A patient with extramedullary acute myeloid leukaemia involving the brachial plexus: Case report and review of the literature

Brakiyal pleksusu tutan ekstramedüller akut miyeloid lösemili bir hasta: Vaka raporu ve literatürün incelenmesi

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Abstract

Extramedullary leukaemic involvement is not uncommon as part of the presentation in acute myeloid leukaemia (AML). However, infiltrative peripheral neuropathy due to AML was rarely reported. We report a case of extramedullary leukaemic infiltration of the brachial plexus in a patient with relapsed AML. *(Turk J Hematol 2008; 25: 98-100)*

Key words: Acute myeloid leukaemia, extramedullary leukaemic infiltration, brachial plexus

Özet

Akut miyeloid löseminin (AML) bir parçası olarak ekstramedüller lösemik tutulum nadir bir durum değildir. Ancak AML'ye bağlı infiltratif periferik nötropati nadiren bildirilmiştir. Burada, relaps AML'li bir hastada brakiyal pleksusun ekstramedüller lösemik infiltrasyonu vakasını sunuyoruz. *(Turk J Hematol 2008; 25: 97-9)*

Anahtar kelimeler: Akut miyeloid lösemi, ekstramedüller lösemik infiltrasyon, brakiyal pleksus

Introduction

Acute myeloid leukaemia (AML) is a heterogeneous group of haematological disorders characterized by uncontrolled proliferation and infiltration of neoplastic haematopoietic cells in the bone marrow and blood. Extramedullary tissue involvement is not uncommon as part of the presentation in AML. Sites of involvement include liver, spleen, skin, gum, bone, testis and central nervous system (CNS). Leukaemia cutis representing leukaemic infiltration to skin occurs in about 10% of patients with AML and it is more common with the monocytic subtypes, including acute monocytic and myelomonocytic leukaemias (AML-M4/M5) [1]. However, CNS involvement in AML is a relatively uncommon manifestation. Leukaemic infiltration to brachial plexus is particularly rare. Herein, we report a case of extramedullary leukaemia infiltration of the brachial plexus and skin in a patient with relapsed AML.

Case Report

The patient is a 62-year old Chinese gentleman with a history of papillary transitional cell carcinoma of the bladder which was first diagnosed in 1998 and was treated with transurethral resection of bladder tumour (TURBT) in March 1998. On surveillance cystocopy, a recurrence of the bladder carcinoma was detected in January 2006.

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Another surgery was planned for resection of the tumour. However, he was found to have pancytopenia and fever. Full blood count showed haemoglobin of 12.5g/dL (normal 14.0-18.0g/dL), white blood cell count of $1.7x10^9$/L (normal 4.0-10.0 $x10^9$/L) and platelet count of $119x10^9$/L (normal 140-$440x10^9$/L).

Bone marrow studies reviewed acute myeloid leukaemia (AML-M5a). Cytogenetic studies showed abnormal karyotype with t(X;10) and structural abnormalities of the long arm of chromosome 11. He was given induction chemotherapy with intravenous idarubicin 12mg/m^2 for 3 days and intravenous cytarabine 100mg/m^2 for 7 days. He achieved complete remission in February 2006.

During chemotherapy, he developed neutropenic sepsis with invasive pulmonary aspergillosis involving the middle lobe of right lung which was treated with intravenous amphotericin followed by oral itraconazole. He subsequently underwent wedge resection of the right middle lobe lesion in March 2006.

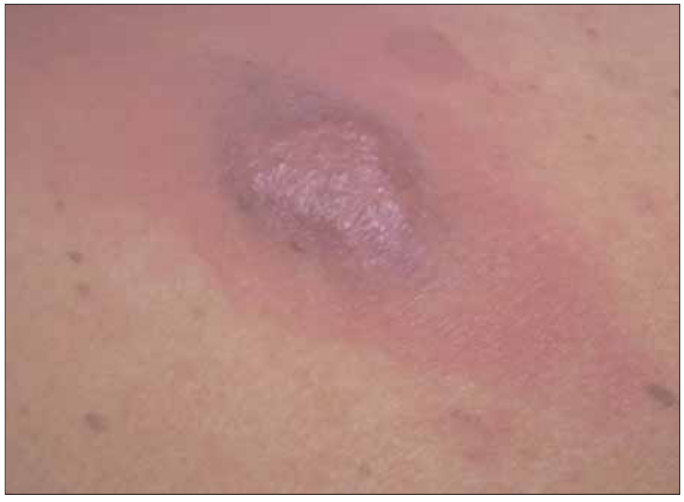

Figure 1. Biopsy of this papular skin lesion showed leukaemic infiltrate

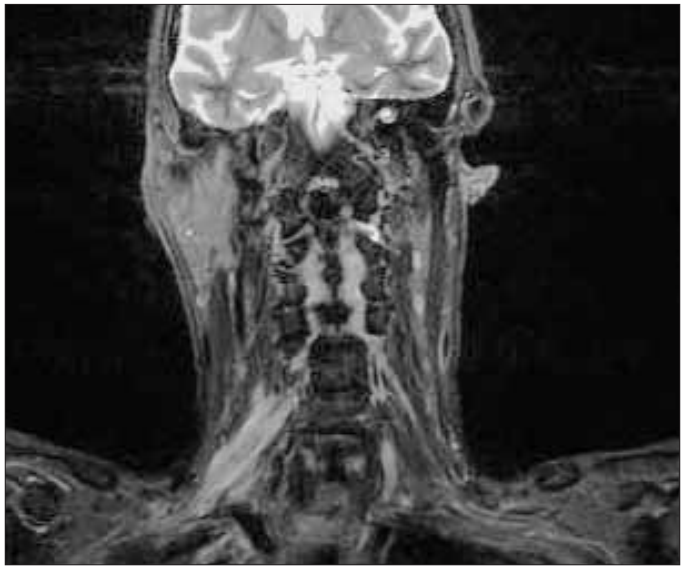

Figure 2. MRI showed perineural enhancing tissue at the right brachial plexus suggesting leukaemic deposits

He underwent TURBT for recurrence of carcinoma of bladder in March 2006. Due to the treatment for recurrent carcinoma of bladder and the wedge lung resection, consolidation chemotherapy was delayed. The consolidation chemotherapy was given in April 2006.

It consisted of intravenous idarubicin 12mg/m^2 for 2 days and intravenous cytarabine 100mg/m^2 for 7 days.

In July 2006, the patient developed multiple skin lesions at his back and groin region, characterized as polymorphous papular nodular lesions of varying intensity of erythema and induration (Figure 1). Punch biopsy of the skin lesion showed blast infiltrate consistent with acute leukaemia. In addition, the patient complained of right arm pain, associated with weakness and numbness of his right upper limb. Clinically his right upper limb power was graded as 3/5 initially and his left upper limb power was normal. His right upper limb weakness progressed over the next few days after his presentation and the power became 1/5 with difficulty in right shoulder abduction, right elbow flexion and finger movements. Magnetic resonance imaging (MRI) of the cervical spine and brachial plexus was performed on 7 July 2006 and showed the presence of perineural enhancing tissue at the right brachial plexus extending from the neural foramina laterally to the mid-clavicular plane, suggesting leukaemic deposits affecting the trunks of the right brachial plexus (Figure 2). Nerve conduction study revealed evidence of right brachial plexopathy. Bone marrow examination was also performed on 6 July 2006. Bone marrow aspirate did not show any morphological evidence of leukaemia, but flow cytometry detected 6% myeloblasts. Hence, the patient relapsed with extramedullary leukaemia to skin and right brachial plexus.

With these new findings and the diagnosis of relapse acute monocytic leukaemia with extramedullary leukaemic involvement, the patient received salvage chemotherapy with intravenous high dose cytarabine 2g/m^2 every 12-hourly for 5 days.

The patient's skin lesions showed significant improvement after the initiation of high dose cytarabine (Figure 3). Complete resolution of the skin lesions was achieved by day 17 of

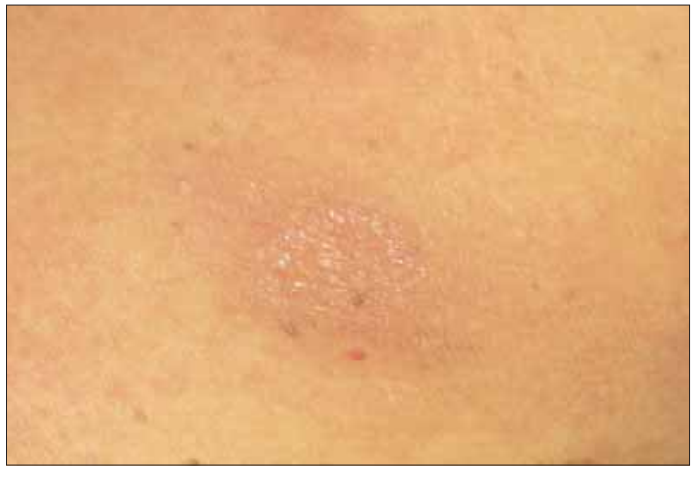

Figure 3. The skin lesion showed significant clinical improvement with chemotherapy

chemotherapy. His right upper limb numbness, weakness and pain had also improved steadily after the initiation of chemotherapy. By day 15 of chemotherapy, the patient was able to flex his right elbow and had reasonably good finger movements. By day 37 of chemotherapy, his right shoulder abduction was full with power graded 4/5. However, residual weakness persisted with the right upper limb power of 4/5.

Discussion

The presenting signs and symptoms of AML are related to the decreased production of normal haematopoietic cells and other organ involvement by the leukaemic cells.

System leukaemic disease is sometimes accompanied by the presence of extramedullary leukaemic infiltration. Skin involvement, also called leukaemia cutis, occurs in about 10% of patients with AML and usually manifests as violaceous, nontender plaques or nodules, which on biopsy are found to be infiltrated by myeloblasts [1]. Skin involvement is more common with the monocytic subtypes of AML [2]. CNS involvement is uncommon, and infiltrative peripheral neuropathy is rarely reported. MRI study of the affected nerve root may demonstrate thickening of the nerve sheath, which is considered as a radiological suggestion of leukaemic involvement [1]. 3 cases of infiltrative peripheral neuropathy have so far been described in the literature. Only one case reported partial response to high dose cytarabine therapy [3-5].

We described a case of relapse AML with leukaemia cutis and leukaemic infiltration to the brachial plexus with disease relapse in bone marrow evidenced only on flow cytometry. Skin biopsy confirmed the cutaneous leukaemic disease. MRI and nerve conduction study suggested that the brachial plexopathy was related to leukaemic infiltration. With the initiation of chemotherapy, the skin lesions resolved and there was obvious neurological improvement of the brachial plexopathy. The clinical improvement after treatment with chemotherapy further confirmed the diagnosis of leukaemic involvement of the brachial plexus.

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