

In conclusion, *IRF4* gene analysis should be considered in patients of any age with Waldenstrom's ring LBCL with germinal center origin, follicular and/or diffuse pattern, and strong *MUM1* expression on pathological examination. The presence of *IRF4* rearrangement may affect the prognosis of the disease and the duration of chemotherapy.

Keywords: Large B- cell lymphoma, Waldenstrom's ring, IRF4, MUM1

Anahtar Sözcükler: Büyük B-hücreli lenfoma, Waldenstrom halkası, IRF4, MUM1

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An Unconventional Presentation of Multiple Myeloma: Bazex Syndrome

Multipl Myelomun Olağan Dışı Bir Prezantasyonu: Bazex Sendromu

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To the Editor,

Multiple myeloma patients may exhibit signs and symptoms of skin involvement secondary to either malignant cell infiltration or disease-specific treatment. There are also anecdotal reports

of paraneoplastic skin diseases including Sweet syndrome, leukocytoclastic vasculitis, and neutrophilic dermatosis associated with multiple myeloma [1]. In this report, we share a patient with a remarkably rare skin presentation of multiple myeloma.

A 57-year-old female patient presented to the dermatology clinic with itching, discoloration, hardening, and exfoliation involving the hands, feet, head, and back. Physical examination revealed scalp scaling, hyperkeratosis of all fingernails on both hands and feet, yellowish discoloration, scaly plaques on the palms, thick yellow-gray crusts on the soles, and ichthyosiform appearance of the trunk (Figure 1a). Laboratory evaluation results were as follows: serum creatinine, 1.5 mg/dL; total protein, 14.4 g/dL; albumin, 2.28 g/dL; corrected calcium, 11.74 mg/dL; lactate dehydrogenase, 322 U/L; hemoglobin, 6.9 g/dL; platelets, 103,000/mm³; and urine protein/creatinine ratio, 3.29 g/day. A skin lesion biopsy showed Bazex syndrome (acrokeratosis paraneoplastica) and she underwent an investigation to reveal a possible underlying malignancy. Upper and lower GI endoscopy and FDG PET-CT results were normal. She was referred to our clinic for hematologic evaluation. Due to her anemia, elevated creatinine level, and high total protein-to-albumin ratio, plasma cell dyscrasias were suspected and related tests were ordered. Serum immunofixation electrophoresis showed an IgG lambda monoclonal band. Serum free kappa light chain was <6.50 mg/L, free lambda light chain was 475 mg/L, and β_2 -microglobulin was 14 mg/dL. The patient did not have polyneuropathy, organomegaly, volume overload, endocrinopathy, papilledema, thrombocytosis, or polycythemia and therefore POEMS syndrome was excluded. Bone marrow aspiration and biopsy confirmed plasma cell myeloma (95% infiltration rate, CD38+, CD118+, CD56+, lambda+, kappa-, and Congo stain negative) and FISH was negative for t(14;16), t(4;14), t(11;14), and del(17p13) mutations. The patient was started on bortezomib, cyclophosphamide, and dexamethasone chemotherapy along with topical steroids for the skin lesions

as suggested by dermatology. After 4 cycles, her skin lesions were markedly regressed (Figure 1b). Bone marrow aspiration and biopsy showed remission and the patient was referred for autologous stem cell transplantation.

Bazex syndrome is characterized by hyperkeratosis of the acral regions, mostly seen in men over 40 years of age. Lesions are often seen on the nose and ears, and less frequently over acral areas such as the nails, hands, feet, knees, and elbows. Lesions appear as erythematous, violet-purple, and symmetrically distributed papulosquamous plaques [2,3]. Even though its exact etiology is unclear, possible proposed mechanisms include antibodies against the tumor cross-reacting with skin antigens, tumors secreting growth factors such as TGF- α that alter epidermal proliferation, and genetic susceptibility due to HLA-A3 and HLA-B8 [4]. Bazex syndrome may be associated with various malignancies, mostly with squamous cell carcinomas of the head and neck. However, it has also been shown to be associated with many other malignancies, including hematological malignancies [5].

In our literature search, we were able to find only one report of the association of multiple myeloma and Bazex syndrome [6]. This anecdotal association should be considered in patients investigated for Bazex syndrome.

Keywords: Bazex syndrome, Acrokeratosis paraneoplastica, Multiple myeloma, Paraneoplastic syndrome

Anahtar Sözcükler: Bazex sendromu, Akrokeratozis paraneoplastika, Multipl myelom, Paraneoplastik sendrom

Informed Consent: Verbal and written informed consent were obtained from the patient.



Figure 1. (a) Patient's lesions at presentation; (b) patient's lesions after chemotherapy.

Authorship Contributions

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Case Report: An Infant with Severe Thrombocytopenia Diagnosed with Type 2B von Willebrand Disease Due To a De Novo p.Val1316Met Mutation

Olgu Sunumu: Ağır Trombositopenili Bir Bebekte De Novo p.Val1316Met Mutasyonuna Bağlı Von Willebrand Hastalığı Tip 2B Tanısı

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To the Editor,

Type 2B von Willebrand disease (vWD 2B) is a rare disease that is difficult to diagnose. It usually presents with a normal or decreased platelet count with bleeding tendency. Clinically, it can be confused with immune thrombocytopenic purpura (ITP), which results in improper treatment. Herein, we report a case of an infant with severe thrombocytopenia diagnosed with vWD 2B.

The female infant was found to have some petechiae on her body when she was 6 days old. A routine blood test in a local hospital showed a decreased platelet count of $19 \times 10^3/\mu\text{L}$, a low hemoglobin level of 100 g/L, and a normal white blood cell count of $12.1 \times 10^3/\mu\text{L}$. Serum bilirubin was elevated to 19 mg/dL. Cranial ultrasound showed subependymal hemorrhage and bilateral intraventricular hemorrhage. After treatment

with platelet transfusion, intravenous immunoglobulin, methylprednisolone, and vitamin K1, the baby's bilirubin and hemoglobin normalized and her platelet count increased to $80 \times 10^3/\mu\text{L}$ initially, but it dropped down to approximately $20 \times 10^3/\mu\text{L}$ thereafter.

At the age of 41 days, the infant was transferred to our hospital. On admission, the physical examination was unremarkable except for some ecchymosis on the body. Her family history included no family members with low platelet levels or with bleeding diathesis. Laboratory evaluations were notable for severe thrombocytopenia (platelet count: $16 \times 10^3/\mu\text{L}$) but otherwise normal hemoglobin and white blood cell count values. The mean platelet volume was 13.4 fL (normal range: 7.4-11 fL). A peripheral blood smear revealed nonspecific thrombocytopenia without platelet clumping. The antinuclear antibody investigation was negative, and the prothrombin time, activated