AN UNUSUAL CASE OF EBV POSITIVE LYMPHADENOPATHY AND CUTANEOUS NODULES

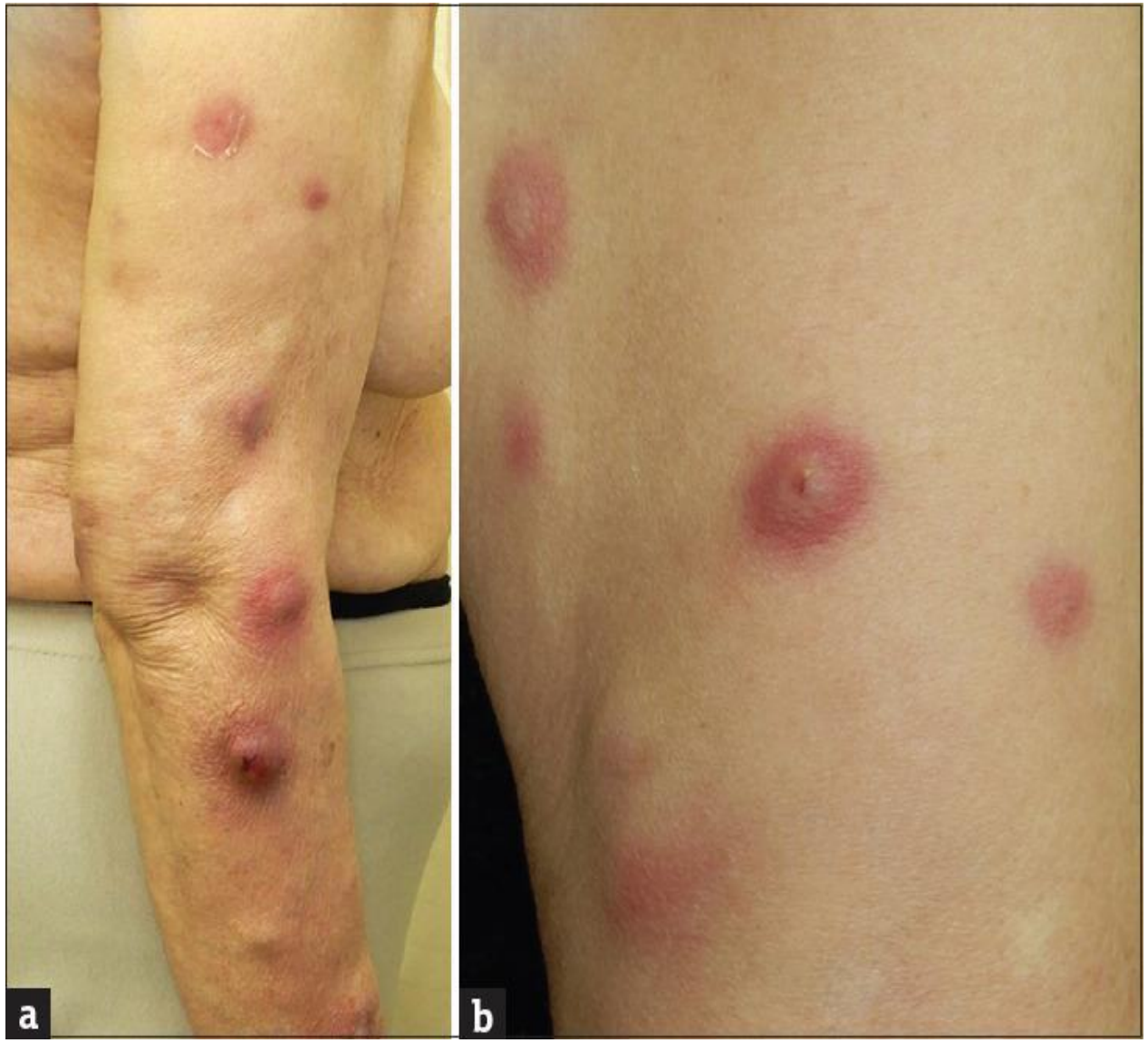
- Lymphomatoid granulomatosis (LYG) is a rare Epstein–Barr virus (EBV) associated B-cell lymphoproliferative disorder.
- It is characterized by vasocentric, angiodestructive and diverse cellular infiltration.LYG involves the lungs, central nervous system, skin and kidneys.
- LYG is intricately linked to host immune function, and its pathogenesis results from defective immune surveillance, mainly in CD8+ cytotoxic T cells, against EBV-infected B cells.
- Pulmonary lesions are observed in >90% of patients with LYG, followed by the central nervous system (40%), skin (34%), kidney (19%) and liver (17%).
- In 10–15% of cases, skin lesions may precede pulmonary lesion development.
- Lymphadenopathy in LYG is typically rare, and other disorders should be considered.

HISTORY

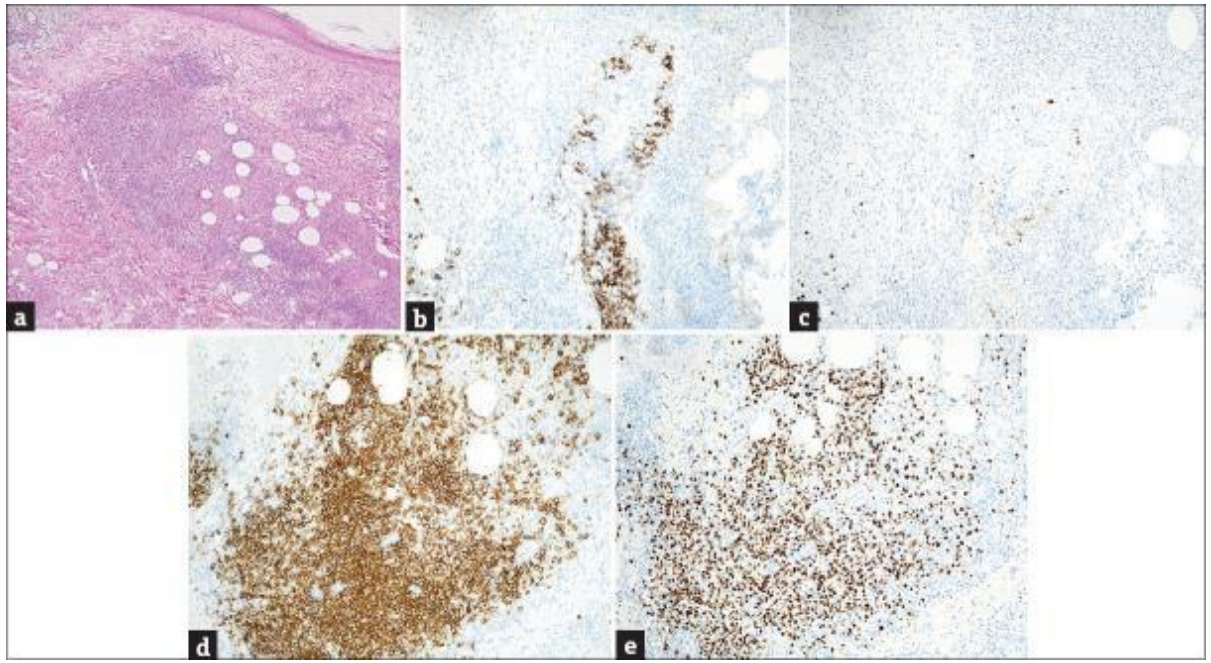
- An 80-year-old female presented with painful cutaneous nodules on her upper extremities and thighs for 2 weeks.
- Medical history revealed osteoporosis, with no immunosuppressive medication use.

ON EXAMINATION

Examination revealed painful erythematous and subcutaneous nodules in the upper extremities.

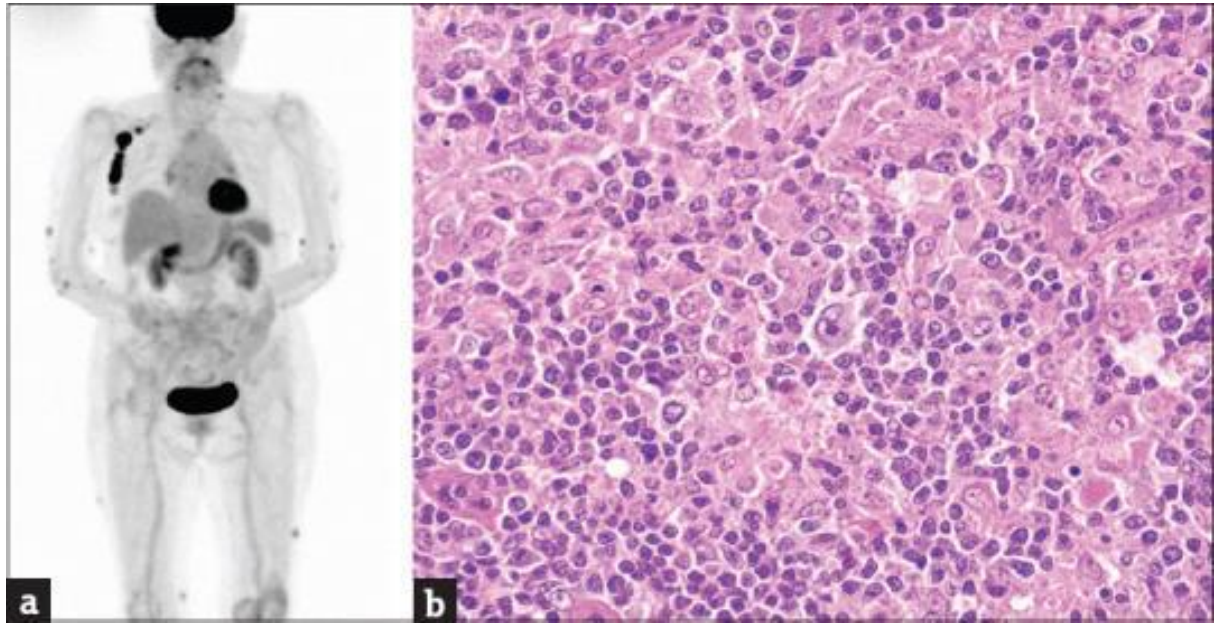


- Malignant lymphoma or nodular erythema was suspected; therefore, a biopsy was performed.
- Blood and biochemical tests were normal. HTLV-1 and HIV test results were negative.
- The patient had previously been immunized against EBV.
- EBER ISH (Epstein Barr virus Encoded RNA Insitu Hybridization Test) is the method of choice for detection of EBV infection at single cell level.



a) Infiltration of tumour and inflammatory cells around blood vessels extending from the dermis to the subcutaneous tissue. Inflammation with necrosis and epithelioid cell infiltration in areas other than that around the blood vessels. (b) EBER-ISH-positive blood vessel infiltrating cells. (c) CD20-positive blood vessel infiltrating cells. In areas with dense proliferation and infiltration of large, atypical lymphocytes, the majority of cells were EBER-ISH (d) and CD20 (e) positive

- Positron emission tomography CT revealed that the brain and lungs were normal.
- However, heavy accumulation was observed in the cervical and right axillary lymph nodes, with punctate accumulation in the skin of the limbs and subcutaneous nodules.
- A right axillary lymph node biopsy revealed loss of germinal centres and scattered epithelioid granulomas.



(a)PET-CT findings demonstrated strong accumulation in the cervical and right axillary lymph nodes, with punctate accumulation in the skin of the limbs and subcutaneous nodules. (b) A small number of Hodgkin-like cells under high magnification.

MANAGEMENT

- More than 50 EBER-positive cells were present in a high-power field, corresponding to LYG Grade 3, and the exacerbation of cutaneous lesions.
- Thus, early therapeutic intervention was necessary. The patient was hospitalized, and doxorubicin, etoposide, vincristine, cyclophosphamide, prednisone and rituximab (**DA-EPOCH-R**) therapy was initiated.
- Following treatment, the skin lesions faded.

LEARNING POINTS

- LYG is a rare EBV-associated B-cell lymphoproliferative disorder.
- Here LYG manifested in the skin and accompanying lymph nodes without any pulmonary lesions, emphasizing the importance of early therapeutic interventions.