Myelodysplasia presenting as erythroderma

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Summary: Erythroderma has not previously been reported to be a feature of myelodysplasia. We report two cases of myelodysplasia presenting with erythroderma, one of which was associated with skin infiltration by blast cells.

Introduction

Cutaneous manifestations of haematological malignancies, although well recognized in acute monoblastic leukaemia and lymphoma, are relatively uncommon in primary myelodysplastic syndrome (MDS), which is a group of heterogeneous disorders formerly known as pre-leukaemia. Previously reported cases of skin involvement in this syndrome have been confined to a sub-type of MDS, namely chronic myelomonocytic leukaemia. We now report 2 cases of primary myelodysplastic syndrome presenting as generalized erythroderma.

Case reports

Case 1

A previously fit 62 year old woman was admitted to hospital in April 1985 with a 6-week history of a generalized itchy rash followed by pleuritic chest pain, cough and progressive tiredness. On examination, she was clinically anaemic with a generalized scaling erythroderma; some of the lesions, especially those in the lower limbs, were purpuric. She also had signs of right middle lobe consolidation. There was no lymphadenopathy or hepatosplenomegaly. Her past medical history was unremarkable, with no exposure to radiation and no recent medication.

Investigations revealed the following: haemoglobin 7.5 g/dl, white cell count $3.1 \times 10^9/l$, and platelets $60 \times 10^9/l$ with occasional blast cells, nucleated red cells and pseudo-Pelger neutrophils in the blood film. Bone marrow aspirate showed features of a myelodysplastic syndrome, being hypercellular with abnormal erythropoiesis and myelopoiesis (20% blast cells). No ringed sideroblasts were seen. Marrow karyotype showed an effective trisomy 14, [46,XX-14 + t(14q/

14q)]. Chest X-ray showed right middle lobe collapse/consolidation. Bronchoscopy and computerized tomographic scan of thorax and abdomen showed no evidence of primary neoplasm or lymphoma. Other investigations including plasma B₁₂ and folate, urea and electrolytes and liver function tests were all normal. Skin biopsy at this stage showed non-specific inflammatory changes. A diagnosis of MDS was made and her erythroderma was thought to be reactive to the underlying marrow disorder.

Treatment was given with blood products, intravenous antibiotics and systemic steroids. However, her erythroderma became more generalized and lesions more erythematous as her pancytopenia became progressively worse. Death occurred from a staphylococcal septicaemia 8 weeks after admission. Post-mortem examination revealed right middle lobe consolidation with multiple abscesses, a dysplastic marrow, and skin infiltration by abnormal blast cells (Figure 1).

Figure 1 Case 1. Skin biopsy post-mortem; $\times 800$.

Case 2

A 76 year old woman presented in March 1986 with a 6-week history of a generalized itchy rash and exertional dyspnoea. Her past health had been good with no history of skin diseases or exposure to chemicals or drugs. On physical examination she was clinically anaemic and had a generalized erythroderma with widespread erythematous macules and scalings, but had no other abnormal findings.

Investigations were as follows: haemoglobin 7.4 g/dl, white cell count $1.62 \times 10^9/l$, platelet count $170 \times 10^9/l$. Blood film was leuco-erythroblastic and contained hypogranular neutrophils. Bone marrow aspirate showed a dysplastic marrow with ringed sideroblasts but no excess of blasts consistent with a diagnosis of myelodysplastic syndrome (refractory anaemia). The marrow karotype was grossly abnormal [43XX -2-5-7-12-13-17-19]. Skin biopsy showed non-specific features with mild parakeratosis, acanthosis and mononuclear cell infiltrates. In view of the marrow findings and the chromosomal abnormalities, a diagnosis of primary MDS was made.

She was then treated with blood transfusion and potent topical steroids. Her skin condition improved initially but this was not sustained. 13-Cis-retinoic acid, a vitamin A analogue, was subsequently given. After 4 weeks of treatment, there was considerable improvement in her skin condition: her erythroderma largely resolved, leaving patchy, pale, scaly areas on her trunk. However, a repeat bone marrow showed no significant improvement and her neutropenia and anaemia persisted.

Discussion

Generalized erythroderma, although well-recognized as a manifestation of internal malignancy, has, to date,

not been reported in association with primary myelodysplastic syndrome. On the other hand, skin manifestations are a known feature of a number of haematological malignancies, especially those with a monocytoid differentiation, such as acute monoblastic leukaemia. These infiltrates frequently present as non-tender, non-itchy nodules which may regress as the patients respond to chemotherapy. However, skin involvement in primary MDS is rare and is usually confined to the chronic myelomonocytic type.^{1,2} Recently a pernio-like infiltrate and cutaneous xanthomata have also been described.^{3,4}

These two cases suggest that primary MDS can present with erythroderma. Moreover, it is probably significant that the second patient, with less severe marrow abnormalities has not, to date, been shown to have skin infiltration by blast cell, while the first patient developed skin infiltration as her marrow became progressively worse with increases in blast cell population. The use of cis-retinoic acid in the second patient is interesting, since this drug has been reported to induce haematological improvement in MDS,⁵ but as yet no favourable results have been reported in the treatment of erythroderma.

In conclusion, we suggest that primary myelodysplastic syndrome, as with other internal malignancies, should be considered in the differential diagnosis of erythroderma.

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