Case Report

Cutaneous myeloid sarcoma: a localised or a systemic disease?

皮膚髓樣肉瘤：局部或全身性疾病？

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Chloroma, also known as myeloid sarcoma, is an extramedullary collection of immature myeloid cells. We describe a patient with myeloid sarcoma preceding the appearance of acute myeloid leukaemia who was treated with radiation therapy with temporary effect. The case emphasises the importance of a systemic treatment for this cutaneous disease.

也稱為骨髓性肉瘤的緣瘤，是未成熟的骨髓細胞在骨髓以外集結。我們描述了一名患者在其急性骨髓性白血病之前發現患有骨髓性肉瘤，該患者接受了放射治療並獲得暫時緩解。此病例突顯了對這種皮膚病進行全身治療的重要性。

Keywords: Chemotherapy, chloroma, leukaemia, myeloid sarcoma, radiation therapy

關鍵詞：化療、緣瘤、白血病、骨髓性肉瘤、放射治療

Case report

A 68-year-old male presented with two subcutaneous asymptomatic nodules on the left shoulder and one in right thigh for four months duration (Figures 1 and 2). The patient was otherwise healthy.

On examination, he had two discrete erythematous purple firm nodules on the lateral left shoulder and one in right medial thigh. There was no evidence of hepatomegaly, splenomegaly or lymphadenopathy. Blood tests showed thrombocytopenia (112 g/L) and a normal white blood cell count and differential. Renal function was mildly elevated with creatinine 118 µmol/L. The LDH was within normal range. Skin biopsies were taken from the lesions. Histopathologically, the dermis and subcutis were infiltrated by neoplastic cells with a moderate amount of granular eosinophilic cytoplasm and round or kidney shaped nuclei with prominent nucleoli (Figure 3). Immunohistochemical stains showed a positive CD43 and CD68 stain and a negative staining to myeloperoxidase CD117, CD3 and CD20.
Following the diagnosis of myeloid sarcoma, the patient was referred to a tertiary hospital where a bone marrow biopsy showed no evidence of acute myelogenous leukaemia (AML) or abnormal populations on flow cytometry. He was treated with localised radiation therapy to the nodules on the left shoulder and right medial thigh. This caused the nodules to regress in size, although multiple new nodules appeared on the limbs and torso. The new nodules were treated locally with radiation as well.

Following complaints of abdominal pain, the patient was admitted to a hospital where blood tests and a second bone marrow biopsy confirmed a diagnosis of acute myelogenous leukaemia. Chemotherapy treatment (fludarabine and cytarabine) was initiated. However, the patient passed away two weeks later following an opportunistic pneumonia.

**Discussion**

Granulocytic sarcoma, also known as myeloid sarcoma (MS), is an aggregation of extramedullary myeloid precursor cells. It is also referred to as chloroma, which is based on the Greek word for green, due to the colour of some of the tumours caused by the action of the enzyme myeloperoxidase in the cells.

Myeloid sarcoma may occur at any age group but is more common in adults in the fifth and sixth decades of life, with a median age of 56.\(^1\) It may involve various organs such as the skin, bone, central nervous system, lymph nodes, gastrointestinal and genitourinary system and is often associated with AML as well as with myelodysplastic syndrome or myeloproliferative neoplasm. It may present de novo without evidence of pathological involvement of the peripheral blood or bone marrow (isolated MS) in 25% of the patients, following the diagnosis in 50% of cases and simultaneously with AML in 15-35%.\(^2,3\)

The association with AML mandates a bone marrow aspiration in every patient with a diagnosis of isolated MS, to rule out a concomitant AML or other haematological malignancies. The prognosis of isolated MS is poor due to the high rate of progression to acute leukaemia. Most patients with MS with no concurrent AML at the time of diagnosis will develop AML within two years.\(^4\) Due to this considerable high frequency of AML in MS patients, the recommended treatment is systemic chemotherapy. This approach is supported by observations showing

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**Figure 1.** Erythematous violaceous firm nodule on right medial thigh.

**Figure 2.** Erythematous violaceous firm nodules on left lateral shoulder.
that 88-100% of the patients who were treated with localised modalities such as radiotherapy and surgery had a higher rate of transformation to AML than patients who were treated with chemotherapy. In addition, the latent period for progression to acute leukaemia is longer in patients treated systemically. Therefore, systemic treatment is recommended to all patients with isolated myeloid sarcoma.

Localised treatments such as surgery and radiotherapy are advocated only in limited cases when tumours cause local organ dysfunction or obstruction or for purpose of debulking, or consolidation together with chemotherapy. In refractory patients or in cases of relapse, bone marrow transplantation can be considered. A limited number of studies have shown an improved overall survival in patients who had stem cell transplantation. It has been speculated that stem cell transplantation concomitantly with chemotherapy has a role in relapsed or refractory cases. Other emerging treatment options include molecular targeted therapies such as imatinib and gemtuzumab ozogamicin.

In conclusion, chloroma is a tumour of immature granulocytic cells isolated to the skin. Due to the high rate of progression to AML, systemic therapies are advised. Nevertheless, since there are no large prospective studies in the literature, new studies are essential to determine the best approach for the treatment of patients with isolated myeloid sarcoma. Myeloid sarcoma patients should be referred to a multidisciplinary cancer care team to involve a haemato-oncologist in management and decision making at an early stage.

References


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