References


Ofuji papuloerythroderma associated with Hodgkin’s lymphoma

Sir, Ofuji described papuloerythroderma for the first time in 1984.1 He presented four cases of erythroderma with solid papules on the trunk and extremities accompanied by severe itch. A striking phenomena was the lack of skin symptoms in skin folds, later called the ‘deckchair’ phenomenon.2 Older South-east Asian men are more often affected. Lymph node swelling, eosinophilia and hyper IgE are associated findings, as well as malignancies.3

A 44-year-old atopic Surinamese female of Indian descent with no dermatological or other medical history suffered from itching papules on the lower legs first noticed 8 months previously. The skin condition rapidly extended over the whole body except the face and was associated with fever. Over the past 7 years she had visited several tropical countries such as Surinam, Pakistan, India and the Arabian Gulf states. Initially, the symptoms disappeared with oral prednisolone but reappeared a few months later. Subsequently she was admitted to hospital. On presentation we saw, on the extremities and trunk, an extensive eruption of prurigo papules, excoriations, lichenification, generalized hyperpigmentation with small hypopigmented macules and xerosis cutis. The ‘deckchair’ sign was present (Fig. 1a). Peripheral lymph nodes where palpable.

Peripheral blood examination showed IgE levels of 25 000 kU L\(^{-1}\), specific IgE for birch pollen, eosinophilia, and no Sézary cells in peripheral blood smears. Examination of stools for parasites, serological tests for parasites and syphilis, skin snaps on microcillaria and mycobacteria, bone marrow cultures on fungi, mycobacteria and Leishmania were all negative. Virological tests for HIV 1 and 2 were both negative. Epstein–Barr virus (EBV) serology was as follows: IgG positive, IgM negative, EBV anti-EBNA positive, EBV DNA copies were undetectable; the conclusion was of a recent EBV infection with good immunological response. Computed tomography showed extensive lymph node swelling in the mediastinum, in the liver ligaments and along the abdominal aorta. Echo examination revealed echo-weak lesions in the liver parenchyma. Pathological examination of several skin biopsies all showed spongiotic dermatitis, without signs of lymphoma. Bone marrow smears on several occasions were all normal. Axillary and inguinal lymph nodes extirpated at different time points were both negative for malignant cells. A lymph node from the mediastinum showed fibrous tissue, Hodgkin’s and Reed–Sternberg cells (Fig. 1b) which were CD 30 positive, and CD 3, CD 15 and CD 20 negative. The conclusion was of a nodular sclerosing form of Hodgkin’s lymphoma.

On admission, the patient was treated with local class III corticosteroids and systemic antihistamine medication, with little effect. Soon after the diagnosis of Hodgkin’s lymphoma was made, chemotherapy with Adriamycin, bleomycin, vinblastin and bacarbazin was started. This made the eosinophilia disappear and improved the skin symptoms and itch dramatically. After the second chemotherapy treatment, the skin symptoms were almost gone.

In several reports, Ofuji’s papuloerythroderma has been associated with malignancies such as mycosis fungoides,4 B- and T-lymphomas,5,6 and carcinomas.7 Papuloerythroderma has also been reported in association with HIV infection.8 We describe for the first time a patient with Ofuji’s papuloerythroderma associated with Hodgkin’s lymphoma. Furthermore, this patient had recently been infected with EBU. There is increasing evidence that EBV infections can induce lymphoproliferative malignancies such as Hodgkin’s lymphoma.

Figure 1. (a) The deck chair sign. Solid flat papules coalescing in large sheets sparing the skin folds of the neck. (b) Nodular sclerosing Hodgkin’s lymphoma. Mediastinal lymph node showing Sternberg-Reed cells (arrow) (haematoxylin and eosin staining, original magnification \(\times 40\)).
phoma. As no DNA copies of EBV were found it is unlikely that the infection played a role in the pathogenesis of the Hodgkin’s lymphoma. It remains unclear whether the EBV infection had a direct effect on the skin symptoms. It is known that amoxycillin can induce a severe rash during an active EBV infection but in our patient there was no relationship between antibiotic therapy and the onset of skin symptoms. In the literature, we could not trace a report of papuloerythroderma in relation to a recent EBV infection.

It is still debated whether papuloerythroderma is a clinical entity with multiple underlying causative factors including paraneoplasia in lymphoma, or a peripheral T-lymphoma distinct from Sézary syndrome and mycosis fungoides. This case report supports the first hypothesis. During the course of our patient’s illness, no signs of cutaneous lymphoma manifestation could be found histologically nor immunohistochemically, in skin biopsies taken at three different time points. Hodgkin’s lymphoma was manifest in central lymph nodes, but not in peripheral nodes. Papuloerythroderma has been associated not only with different lymphoreticular malignancies but also with HIV infection. Moreover, the incidence of lymphoma in papuloerythroderma seems lower but that of cancer seems higher compared with ordinary erythroderma. In conclusion, we believe that our patient suffered from Hodgkin’s lymphoma, with Ofuji’s papuloerythroderma as a paraneoplastic manifestation.

References

Is the search for serum antibodies to gliadin, endomysium and tissue transglutaminase meaningful in psoriatic patients? Relationship between the pathogenesis of psoriasis and coeliac disease

Sirs, Enteropathy and malabsorption, documented by abnormal jejunal histology with short villi, have been found in patients with psoriasis. Furthermore, six subjects with longstanding psoriasis and gluten intolerance showed marked improvement on a gluten-free diet (GFD). These observations seem worthy of attention, given that psoriasis patients often have increased serum levels of IgA antigliadin antibodies (AGA). These antibodies are a mark of coeliac disease, but they are also seen in IgA nephritis, sickle-cell anaemia, hepatic disorders, juvenile rheumatoid arthritis and autoimmune thyroidism, and in persons who come into contact, usually for occupational purposes, with large amounts of wheat.

The assessment of AGA and other antibodies, such as antireticulin antibodies, antiendomysium antibodies (EMA) and antitissue transglutaminase (tTG) antibodies, is important in the diagnosis and follow-up of coeliac disease and dermatitis herpetiformis. Recent studies have also investigated the possible finding of these antibodies in psoriatic subjects. In a screening study of 302 subjects with psoriasis, 16% had IgA AGA; only one was positive for antireticulin antibodies, and none was positive for EMA. In another study, 10 of 32 (31%) subjects affected by various clinical forms of psoriasis had positive antireticulin antibodies with titre variations related to the extent of the disease. In addition, it has been demonstrated that AGA-positive psoriatic subjects experienced improvement with a 3-month GFD, while worsening was observed with resumption of the ordinary diet.

On the other hand, a pronounced increase of EG2-positive eosinophils in duodenal mucosa associated with elevated serum levels of eosinophil cationic protein (ECP) has been documented in psoriatic subjects without AGA, but no tendency to higher ECP values has been revealed in those with AGA. The increased number of eosinophils in the gastrointestinal tract has been explained as being part of some unknown process that may be related to the development of psoriatic lesions.

To date, the role of tTG, the supposed endomysial autoantigen in coeliac disease, has not been investigated in psoriasis. We performed a study to verify if gluten intolerance is more frequent in Mediterranean subjects with psoriasis than in healthy controls. Sera of 39 psoriasis (22 women, 17...