A rare case of peripheral T-cell lymphoma, not otherwise specified presenting as a solitary abdominal mass

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LEARNING OBJECTIVE
To highlight the unusual presentation of peripheral T-cell lymphomas, not otherwise specified (PTCL, NOS)

HISTORY OF PRESENTING COMPLAINT
- 70 year old Caucasian male
- 4-month history of rapidly enlarging tumor on lower abdomen, episodes of night sweats and generalized pruritus
- No other relevant medical or drug history

CLINICAL EXAMINATION
- 16 by 13 centimeters fungating infiltrative tumor on right lower abdomen (see Fig. 1) with palpable inguinal lymphadenopathy

BLOOD TESTS AND IMAGING
- Raised serum lactate dehydrogenase level of 1015 units/L (normal range 90-500 units/L)
- Computed tomography (CAT) scan showed large mass on right anterior abdominal wall infiltrating into underlying subcutaneous fat, along with widespread lymphadenopathy

HISTOLOGICAL EXAMINATION
- Incisional biopsy sample was taken from the abdominal mass
- Histological examination revealed pandermal infiltration of atypical medium- to large-sized lymphoid cells, separated from the epidermis by a Grenz zone (see Fig. 2)
- Immunostaining showed cells were positive for pan-T-cell antigens (CD3 and CD5), and predominantly CD4 positive. Some of the large cells stained positive for CD30, but were negative for ALK-1 and EMA. CD56 and EBER were negative.

DIAGNOSIS
Peripheral T-cell lymphoma, not otherwise specified (PTCL, NOS) based on World Health Organization (WHO) classification of lymphomas

TREATMENT AND CLINICAL COURSE OF PATIENT
- Received treatment with six-cycle course of CHOP (cyclophosphamide, doxorubicin, vincristine, prednisolone) chemotherapy supervised by the local Hemato-Oncology team
- Post-treatment examination revealed almost complete regression of the abdominal mass (see Fig. 3)
- Previous widespread lymphadenopathy was no longer apparent on subsequent CAT scan
- Prognosis remains guarded and requires regular surveillance for relapse

MANAGEMENT AND PROGNOSIS
- Combination chemotherapy regimens such as CHOP regimen commonly employed to treat NHLs
- Prognosis is generally poor
- Reported 5-year survival rates are less than 20%

DISCUSSION
- PTCL is a rare and heterogeneous group of T-cell non-Hodgkin lymphomas (NHLs)
- Classification is complex and PTCL, NOS is used when a tumor does not fit into any specific subgroup

PRESENTATION
- Can present with nodal and/or extra-nodal disease
- Commonly involves bone marrow, liver and gastro-intestinal tract
- Cutaneous involvement reported in only 10 to 18% of cases as a primary or secondary manifestation
- Multiple, generalized or solitary skin tumors may be seen

DIAGNOSIS
- Diagnosis confirmed via histological examination with immunostaining
- Variable numbers of pleomorphic or immunoblast-like T cells, with little (if any) epidermotropism
- Cells show variable loss of pan-T-cell antigens, and often CD4 positive

Fig. 1. Fungating tumor on lower abdomen at initial presentation

Fig. 2. Skin biopsy showing (a) pandermal, diffuse infiltration of atypical lymphocytes which were large and pleomorphic with prominent nucleoli (haematoxylin and eosin, original magnification x 4). These cells showed diffuse positivity to T-cell markers (b) CD3 and (c) CD5 and were negative for B-cell marker (d) CD20.

Fig. 3. Residual hyperpigmentation with no textural change or induration on patient’s right lower abdomen post-chemotherapy