Primary Cutaneous Large B Cell Lymphoma Masquerading as Lupus Vulgaris

[1]Pooja Gupta, [1]Shruti Sharma, [2]V Ramesh, [3]Aradhana Bhargava

Departments of [1]Pathology, [2] Dermatology, [3] Microbiology

National Institute of Pathology- ICMR, VMMC and Safdarjung Hospital, New Delhi

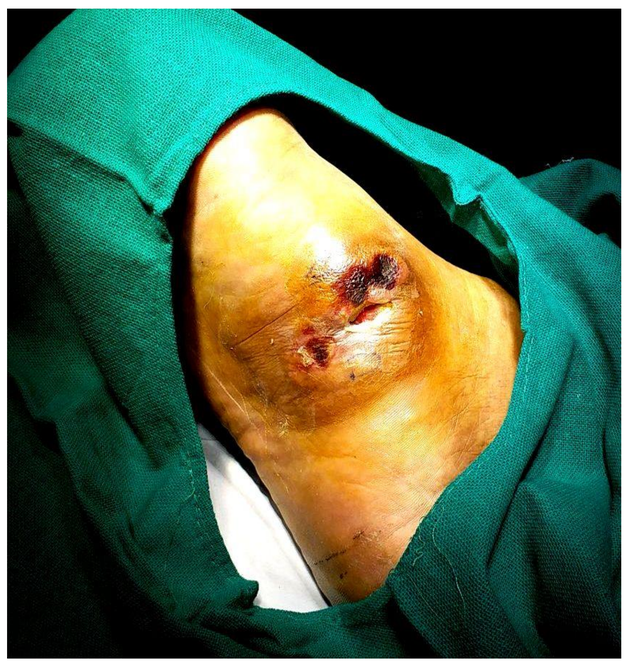
POSTER ID 62

Introduction

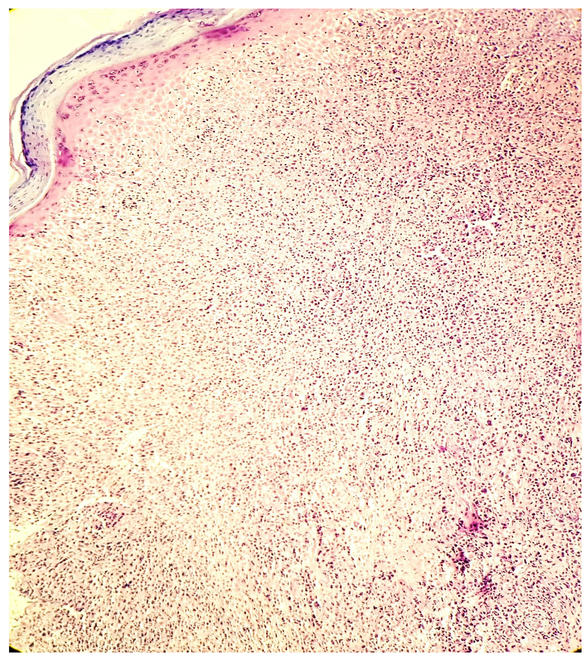
- Primary cutaneous large B-cell lymphoma, leg type (PLBCL, LT) is a rare and aggressive cutaneous neoplasm.
- It predominantly affects elderly women, with lower limb (leg) being the most common site of presentation.
- The overall prognosis is poor compared to other cutaneous B cell lymphomas.

Case Presentation

- A 47-year-old male presented with history of swelling on medial aspect of the left foot for 1.5 months.
- Examination revealed a nodular plaque measuring 7 X 7 cm, firm in consistency, and with ill defined margins. An ulcer was noted in the centre of this lesion.
- The initial working diagnosis was lupus vulgaris.



Clinical photograph showing ill defined nodular plaque with ulceration on medial aspect of left foot



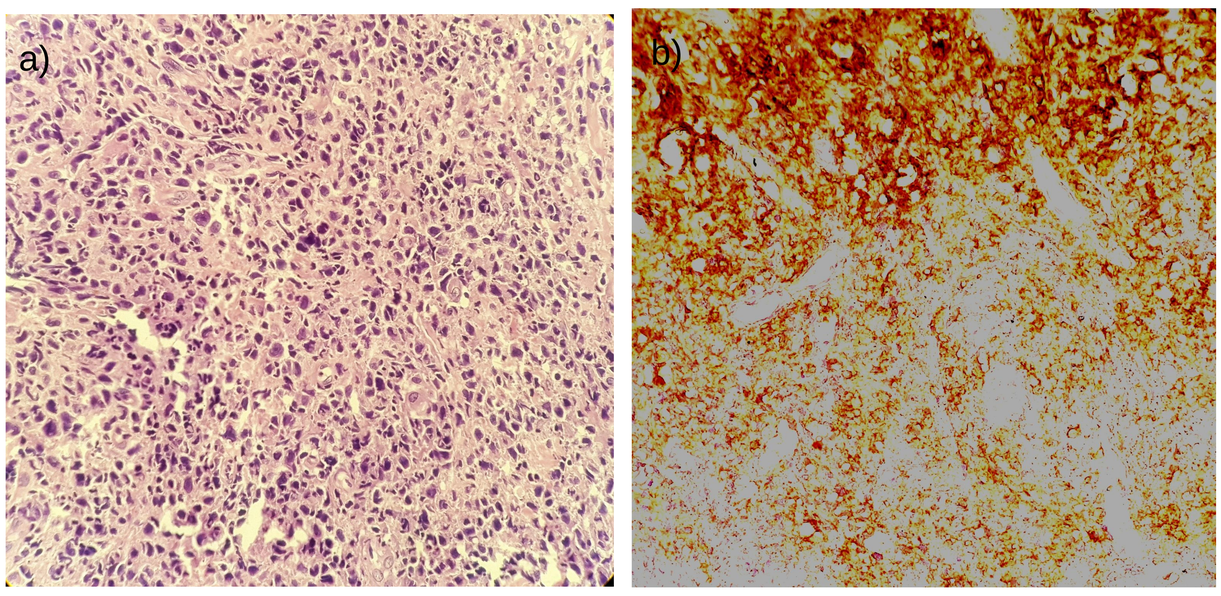
Microphotograph showing normal epidermis with dermal infiltration by lymphoid cells (H & E, 40X)

Investigations

- Complete blood count, liver and kidney function tests were unremarkable. An incision biopsy was done, which revealed mildy acanthotic epidermis. Entire dermis showed sheets of atypical cells. Cells showed moderate nuclear atypia, high nuclear-cytoplasmic ratio and prominent nucleoli.
- Immunohistochemistry revealed diffuse CD20 and MUM1 positivity, with very high Ki-67 proliferation index (50%). The cells were negative for CD3, CD68, CD5, cyclin D1, CD10 and CK.
- Clinical and radiological examination did not reveal any primary malignancy or enlarged lymph nodes.
- Bone marrow aspiration and biopsy were normal.

Diagnosis and treatment outcome

- A rare diagnosis of primary cutaneous large B cell lymphoma, leg type was made and patient started on chemotherapy.
- Unfortunately, patient died in June 2019, about one year after the index presentation



Microphotograph showing dermal infiltration by atypical lymphoid cells, 400x a) H & E b) CD 20

Discussion

- Cutaneous lymphomas are uncommon neoplasms, with an approximate annual incidence of 0.5 to 1 per 100,000.
- Primary cutaneous B-cell lymphomas are a heterogeneous group of mature B-cells neoplasms with tropism for the skin.
- Primary cutaneous large B-cell lymphoma, leg type is the most aggressive form of this neoplasm.
- Exact histogenesis of primary cutaneous large B-cell lymphoma, leg type is unclear but it is hypothesized to begin as reactive inflammatory lymphoproliferative processes due to chronic antigenic stimulation .
- Clinical manifestations include single or multiple patches, plaques and non-ulcerated nodules, with firm consistency.
- Histology is characterized by a nodular or diffuse, often sharply demarcated lymphoid infiltrate, located predominantly in the dermis.
- The study of molecular rearrangements of genes encoding leg heavy chains (IGH) is useful to differentiate PCBCL from pseudolymphoma.
- Treatment includes CHOP regimens associated with rituximab with or without radiotherapy.

References

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CONCLUSIONS

- PCLBCL, LT is a rare and aggressive neoplasm which may masquerade clinically as lupus vulgaris.
- A detailed histopathological and immunohistochemical analysis helps in achieving the correct diagnosis and initiating appropriate management