Atypical pyoderma gangrenosum in a patient with post-polycythaemic myelofibrosis

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A 65 year old lady was diagnosed to have Polycythemia Vera in 1995. She was treated with hydroxyurea and her disease remained under control until early 2003 when it progressed to myelofibrosis. In October 2003, she was admitted to our hospital with fever and a vesicopustular eruption on the lateral surface of her right hand (panel A). Blood counts on admission were as follows: Hct 33.7%, Hb 11.2 g/dL, WBC 46.9×10^6/L with 79% neutrophils, 2% myelocytes, 2% metamyelocytes, 3% monocytes, 6% eosinophils, 8% lymphocytes and platelets 79.0×10^6/L. Physical examination revealed a reddish-violaceous, slightly edematous plaque, which covered the external site of the right hand and part of the right wrist. On the plaque were confluent vesicopustules with seropurulent and hemorrhagic content. There were also erosions covered with hematogenous crusts (A). The patient had an enlarged spleen, 10 cm below the left costal margin, but no lymphadenopathy or hepatomegaly. Differential diagnosis included an infectious process, myeloid sarcoma, pyoderma gangrenosum and Sweet's syndrome.

Multiple cultures remained negative for pathogenic microorganisms and treatment with broad-spectrum antibiotics failed to improve the skin lesions. Biopsies from the erythematous nodules revealed clusters of neutrophils and hemorrhage in the upper dermis, without any vascular changes (B). Based on the above findings we felt that the most probable diagnosis was an atypical pyoderma gangrenosum. The patient was treated with a course of steroids, which resulted in prompt improvement and healing of the lesions in a short period of time (C).

M. Papaioannou,[1] E. Georgiou,[1] F. Chrysomallis,[2] J.I.Christakis[1]

[1]Department of Haematology, Theagenion Cancer Center, Thessaloniki, Greece; [2]Department of Dermatology, Medical School, University of Thessaloniki, Greece

E-mail:marygpap@otenet.gr

Figure A.

Figure B.