Malignant cutaneous lesions

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DESCRIPTION

An 81-year-old woman presented with fatigue, loss of appetite and painless skin lesions, which had developed suddenly. Other than type 2 diabetes, her history was unremarkable; she did not use any medication. On physical examination, several erythematous purple-reddish papules and plaques were observed on her leg (figure 1A) and trunk (figure 1B). Blood count showed anaemia (7.1 mmol/L), thrombocytopenia ($36 \times 10^9/L$) and leucocytosis ($17.2 \times 10^9/L$) with $5.2 \times 10^9/L$ blast cells in the peripheral blood smear (figure 1C). Biopsy of a skin lesion revealed a dense dermal infiltration by medium-sized monomorphic cells with blast-like appearance (figure 1D). Through additional immunohistochemistry, the diagnosis of a blastic plasmacytoid dendritic cell neoplasm (BPDCN) was made.

BPDCN is a rare, aggressive malignancy with approximately 150 described cases and only recently has the disease been appreciated as a separate entity.¹ The origin of the tumour is unclear, but has been linked to both natural killer (NK) and, more recently, to dendritic cells, and is therefore called BPDCN.² All patients develop skin lesions that range from nodules to patches and bruise-like areas. Skin biopsies reveal dermal infiltration with monomorphic poorly

differentiated medium-sized cells. The epidermis is typically spared. Lymphadenopathy and leukaemic transformation are less common; therefore, it has been debated whether it is primarily a skin or haematological disease.

Several other malignancies may infiltrate the skin to cause similar lesions. The most important differential diagnoses are acute myeloid leukaemia (AML) and myeloid sarcoma, which may similarly present with quickly developing, nodular and grey-blue skin lesions, usually referred to as leukaemia cutis. Since BPDCN expresses CD4, CD45, CD56, CD68, CD123 and T-cell lymphoma 1 and AML usually demonstrates negativity for these markers, but instead expresses several myeloid markers such as myeloperoxidase, a distinction is possible.³ Other malignancies that may produce similar skin lesions are a number of T-cell lymphomas, including cutaneous T-cell lymphoma (CTCL), subcutaneous panniculitis-like T-cell lymphoma (SPTL) and extranodal NK/T-cell lymphoma. The clinical presentation of these malignancies, however, may be very different from BPDCN, as these tumours develop more slowly over time, often with relapsing and recurrent atypical skin lesions prior to the diagnosis. CTCL usually presents with painless, pruritic patches or plaques. Erythroderma may also

Figure 1 (A and B) Macroscopic appearance of the cutaneous lesions on the patient's left leg and on the right side of the trunk, respectively. (C) Peripheral blood smear showing blast cells. (D) Histology of the truncal skin lesion showing dense infiltration of the entire dermis by monomorphic cells with blast-like appearance.

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occur. Skin biopsies reveal the presence of lymphocytes with indented contours in the upper dermis and epidermis. T-cell markers are usually only partly expressed, indicating immature lymphocytes. Typical BPDCN markers are negative.⁴ SPTL presents with subcutaneous nodules or plaques that have a panniculitis-like appearance. Biopsy shows infiltrate of atypical lymphocytes in fat lobules. SPTL expresses T-cell markers (CD3 and CD8), but does not show CD4 or CD56 positivity. Nasal-type extranodal NK/T-cell lymphoma typically presents with upper respiratory tract symptoms due to local tumour formation, however, extranodal presentation in the skin may resemble the lesions observed with BPDCN. Owing to their strong relation with NK cells, nasal-type extranodal NK/T-cell

lymphoma stains positive for cytotoxic granule proteins such as granzyme B, T-cell intracellular antigen-1 and perforin, which are not expressed by BPDCN. Also, the diseases differ from a morphological point of view as nasal-type extranodal NK/T-cell lymphoma typically induces a polymorphic infiltrate. Finally, NK/T-cell lymphoma usually demonstrates Epstein-Barr virus positivity.^{3 5}

The prognosis of a BPDCN is poor, with median overall survival of 14 months, even if treated with polychemotherapy. The survival rates for AML are significantly better, and also the mentioned T-cell lymphomas usually have a better prognosis, particularly in the earlier stages of the disease. Given the age of our patient, no additional diagnostic tests were performed and supportive care was initiated; she died a few weeks later.

Competing interests None declared.

Patient consent Obtained.

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Learning points

- ▶ When a patient presents with extensive skin lesions, a haematological malignancy, predominantly acute myeloid leukaemia, should be early considered in the differential.
- ▶ A blastic plasmacytoid dendritic cell neoplasm is a very rare aggressive malignancy that always presents with skin lesions. Since all patients have cutaneous involvement and a minority have bone marrow localisation at the time of diagnosis, the disease is considered by many to be primarily a skin disease and not a haematological disorder.

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