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CORRESPONDENCE

Neutrophilic Erythrophagocytosis in MDS-MLD

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Dear Sir
An 85 years old male presented with generalized weakness and numbness of feet since 6 months. Complete blood count (CBC) showed pancytopenia (Hemoglobin-96 g/l, WBC-2.5 $\times$ 10^9/l) and platelets-47 $\times$ 10^9/l with an absolute neutrophil count of 0.675 $\times$ 10^9/l). Peripheral blood smear showed mild dysgranulopoiesis with few pelgeroid neutrophils. Bone marrow aspirate smears were hypercellular and showed marked erythroid hyperplasia (Myeloid: Erythroid ratio = 1:2.4). There was borderline (10%) dyserythropoiesis with significant dysgranulopoiesis (17%) and dysmegakaryopoiesis (40%). Few neutrophils (5 per 10 oil immersion fields) showed erythrophagocytosis (Fig. 1a, b) with engulment of mature red cells while others showed dysplastic changes with pelgeroid forms (Fig. 1 c, arrow) and hypogranularity. Rare erythroblasts also showed engulfment of RBCs (Fig. 1d). No ring sideroblasts were seen. There was no significant increase in blasts (2%) on aspirate smears or trephine biopsy. The same was also confirmed on immunostaining by CD34 and CD117 on trephine biopsy sections. Lactate dehydrogenase, vitamin B12, folic acid and ferritin levels were raised. Conventional cytogenetics showed normal 46,XY[20/20] karyotype. A diagnosis of myelodysplastic syndrome with multi-lineage dysplasia (MDS-MLD) with neutrophilic erythrophagocytosis was made.

The patient was lost to follow up and presented 16 months later with fever and cough since 5 days. CBC showed pancytopenia. Bone marrow aspirate smears were hypercellular and showed megaloblastoid erythroid hyperplasia, frequent erythrophagocytosis by dysplastic neutrophils and occasional erythroblasts similar to the initial presentation. There was no significant increase in blasts (3%) or disease progression. Significant (> 10%) bilineage dysplasia (dysgranulopoiesis and dysmegakaryopoiesis) was seen.

Erythrophagocytosis by neutrophils in the peripheral smears has been reported in patients with paroxysmal cold hemoglobinuria, poisoning with potassium chlorate, sickle cell disease, incompatible blood transfusion and cold agglutinin disease [1].

The phenomenon of erythrophagocytosis by dysplastic myeloid elements is rare and has been reported in MDS with multilineage dysplasia (MDS-MLD) associated with del20q [2].

In a study of 1408 patients with MDS, 28% had autoimmune diseases. Autoimmune diseases were more common in female patients with MDS and in patients with refractory anemia or refractory cytopenia with multilineage dysplasia (WHO 2008) [3].

A multi-centric study from France assessed 123 patients with MDS and CMML [WHO 2008 classification: Refractory cytopenia with multilineage dysplasia (26%), refractory anemia with excess blasts 1 (15%), and CMML-1 in 16% patients]. The authors also collected information on autoimmune and inflammatory conditions in these patients. Two-thirds of the patients were males. The most common systemic disorders seen were systemic vasculitides (32%), connective tissue disorders (25%) and inflammatory arthritis (23%) among others [4].

Studies have shown that immune dysregulation and abnormal T cell hemostasis plays a role in the pathogenesis of myelodysplastic syndromes (MDS). Oligoclonal or monoclonal T-cell expansions of CD8 +/CD57 +/CD28-

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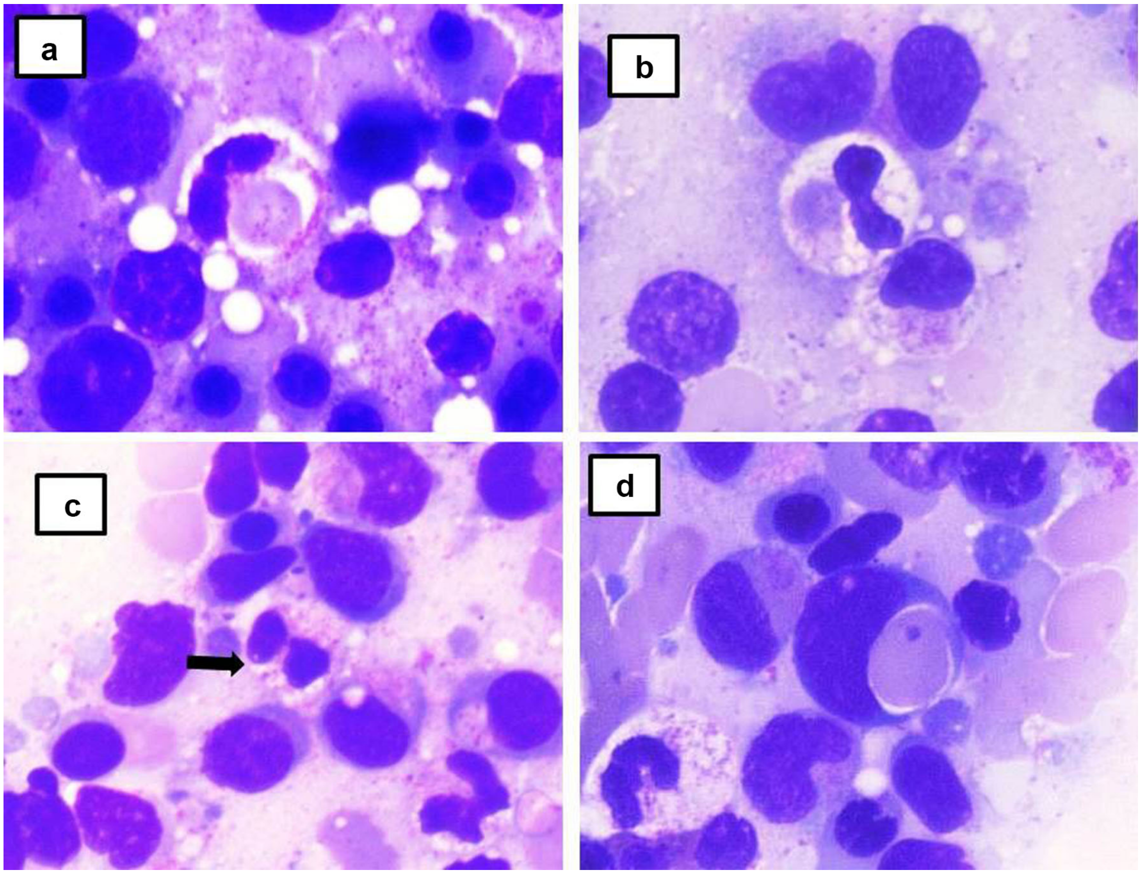

Fig. 1 **a**, **b** Show dysplastic neutrophils engulfing intact RBCs. **c** Shows a pelgeroid neutrophil (arrow). **d** Shows an erythroblast engulfing a red blood cell

effector cells have been reported in about half of patients with MDS. There is also increased telomere attrition of T cells in patients with MDS compared to controls. Alteration of specific T-cell subpopulations has also been reported which is associated with autoimmunity. Other mechanisms contributing to immune dysregulation in MDS include increase in inflammatory cytokines and number and activity of macrophages and natural killer cells. Genetic alterations leading to clonal T cell expansions are also thought to play a role [5].

The unusual phenomenon of neutrophilic erythrophagocytosis may act as a surrogate indicator of an autoimmune component in the pathogenesis of MDS.

Compliance with Ethical Standards

Ethical Approval No animals were used in this study. All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Informed Consent Informed consent was not taken as this is an image based report with no patient intervention specific to the study.

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