RESIDENT’S FORUM

Peculiar brownish eruption and superficial lymphadenopathy in a middle-aged man

Case report

A 49-year-old Taiwanese male developed many discrete irregularly shaped brownish macules and patches (diameter 0.3–4.0 cm) that had disseminated over his entire body, predominantly on the head and back; these macules and patches persisted for 8 months (Figures 1A and 1B). The lesions had begun on his face and had gradually spread to his trunk and extremities. A computed tomography scan revealed multiple palpable and non-tender lymph nodes in the occipital, periauricular and submandibular regions and in the neck. A systemic physical examination revealed that he was healthy, that he had not lost body weight, and that he was without constitutional discomfort and systemic disease. The histopathologic findings of trunk skin biopsy showed acanthosis with elongation of epidermal rete ridges, basal hyperpigmentation, and moderate perivascular infiltration of predominantly mature plasma cells with a few lymphocytes and histiocytes in the dermis (Figure 2). The immunohistochemical staining revealed that these plasma cells were positive for both kappa and lambda light chains. The pathologic features of the lymph node of the neck revealed reactive follicular hyperplasia with many mature plasma cells in the interfollicular area (Figures 3A and 3B).

Laboratory examination yielded the following remarkable findings: elevated serum total protein concentration (9 g/dL; normal: 6.4–8.4 g/dL), low albumin level (3.6 g/dL; normal: 3.7–5.3 g/dL), and elevated levels of serum immunoglobulins (Igs) G and A (IgG, 3550 mg/dL; normal: 751–1560 mg/dL); IgA, 675 mg/dL (normal: 82–453 mg/dL); and normal IgM level. The patient had mild leukocytosis, elevated erythrocyte sedimentation rate, C-reactive protein level, and lactate dehydrogenase level. Serum protein electrophoresis showed reversed albumin/globulin ratio (A/G: 0.4) and polyclonal gammopathy. Elevated levels of serum kappa (3000 mg/dL; normal: 542–1248 mg/dL) and lambda light chain (1300 mg/dL; normal: 293–725 mg/dL) and absence of monoclonal Ig light chains were observed. Complete blood cell counts revealed absence of anemia, and the biochemistry panel showed normal findings. A trace amount of protein was detected in the urine. Serologic surveys revealed an absence of possible disease with cutaneous predominant plasma cell infiltration, including autoimmune connective tissue diseases (ANA, anti-ds DNA). Moreover, chronic infections such as syphilis and infections with human immunodeficiency virus, hepatitis B virus, hepatitis C virus, Epstein-Barr virus, and Borrelia (Lyme disease) were absent. Plain radiography of the chest, skull, and long bones revealed absence of osteolytic lesions. Ultrasonography revealed that hepatosplenomegaly was absent.

Figure 1 Diffuse discrete irregularly shaped brownish macules and patches, 0.3–4 cm, on the face, neck (A) and back (B). Note the palpable lymphadenopathy on the neck (arrow in A).
Histopathological analysis revealed dense perivascular and periadnexal infiltrates of predominant mature plasma cells with a few lymphocytes and histiocytes in the dermis. Immunohistochemical analysis revealed that the plasma cells are polyclonal, as indicated by the positive staining of both kappa and lambda chains. 

Cutaneous plasma cell infiltration may be observed in a variety of disorders such as connective tissue disorder, plasma cell neoplasm and chronic infections, including syphilis and Lyme disease, but the cutaneous manifestations in these disorders are different from those in CP.

CP usually follows a chronic and benign course without spontaneous resolution and is refractory to various treatments. Most of these patients had no detectable underlying diseases. Systemic plasmacytosis (SP) was first proposed by Watanabe et al in 1986. Cases with SP had more than two organs involved, including skin and lymph nodes. Most authors agree that SP should not be diagnosed in patients if only the skin and lymph nodes are involved. A few reported patients had systemic involvement such as fever, dyspnea, malaise, weight loss, anemia and hepatosplenomegaly. Some had benign conditions initially, which become aggressive years later as lymphoid interstitial pneumonia, squamous cell carcinoma of the anal region and the lung, or T-cell lymphoma.

The treatments for CP have variable efficacy. Topical or intradermal injection of corticosteroids, topical tacrolimus, or administration of topical phototherapy (PUVA), photodynamic therapy, lead to partial remission in cases with focal lesions; while systemic corticosteroids as well as systemic chemotherapy are useful for cases with systemic involvement.

In conclusion, our patient had a typical clinical triad of CP with mature plasma cell infiltrates in the skin and lymph nodes. He was asymptomatic without any other systemic involvement. No evidences of infection or connective tissue disease were noted. Plasma cell neoplasm could be excluded because of absence of cell atypia and monoclonal cells in the skin and blood. He underwent a few sessions of psoralen + UVA (PUVA) therapy, but did not show a remarkable improvement. As per the patient’s request, no further treatment plan was arranged. At the 6-month follow-up, the skin lesions and lymph node inflammation were found to be persistent without any other systemic involvement.

Pei-Chun Chen, Chieh-Shan Wu, Hui-Wen Tseng
Department of Dermatology, Kaohsiung Veterans General Hospital, Kaohsiung, Taiwan

Hui-Hwa Tseng
Department of Pathology and Laboratory Medicine, Kaohsiung Veterans General Hospital, Kaohsiung, Taiwan

*Corresponding author. Hui-Wen Tseng, 386 Ta-Chung 1st Road, Kaohsiung 813, Taiwan. Tel.: +886 7 3422121 4304; fax: +886 7 3468209. E-mail address: hwtseng@vghks.gov.tw (H.-W. Tseng)

References

Received: Aug 27, 2010
Revised: Nov 19, 2010
Accepted: Mar 1, 2011