Acta Haematologica Polonica 2010, 41, Nr 2, str. 317–322

PRACA KAZUISTYCZNA – Case Report

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Pyoderma gangrenosum as a risk factor of acute myeloid leukaemia

Nieinfekcyjne zgorzelinowe zapalenie skóry jako czynnik ryzyka ostrej białaczki szpikowej

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SUMMARY

Pyoderma gangrenosum (PG) is an abacterial necrosis. The etiopathogenesis is unknown [1]. It is often associated with a systemic disease such as inflammatory bowel disease (IBD), myeloproliferative disorders or monoclonal gammapathy [2]. Up to now, there is no characteristic serologic or hematologic marker of the PG [2]. We would like to present our recent experience in the treatment of a patient with PG and difficulties in diagnosing of coexisting massive leucocytosis.

KEY WORDS: Pyoderma gangrenosum – Acute leukemia – Hyperleucocytosis – Flow-cytometry

STRESZCZENIE

Zgorzelinowe zapalenie skóry (ZZS) jest przykładem niebakteryjnej martwicy o nieustalonej etiopatogenezie. Dotychczas nie odkryto markerów serologicznych oraz hematologicznych tej choroby. Zaobserwowano współwystępowanie ZZS z zapaleniem jelit, chorobami mieloproliferacyjnymi oraz monoklonalnymi gammapatiami. W naszej pracy przedstawiamy problemy diagnostyczne i terapeutyczne u pacjenta ze zgorzelinowym zapaleniem skóry i współistniejącą hyperleukocytozą.

SŁOWA KLUCZOWE: Zgorzelinowe zapalenie skóry – Ostra białaczka – Hiperleukocytoza,cytometria przepływowa

CASE REPORT

We report a 22-year-old man with a history of idiopathic thrombocytopenia in childhood. From 10th August 2009, he was complaining about abdominal pain. The next day he was bitten by an unidentified insect in both shins. Because of topical inflammatory reaction, a dermatologist prescribed antiallergic pills and an ointment with antibiotic and steroid, what led to transient improvement. On 14th August, he was operated on due to abdominal pain. Histopathological diagnosis was appendicitis. In perioperative period the patient was in severe general condition with high fever. Also, he developed necrotic dermatitis on his both shins (see Fig. 1 to Fig. 3). Leucocytosis increased from 19.6 G/L to 87 G/L with 61.8% and 91.7% neutrocytes respectively. C-reactive protein was over 300 mg/L. Pericarditis, fluid in pleura and peritoneum was found. Microscopic, flow cytometry and molecular biology testing of peripheral blood excluded leukemia. The therapy with prednisolone (60 mg/day) and cyclosporine (100 mg/day) started on 28 August 2009 resulted in complete improvement of general condi-

tion, disappearing of PG symptoms (see Fig. 4 to Fig. 6) and reducing of white blood cells to 11G/L during two weeks. Extensive check-up didn't reveal any systemic disease. The skin of the body was clear, excluding 2 tattoos made a few years ago. The therapy was continued but progression of leukocytes up to 96.6 G/L with 59–70% neutrocytes and anemia up to 7.5 g/dl were observed. The patient had again fever with shivers and drenching sweats. Bone marrow aspiration revealed 60% of blasts of monoblastic morphology. Histopathology of bone marrow revealed 15% leukemic infiltration. A karyotype was normal.

Three-colour, fluorescence based on CD45/SSC gating, flow cytometry analysis of bone marrow indicated the presence of predominant cell subset (approximately 90%) with the myelomonoidal phenotype category: CD33+ – 88,7%, CD13+ – 17,1%, CD15+ – 61,5%, CD65+ – 81,1%, CD14+ – 46,6%, CD64+ – 66,1%, CD114+ – 79,7%, CD116+ – 76,4%, CD38+ – 65,2%, MPO+ – 36,3%, HLA-DR+ – 67,2%, CD34+ – 1,7%, CD117+ – 4,1%,, CD11b+ – 34,7%, CD11c+ – 51,9%, CD16+ – 32,9%. The cells did not express the coexpression of lymphoid cell markers (CD7, CD2, CD19, CD22, cytCD79a).

The patient's final diagnosis was acute myeloid leukemia M4. After three days of induction chemotherapy, recurrence of PG symptoms was observed, patient's general condition began to deteriorate, finally leading to patient's death. A direct cause of death was myocardial infarction. Autopsy showed neutrophilic infiltration of internal organs with focal necrosis of myocardium.

DISCUSSION

The association of pyoderma gangrenosum with haematological diseases was first reported in 1972 by Perry and Winkelmann [3,4]. Our case showed acute leukemia which was preceded by PG. Patients with myeloid malignancies and pyoderma gangrenosum have been observed to have poor prognosis [4]. PG can be an immune complex disorder [1, 5] and the presence of immune complexes has been related to fatal prognosis in acute myeloid leukemia [5]. Cyclosporine and steroids proved to be effective in treatment our patient with PG. Maybe the continuation of steroids and CyA during the chemotherapy would change the fatal prognosis in patients with acute leukemia with coexisting PG? It may be a fact that immune complexes are the aetiological link between atypical PG and poor prognosis myeloid malignancies [6].

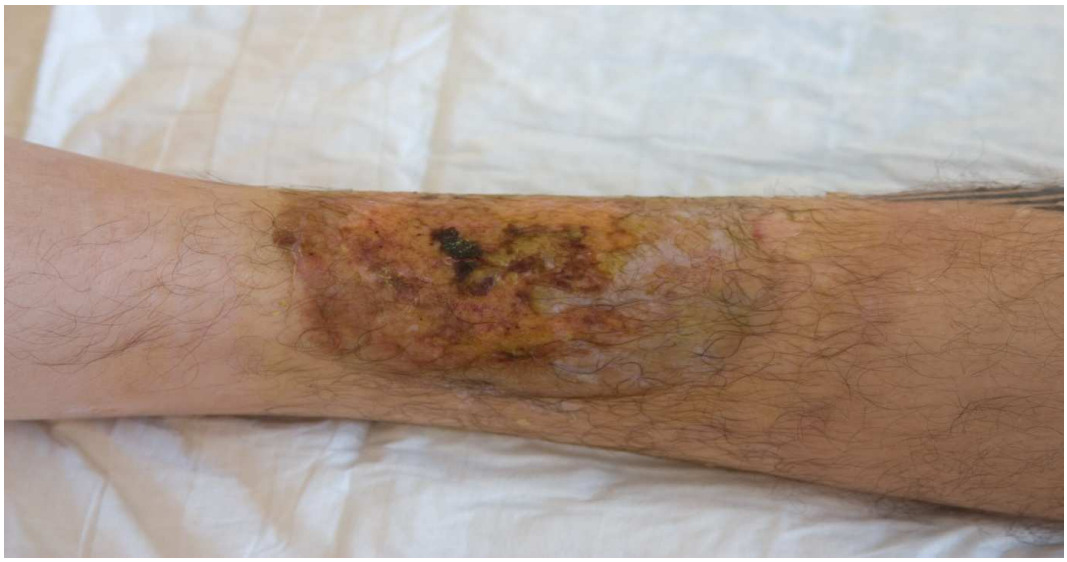


Fig. 1. The smaller ulcer on the anterior surface of right shin (tattoo in the upper side part of tibia).

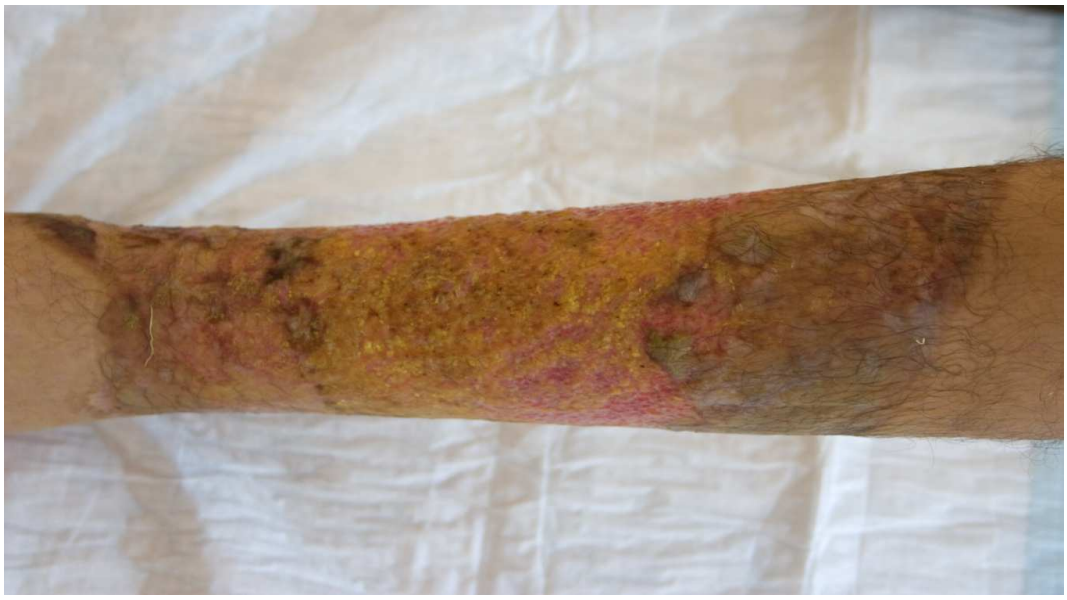


Fig. 2. The enormous ulcer on the left shin (extending from knee to ankle).

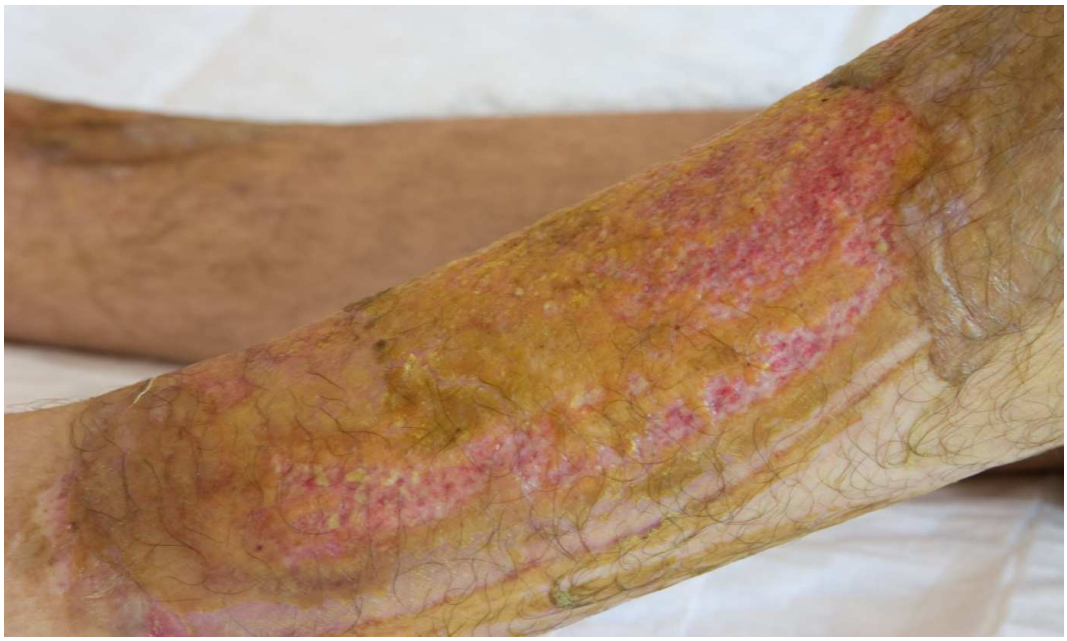


Fig. 3. The left shin in enlargement.

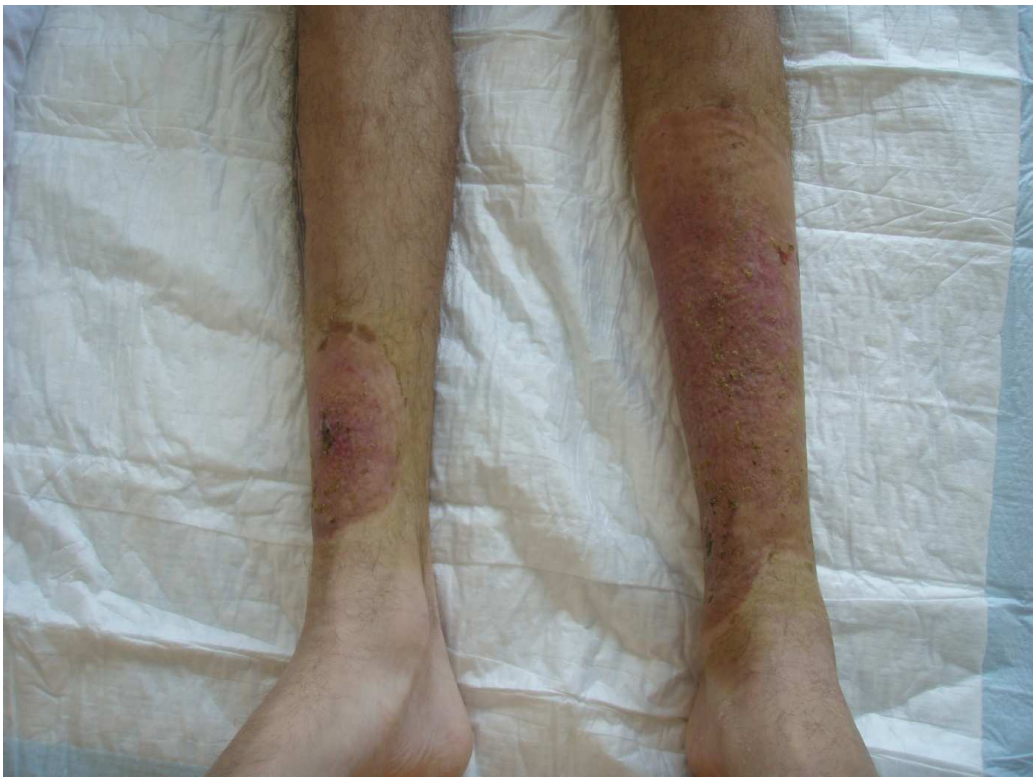


Fig. 4. Both shins after 2-weeks treatment with prednisone and cyclosporine.

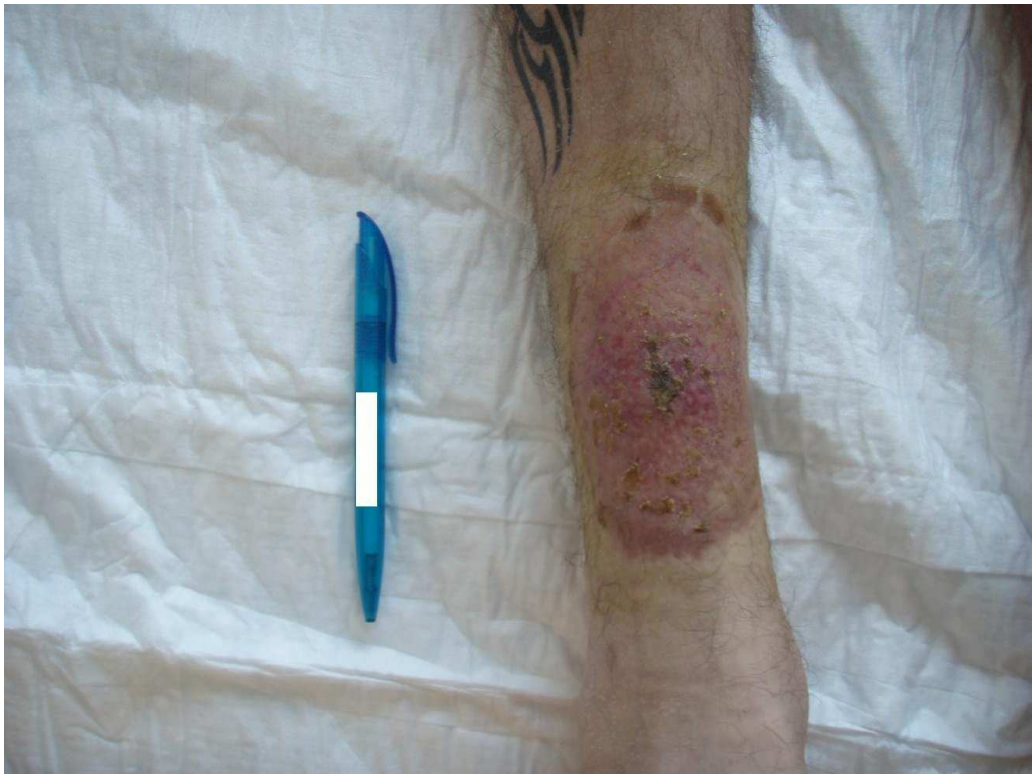


Fig. 5. The right lower leg with the healing ulcer.

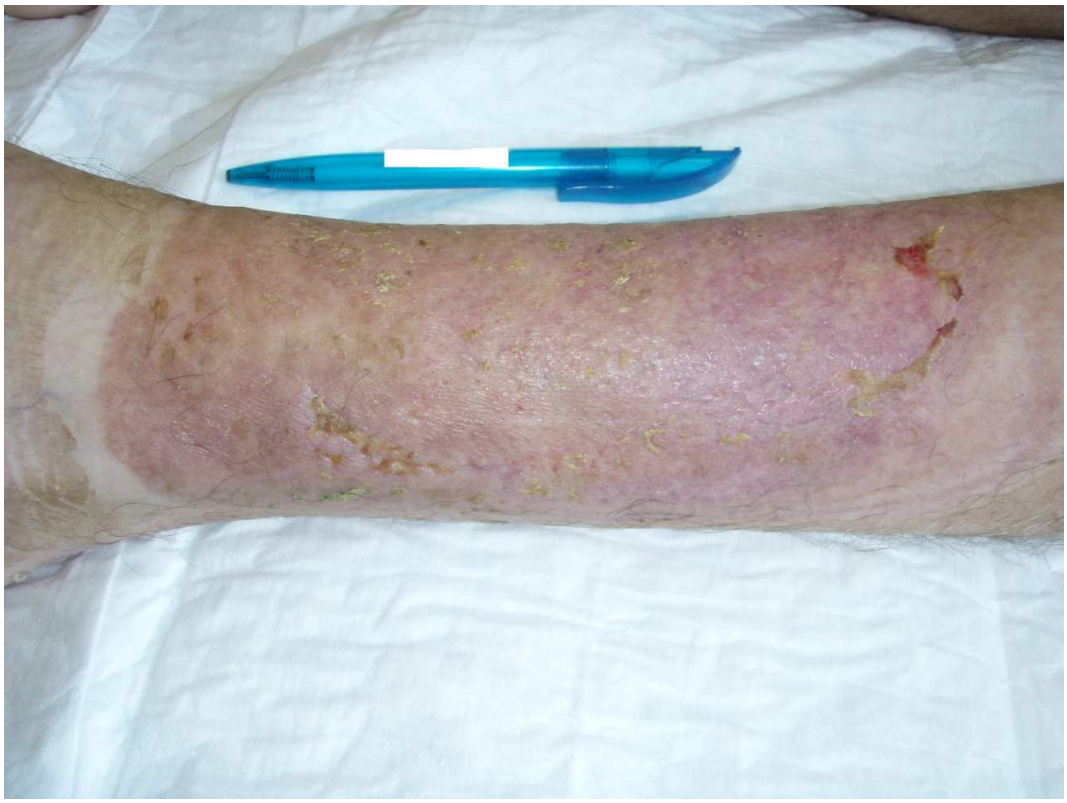
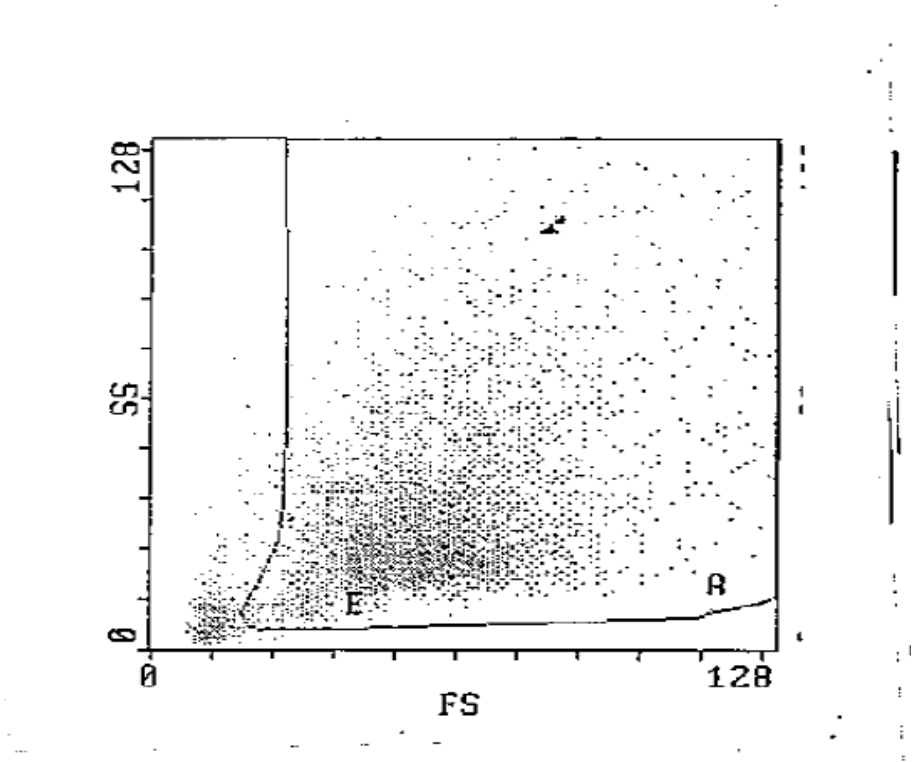


Fig. 6. The left lower leg after treatment.



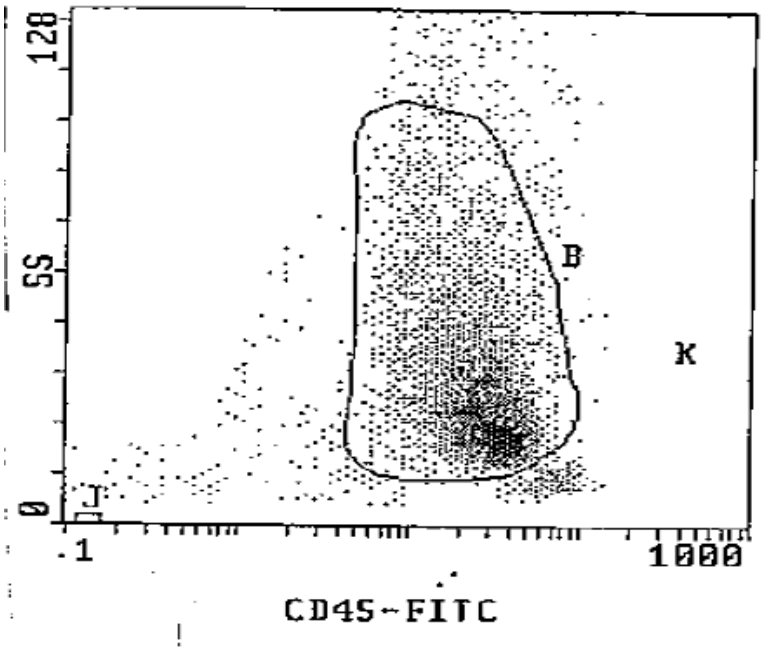


Fig. 7. Flow-cytometry of bone marrow

CONCLUSIONS

1. Pyoderma gangrenosum can precede an acute myeloid leukemia
2. Patients with acute leukemia with coexisting PG have a fatal prognosis.

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Disclosures: No relevant conflicts of interest to declare.

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