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SHORT COMMUNICATION

Prurigo Pigmentosa-like Persistent Papules and Plaques in a Patient with Adult-onset Still's Disease

Yung-Tsu Cho and Yi-Hua Liao

Department of Dermatology, National Taiwan University Hospital and National Taiwan University College of Medicine, 15 F, No. 7, Chung-Shan S. Road, Taipei 10002, Taiwan. E-mail: yihualiao@ntu.edu.tw

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Adult-onset Still's disease (AOSD) is a systemic inflammatory disease characterized by high fever, arthralgia, myalgia, skin rash, lymphadenopathy, and splenomegaly. Marked hyperferritinaemia is an indicator of disease activities (1). The typical skin presentation is an evanescent salmon-pink skin rash, appearing with fever episodes, which has been observed in up to 87% of patients (2). In addition, persistent papules and plaques with a variety of clinical manifestations have also been reported in AOSD. We report here a case with a prurigo pigmentosa-like eruption.

CASE REPORT

A 38-year-old woman with a history of Graves' disease, controlled by daily oral hydroxychloroquine with euthyroid status, presented with fever, sore throat, arthralgia and itching skin eruptions for more than 2 weeks. Physical examination revealed fever up to 39.2ºC, enlarged lymph nodes on the neck, and swelling of the ankles and hand joints. In addition, numerous reticulated, erythematous and brownish papules and plaques, which were pruritic and persistent, were found on her anterior chest wall, abdomen and back (Fig. 1A and B). In addition, she also had evanescent urticated eruptions occurring with fever. A skin biopsy from one of the itching persistent plaques on her back revealed parakeratosis, mild acanthosis, some foci of eosinophilic spongiosis, scattered dyskeratotic cells in the upper epidermis, and a mixed dermal perivascular infiltrate composed of lymphocytes, eosinophils and neutrophils (Fig. 1C and D). Leukocytosis (white blood cell count: 17,500/μl (normal 3,540–9,060/μl)), abnormal serum aspartate aminotransferase (359 U/l (normal <31 U/l)) and alanine aminotransferase (363 U/l (normal <31 U/l)), and elevated serum ferritin (2,244 ng/ml (normal 3.0–151 ng/ml)) were noted. Results of renal function tests and the autoimmune profile were unremarkable. Whole body computerized tomography (CT) revealed hepatosplenomegaly and enlarged cervical lymph nodes. The lymph nodes had later been shown to be a reactive hyperplasia by histopathological examinations. Subsequent positron emission tomography (PET) demonstrated intense fludeoxyglucose (FDG) hypermetabolism at bilateral neck, axillary, mediastinal, para-aortic, and inguinal areas, suggesting enlarged lymph nodes, which favoured a diagnosis of AOSD. Serum electrophoresis revealed polyclonal gammopathy. In addition, comprehensive investigations failed to find any infectious source. Therefore, AOSD was diagnosed. She received a 3-day course of 1,000 mg daily intravenous pulse methylprednisolone treatment and systemic prednisolone at a dose of 0.5–1.0 mg/kg/day over a period of 3 weeks. The symptoms, including fever and skin eruptions, improved gradually. She was then given maintenance therapy with methotrexate, hydroxychloroquine and prednisolone. To date, she has had no relapses of the disease for 1.5 years.

DISCUSSION

Despite several characteristic clinical presentations, it is difficult to make a diagnosis of AOSD, due to a lack of pathognomonic histopathological and serological findings. The Yamaguchi criteria are the most commonly used, with high sensitivity and specificity (3). However, exclusion of infectious diseases, other in-

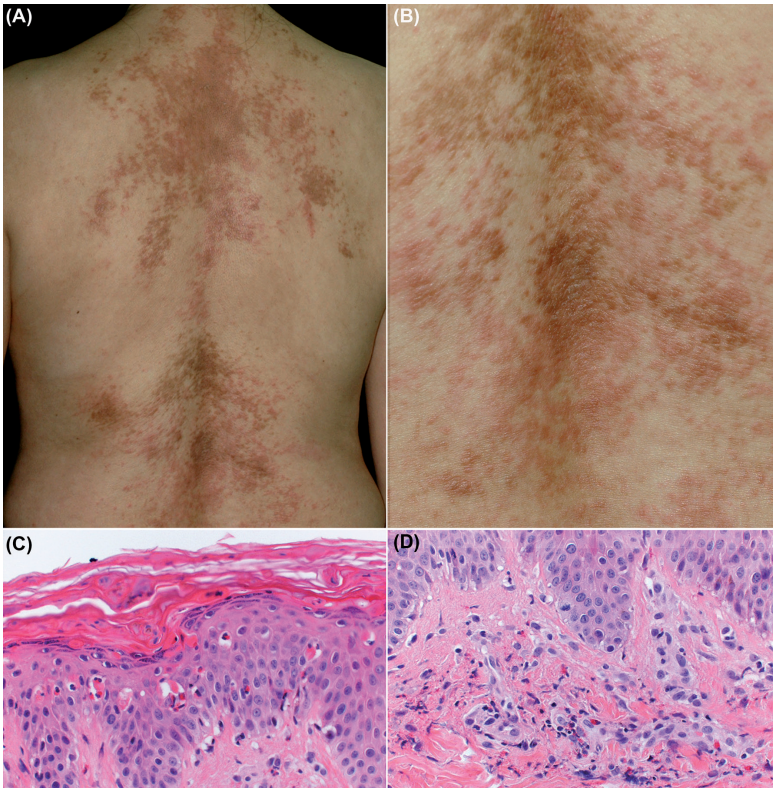

Fig. 1. Prurigo pigmentosa-like persistent papules and plaques in a patient with adult-onset Still's disease. (A, B) Numerous pruritic, reticulated, erythematous and brownish papules and plaques are located on the anterior chest wall, abdomen, and back. Histopathology examination from one of the itching plaques on her back showed parakeratosis, mild acanthosis, some foci of eosinophilic spongiosis, scattered dyskeratotic cells in the upper epidermis, and mixed perivascular infiltrations composed of lymphocytes, eosinophils and neutrophils in the upper dermis. (C, D) H&E; original magnifications × 200.

flammatory diseases, and malignancies is still required before a diagnosis of AOSD to be made. On the other hand, more and more evidence has demonstrated that persistent papules and plaques of AOSD have important diagnostic values (4). The reported rates of persistent eruptions in AOSD patients varied from 25% to 78% (5–7). Clinically, it usually appeared at the onset of the disease and presented as pruritic, erythematous or brownish, urticarial or flat-topped papules and plaques over the trunk, face, neck, and extensor surface of the extremities (6). However, several uncommon presentations have been reported (8), such as fixed plaques or persistent generalized erythema. In spite of various clinical presentations, the histopathological findings are characteristic, including scattered or grouped dyskeratotic cells in the upper epidermis and a dermal infiltrate containing neutrophils, which usually exclude other differential diagnoses (6, 7).

The cutaneous lesion of our patient was unique. The reticulated erythematous and brownish papules and plaques on the chest, abdomen and back resemble those seen in prurigo pigmentosa, which is a severe pruritic skin disease with a characteristic skin presentation and a predilection in young adulthood (9). In addition, the histopathological findings included features seen in both AOSD and prurigo pigmentosa. In persistent papules and plaques of AOSD, scattered or grouped dyskeratotic cells in the upper epidermis are diagnostic, but eosinophil infiltration was seldom seen (6, 7). However, in fully developed lesions of prurigo pigmentosa, eosinophils and lymphocytes may predominate in upper dermal and epidermal infiltrations, along with scattered dyskeratotic cells at the basal layer (10). In addition, mild epidermal hyperplasia and neutrophil infiltrations could be seen in both diseases. Although there is a case report describing a patient with AOSD having clinical and histological presentations compatible with prurigo pigmentosa (11), less characteristic histopathological pictures of persistent eruptions of AOSD, including scattered or grouped dyskeratotic cells in the upper epidermis and a dermal infiltrate containing neutrophils, could be found in that case.

The reason why AOSD patients presented with prurigo pigmentosa-like persistent eruptions is uncertain (11). Interleukin-6 (IL-6) was found to be significantly increased in the lesional skin of prurigo pigmentosa (12), and IL-1, IL-6, IL-18 and tumour necrosis factor-α have been shown to be associated with AOSD (4). Interestingly, IL-6 has been shown to correlate with disease activity (13), and a novel IL-6 antagonist, tocilizumab, has been proved to be a promising treatment for AOSD (14). Induction of clinical remission by systemic anti-inflammatory therapies in our patient may provide indirect evidence that systemic inflammation in AOSD may contribute to the formation of prurigo pigmentosa-like eruptions. Further investigation is needed to confirm our observations.

The authors declare no conflicts of interest.

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