improvement. He developed a painless scrotal swelling for the past 2 days. He had neither systemic complaints nor, a history suggestive of high-risk sexual behavior.

Cutaneous examination revealed well-circumscribed, ulcerated plaque with few satellites such as erythematous and infiltrated nodules [Figure 1]. There was stony hard induration of the underlying and surrounding skin with a local rise of temperature. Bilateral inguinal, left cervical and submandibular lymph nodes were enlarged and nontender.

His hematological and biochemical parameters were within normal range. Routine blood analysis revealed anemia and leukocytosis. Histopathological examination revealed a dermal diffuse infiltrate with sheets of small-to-intermediate abnormal lymphoid cells with focal epidermotropism [Figures 2 and 3]. Immunohistochemistry of tumor cells was positive for CD30 (strong and equal intensity staining at the cell membrane and Golgi region), LCA, and MUM-1 and negative for CK, CD3, CD5, CD20, CD138, PAX-5, ALK, and EMA. Ki67 index was >80%. No abnormal cells were seen on bone marrow aspiration. Fluorodeoxyglucose positron emission tomography scan revealed multiple liver, lung spleen, and bone metastases. A diagnosis of Stage IV metastatic Non-Hodgkin's lymphoma; anaplastic large cell lymphoma (ALCL)-anaplastic lymphoma kinase (ALK)-negative subtype was made.

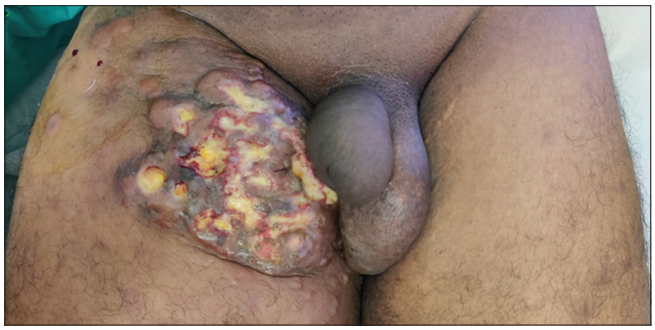

Figure 1: Infiltrated plaque (6 cm × 6 cm size) with confluent ulcers with yellow slough and areas of hyperpigmentation; surrounding satellite erythematous, infiltrated nodules (0.5–2.5 cm in diameter)

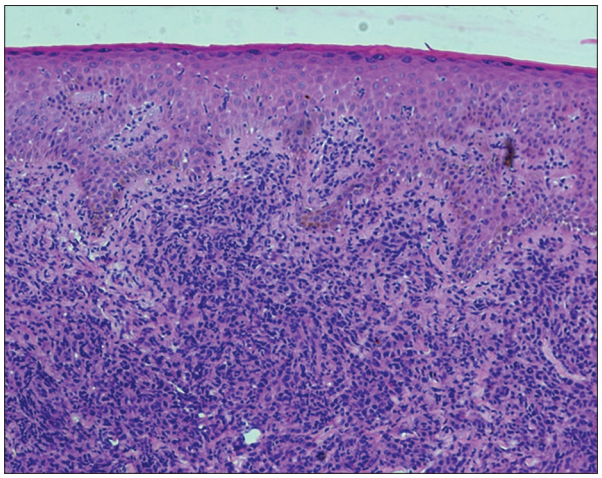

Figure 2: Focal epidermotropism, dermis shows an infiltrating neoplasm composed of diffuse sheets of abnormal lymphoid cells

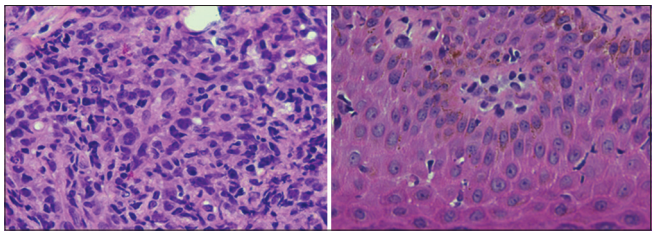

Figure 3: Intermediate to large-sized cells with oval and round nuclei with multiple peripherally placed nucleoli, few centroblasts, and immunoblasts, few with nuclear angulations, scant cytoplasm, and numerous mitotic figures

ALCLs (CD30 positive) may be classified into primary cutaneous ALCL (PC-ALCL), systemic ALK-positive, and ALK-negative ALCL.[1] It is very important for clinicians to delineate the origin of ALCL as the management and prognosis are different for PC-ALCL and the systemic variants with cutaneous involvement.[2] PC- ALCL is the second most common type of cutaneous T-cell lymphoma with a skin-limited malignancy, extracutaneous dissemination occurring mainly to lymph nodes in up to 10% of cases.[3]

Systemic ALCL presents with lymphadenopathy and is often associated with extra nodal disease; cutaneous involvement has been reported in 20% of cases. Most patients present with advanced (stage III-IV) disease, B symptoms, and often with generalized skin lesions.[2,4] However, skin lesions were localized in our patient and he had no systemic symptoms. Hence, a diagnosis of donovanosis was considered initially. Donovanosis uncommonly presents with extragenital lesions in <10% of the cases where the diagnosis may be missed when the disease is confined to the extra genital site.[5]

Around 16%–21% of cutaneous metastases are discovered before the underlying primary tumor. They have varied clinical presentations mimicking inflammatory and benign skin conditions.[6] Hence, a high index of clinical suspicion and a thorough workup including immunostaining is crucial in arriving at a diagnosis and initiating timely treatment. Although a morphological and immunophenotypic overlap exists, short duration of complaints and systemic organ involvement at presentation favored the diagnosis of ALK-negative ALCL to PC-ALCL in our patient.[1,4]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

Anusha Potula, M. Deepak Nayak[1], Pallavi Hegde, Deepti Jaiswal, Raghavendra Rao

Departments of Dermatology and Venereology and [1]Pathology, Kasturba Medical College, Manipal, Karnataka, India

Address for correspondence:
Dr. Anusha Potula,
Department of Dermatology and Venereology, Kasturba Medical College, Manipal - 576 104, Karnataka, India.
E-mail: anusha.potula@gmail.com

References

1. Montes-Mojarro IA, Steinhilber J, Bonzheim I, Quintanilla-Martinez L, Fend F. The pathological spectrum of systemic anaplastic large cell lymphoma (ALCL). Cancers (Basel) 2018;10:107.
2. Feldman A, Gaulard P, Pileri SA, Willemze R. Secondary cutaneous involvement in T-cell Lymphomas and Leukemias. In: Elder DE, Massi D, Scolyer RA, Willemze R, editors. WHO Classification of Skin Tumours. 4th ed. France: IARC; 2018. p. 254.
3. Feldman A, Gaulard P, Pileri SA, Willemze R. Primary cutaneous CD30+ lymphoproliferative disorders. In: David E, Elder, Massi D, Scolyer RA, Willemze R, editors. WHO Classification of Skin Tumours. 4th ed. France: IARC; 2018. p. 238-39.
4. Feldman AL, Harris NL, Stein H, Campo E, Kinney MC, Jaffe ES *et al*. Anaplastic large cell lymphoma, ALK-negative. In: Steven H. Swerdlow, Campo E, Harris NL, Jaffe ES, Pileri SA, Stein H *et al*. WHO Classification of tumours of haematopoietic and lymphoid tissue, Revised 4th ed. France: IARC;2017. p. 418-421.
5. O'Farrell N. Donovanosis. Sex Transm Infect 2002;78:452-57.
6. Choate EA, Nobori A, Worswick S. Cutaneous metastasis of internal tumors. Dermatol Clin 2019;37:545-54.

Access this article online

Quick Response Code:

Website: www.ijstd.org

DOI: 10.4103/ijstd.ijstd_106_22

How to cite this article: Potula A, Nayak MD, Hegde P, Jaiswal D, Rao R. An intriguing case of cutaneous noduloulcerative lesion in the vicinity of genitalia. Indian J Sex Transm Dis 2023;44:100-1.

Submitted: 30-Oct-2022 **Revised:** 03-Jan-2023
Accepted: 19-Jan-2023 **Published:** 06-Jun-2023