

Cutaneous Myeloid Sarcoma Presenting as Grey Pigmented Macules

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According to previous reports, cutaneous myeloid sarcoma often manifests as a red nodule on the skin. We report here a patient with cutaneous myeloid sarcoma presenting with a unique skin lesion.

CASE REPORT

A 71-year-old man presented with pigmented macules of one month duration. Physical examination showed multiple grey pigmented macules, 10–30 mm in diameter, on the face and trunk (Fig. 1a). His white blood cell count was $2.3 \times 10^9/l$, with 8.5% blast cells, 1.0% myeloid cells, 3.5% stab cells, 23.0% segmented neutrophils, 56.0% lymphocytes, 6.5% monocytes, 0.5% eosinophils, and 0.5% basophils. His platelet count was $7.2 \times 10^9/l$, red blood cell count $3.75 \times 10^{12}/l$, and haemoglobin level 10.3 g/dl. Other routine biochemical tests and urinalysis were normal. Microscopic examination showed sheets of cells with abundant eosinophilic cytoplasm, enlarged, frequently reniform, nuclei, and numerous mitotic figures (Fig. 1b). Immunohistochemical studies were positive for CD43, CD45, myeloperoxidase, and lysozyme, and negative for CD3, CD4, CD8, CD20, and CD56, confirming the diagnosis of myeloid sarcoma. When examined at the department of haematology at our request, the patient was diagnosed as having acute myeloid leukaemia with the 7;21 translocation. On the basis of this diagnosis, the patient was administered induction chemotherapy with intravenous

enocitabine and aclarubicin hydrochloride. The macules promptly disappeared with this treatment. After 3 courses of chemotherapy, the patient was in complete clinical remission and remained disease-free during a follow-up period of 18 months.

DISCUSSION

The cutaneous manifestations of leukaemia can be non-specific (containing no leukemic cells), e.g. panniculitis and generalized pruritus; or specific (leukaemia cutis). Non-specific lesions are found in up to 30% of leukaemia patients (1), but leukaemia cutis is much less common. Skin involvement is usually a late occurrence, and leukaemia cutis preceding marrow or peripheral blood abnormality is extremely rare (2). The clinical manifestations of leukaemia cutis are variable, including macules, nodules, purpura, and erythroderma, and the condition often resembles cutaneous lymphoma.

Myeloid sarcoma is an extramedullary tumour of immature cells of granulocytic series, generally occurring in approximately 2% of patients with acute myeloid leukaemia (3). Myeloid sarcoma occurs mostly in adults aged 45–55 years, and it has a predilection for the bone, soft tissue, and skin (4), but they have been found in many other organs, including the abdominal organs, testis, and lacrimal gland. The skin lesions most commonly occur on the trunk, scalp and face. In general, they are solitary, but may be multiple and disseminated as firm nodular masses of variable sizes, which do not ulcerate. The patient with cutaneous myeloid sarcoma reported here presented with macules instead of nodules. However, to date, there are no reports of cutaneous myeloid sarcoma presenting as macules. Further study of additional cases is needed to determine definitively if the macule might represent an early sign of nodule formation, or if a macule might represent a new clinical manifestation of cutaneous myeloid sarcoma.

It was earlier termed chloroma, due to the green appearance resulting from the increased level of myeloperoxidase enzymes in the immature myeloid cells. The term myeloid sarcoma was coined following the description of lesions, which were not green. In the



Fig. 1. (a) Grey pigmented macules on the back. (b) Sheets of cells with abundant eosinophilic cytoplasm, enlarged, frequently reniform nuclei, and numerous mitotic figures (haematoxylin-eosin stain; original magnification $\times 400$).

present case, the lesion was grey in colour. To date, one report has been published of a grey nodule, in a Japanese patient with myeloid sarcoma (5). This lesion colour may be a characteristic of myeloid sarcoma in Mongoloid people.

The most commonly confused tumours include diffuse large cell lymphoma and Ewing's sarcoma. The diagnosis of sarcoma is based on combined haematoxylin and eosin assessment, and phenotypic studies with or without molecular and cytogenetic studies.

The pathogenesis of cutaneous myeloid sarcoma is unknown. In general, homing to specific tissues is intricately controlled by expression of different chemokine receptors and adhesion molecules. Blast neural cell adhesion molecule (CD56) has long been implicated in extravasation of myeloid precursor cells (6). Supporting this, neural cell adhesion molecule blast expression has been associated with a high incidence of myeloid sarcoma (7), and is common in patients with t(8;21) (8). Mechanistically, neural cell adhesion molecule is also highly expressed in testicular, ovarian, and gut tissue, which could account for extramedullary homing to these sites (9).

The authors declare no conflicts of interest.

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