Granuloblastic sarcoma: onset with cutaneous polymorphic lesions

Haematologica 2007; 88:(3)e43

A 54 year-old-woman developed papulo-erythematous lesions on her face, arms, and trunk .These lesions showed polymorphic features. (Figure 1) Biopsy revealed an extramedullary (cutaneous and subcutaneous) infiltration of poorly differentiated myeloid cells CD34+, CD68+, LYSOZIME+, MPO+, CAE-, CD3-.(Figure 2-3) The bone marrow aspiration was compatible with chronic myelomonocytic leukemia with normal cytogenetics (46 XX), and normal peripheral blood cell count. She was given standard induction chemotherapy for acute myeloid leukemia with Ara-c 100 mg/mq for 7 days (continuous infusion) and Idarubicine 10 mg/mq for 3 days. She obtained complete regression of cutaneous lesions, but bone marrow aspiration showed an increase of blastic cells (10-15%). After discharge she developed acute cholecystitis and bronchopneumonia. The following cycle of chemotherapy was postponed because she underwent cholecystectomy.

Complete remission of cutaneous lesions lasted 62 days. Afterwards she developed acute myeloid leukemia with complex cytogenetics (48 XX, +8, +8, der(17), t(1;17)q21p13 [9]/ 49 idem, +del(4)q21 [8]/ 49 idem, +4 [14]/46 XX [1]) without cutaneous relapse.

The patient died during salvage chemotherapy in aplasia because of multiorgan failure.

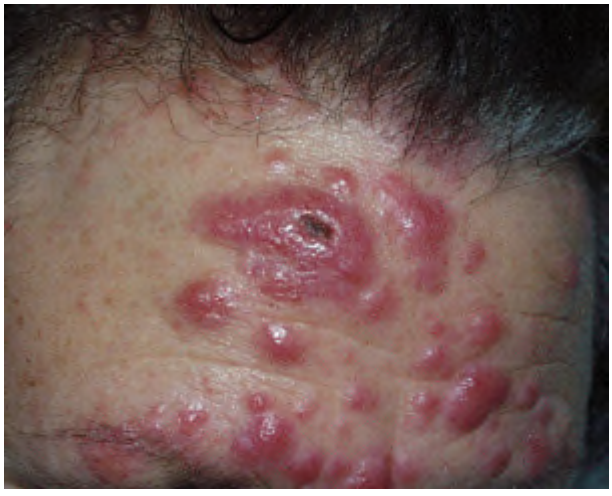

Figure 1. polymorphic papulo-erythematous lesions on the face

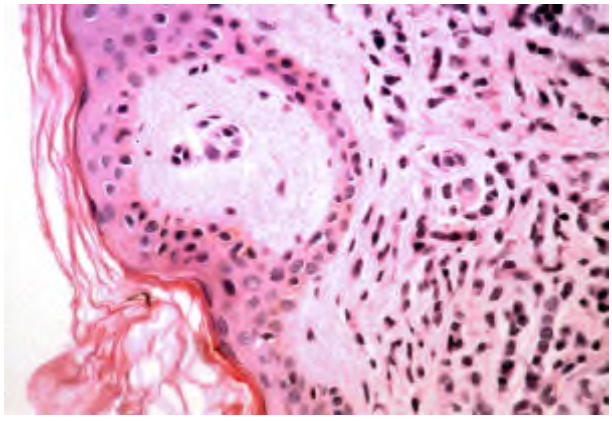

Figure 2. Diffuse sub-epidermal and epidermal infiltration of monomorphic monocytoid blasts (400x)

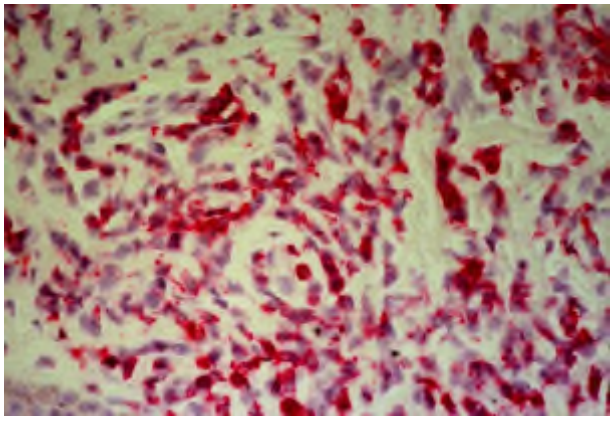

Figure 3. Blastic cells showed monoclonal antibody CD68 positive reaction (400x).

G. D'Avanzo, A. Nosari, P. G. Oreste, E. Morra

Haematology and Pathology Unit, Niguarda Ca' Granda Hospital, Milan, Italy

References

1. Cavadar AO, Babacan E: High risk subgroup of acute myelomonocytic leukemia (AMML) with orbito-ocular granulocytic sarcoma (OOGS) in Turkish children. Retrospective analysis of clinical, haematological, ultrastructural and therapeutical findings of thirty-three OOGS. Acta Haematol 1989:81(2),80-5;
2. Pero Silva A, Garcia Saavedra V: Granulocytic sarcoma, disclosing myelomonocytic leukemia. Ann Med Interne (Paris) 1990:141(6),559-60.