Answers to Dermato-venereological Quiz on pages 43-44

1. The clinical differential diagnoses include: (a) Malignancy: cutaneous lymphoma, leukaemia cutis, Sweet' syndrome. (b) Cutaneous infections: ecchyma gangrenosum, cellulitis/erysipelas/ necrotising fasciitis. (c) Cutaneous inflammatory dermatoses: pyoderma gangrenosum, discoid lupus erythematosus.
2. The initial management includes complete blood picture with flow cytometry, LDH (lactase dehydrogenase) level, urgent haematology consultation in view of the pancytopenia for bone marrow biopsy/examination. Septic work up in view of fever and immunocompromised state should be performed and empirical antibiotics treatment should be considered to cover for common skin pathogens. The investigations include skin biopsy for histology, tissue for culture, AFB, mycology examination and culture, T-cell receptor (TCR) gene rearrangement, immunostaining/phenotyping. PET-CT scan to screen for possible organ involvement and lymph node, Fine-needle aspiration for cytology(FNAC)/biopsy for histology in view multiple lymphadenopathy should also be considered.

The diagnosis is Angioimmunoblastic T cell Lymphoma with prominent EBER associated large B-cell proliferation. Histology shows diffuse infiltrate of mixed population of atypical lymphoid cells in skin with no intraepithelial spread or lack of Pautrier microabscess. It is angiocentric with invasion of vessel wall accompanied by tissue necrosis. The medium sized cells are tumour T-cells, while large pleomorphic cells are EBER positive B-cells (Figures 2-5). PET/CT scan showed multiple hypermetabolic lymph nodes over neck, thorax, abdomen and increased uptake in spleen and marrow. There were also pulmonary opacities affecting left upper lobe and right middle lobe. The biopsies from lymph node, bone marrow trephine and bronchial lung showed similar morphology. She was started on chemotherapy with guarded prognosis.

The peripheral T cell lymphomas (PTCL) are a heterogeneous group of generally aggressive neoplasms that constitute <15% of all non-Hodgkin lymphomas (NHLs) in adults. Among these, in decreasing frequency of occurrence, are: Peripheral T cell lymphoma (not otherwise specified), Anaplastic large cell lymphoma (primary systemic type), Angioimmunoblastic T cell lymphoma, Extranodal NK/T cell lymphoma (nasal type), Subcutaneous panniculitis-like T cell lymphoma, Enteropathy associated T cell lymphoma, and Hepatosplenic T cell lymphoma.

Angioimmunoblastic T cell lymphoma (AITL) is thought to arise from a subset of peripheral CD4 positive T cells corresponding to follicular helper T cells. Patients typically present with the acute onset of a systemic illness characterized by generalized lymphadenopathy, hepatosplenomegaly, B systemic symptoms (fever, night sweats, weight loss), with or without a rash. It can also be associated with immunological abnormalities, such as autoimmune haemolytic anaemia, plasmacytosis, or polyclonal hypergammaglobulinemia. A polymorphous infiltrate is seen on lymph node biopsy with prominent proliferation of high endothelial venules and follicular dendritic cells.

The survival rate of PTCL depends on the subtype. In general, without treatment the survival is in terms of months. There is no general consensus regarding the optimal treatment regimen for these patients. The overall response rates with CHOP (cyclophosphamide, doxorubicin, vincristine, prednisone) chemotherapy for T cell NHL ranges from 50 to 70%. Due to the poor outcome, more aggressive strategies such as the use of autologous haematopoietic cell transplantation (HCT) or radiation therapy as consolidation have been proposed but their roles remain controversial and their use varies depending on the lymphoma subtype.

Further reading

1. World Health Organization Classification of Tumours of Haematopoietic and Lymphoid Tissues, Swerdlow SH, Campo E, Harris NL, et al. (Eds), IARC Press, Lyon 2008.
2. Swerdlow SH, Campo E, Pileri SA, Harris NL, Stein H, Siebert R, et al. The 2016 revision of the World Health Organization classification of lymphoid neoplasms. Blood 2016;127:2375-90.
3. Willemze R, Cerroni L, Kempf W, Berti E, Facchetti F, Swerdlow SH, et al. The 2018 update of the WHO-EORTC classification for primary cutaneous lymphomas. Blood 2019, pii: blood-2018-11-881268. doi: 10.1182/blood-2018-11-881268.