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Rika Sato, Yukiko Kiniwa, Takushi Shirai, Ryuhei Okuyama

Dans European Journal of Dermatology 2019/3 Vol. 29 , pages 331 à 332
Éditions JLE

ISSN 1167-1122
DOI 10.1684/ejd.2019.3551
Date de mise en ligne : 23/09/2024

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doi:10.1684/ejd.2019.3541

Sarcoidosis mimicking malar rash: a case of lupus pernio

Cutaneous sarcoidosis is known as "the great imitator" [1] and exhibits a variety of macroscopic presentations such as maculopapular lesions, subcutaneous nodules, plaques, and lupus pernio, which is one of the most characteristic features. We describe a case of lupus pernio on both cheeks that mimicked a butterfly rash.

A 43-year-old Japanese man noticed subcutaneous nodules on his upper limbs five years ago. The nodules were suspected to be sarcoidal granulomas based on skin biopsy at another hospital, but regressed spontaneously within a few years. Two years later, the patient noticed reddish swelling on his cheek and subcutaneous nodules on the upper limbs, and was referred to us. Physical examination revealed diffuse erythema with induration on the bridge of the nose and both cheeks (*figure 1A*). There were several subcutaneous nodules of 1-2 cm in size on both upper limbs. He had no extracutaneous manifestations and was afebrile.

Skin biopsy of the right cheek revealed multiple non-caseating granulomas with a few Langhans-type giant cells throughout the whole dermis and subcutis with fibrosis (*figure 1B-C*). Some granulomas were surrounded by lymphocytes and eosinophils. Neither interface dermatitis nor atypical lymphoid cells were present. Laboratory test results were as follows: normal blood cell count, calcium at 9.4 mg/dL (normal range: 8.7-10.3), complement component 3 at 113 mg/dL (86-160), anti-nuclear antibody (homogenous pattern) at 1:40 (<1:40), soluble IL-2 receptor at 1,466 U/mL (135-421), and angiotensin converting enzyme at 24.0 U/L (8.3-21.4). Chest computed tomography revealed lymph node enlargement in the mediastinum and small nodules in both lungs. Bronchoalveolar lavage fluid contained an infiltrate predominantly made of macrophages and lymphocytes (62.4% and 34.1%, respectively), with an increased CD4/CD8 ratio (6.28; normal <3.5). Needle biopsy of a subcarinal lymph node obtained during bronchoscopy also revealed granulomatous changes. Normal sinus rhythm without signs of cardiac ischaemia was found on the electrocardiogram. Ophthalmological examination showed bilateral uveitis. Based on these findings, the patient was diagnosed with sarcoidosis. To prevent systemic involvement, he was treated with 30 mg/d of oral prednisolone (PSL). After a month of treatment, the erythematous induration started regressing gradually without deformity (*figure 1D*), the subcutaneous nodules on the upper limbs began decreasing and the soluble IL-2 receptor level normalised (256 U/mL). PSL was tapered to 5 mg/day over six months, and the disease is being controlled.

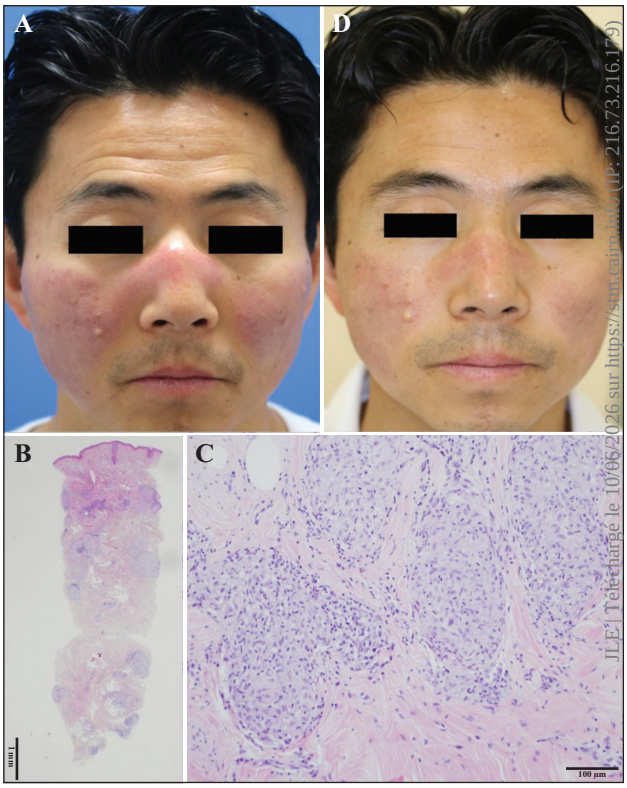

Figure 1. A). Clinical features on the first visit, including indurated erythema on the bridge of the nose and both cheeks. **B**) Histopathological features of the right cheek; high-power magnification shows non-caseating granulomas with inflammatory cell infiltrates and fibrosis in the dermis and subcutis (haematoxylin-eosin staining; ×20). **C**) Low-power magnification showing non-caseating granulomas, some of which are surrounded by lymphocytes and eosinophils (haematoxylin-eosin staining; ×200). **D**) Improvement of indurated erythema on the nose bridge and bilateral cheeks is seen after treatment with oral prednisolone (30 mg/d).

Lupus pernio has been reported in 2-6% of all cases of cutaneous sarcoidosis [1-3]. In this type, the deep tissues are commonly affected, which may lead to deformities and/or functional impairment [1]. To avoid these complications, therapeutic intervention before disease progression may be needed, and oral PSL is the first-line treatment [3]. The present case was successfully treated by a moderate dose of PSL.

Malar rash is a characteristic skin manifestation of systemic lupus erythematosus, but occurs in several other conditions, including rosacea, seborrheic dermatitis, dermatomyositis, pellagra, erysipelas, Lyme disease, photosensitivity, and Bloom syndrome [4]. The macroscopic features of lupus pernio generally include violaceous or erythematous

plaques with induration, and are distributed on the central face, *i.e*. the nose, cheeks, earlobes, lips, and forehead [1]. The most common localisation of lupus pernio is the nose, moreover, in most of the reported cases with nasal involvement, the lesions affect the tip of the nose [1, 2]. Our patient presented with a unique feature of symmetrical bilateral erythematous induration on the cheeks across the nasal bridge, sparing the tip of the nose, thereby mimicking a butterfly rash. This observation shows that lupus pernio may present with a malar rash. ■

Disclosure. *Financial support: none. Conflicts of interest: none.*

Department of Dermatology,
Shinshu University School of Medicine., 3-1-1 Asahi, Matsumoto, Nagano 390-8621, Japan
<ykiniwa@shinshu-u.ac.jp>

Rika SATO
Yukiko KINIWA
Takushi SHIRAI
Ryuhei OKUYAMA

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doi:10.1684/ejd.2019.3551

Acantholytic Bowen's disease histopathologically showing the Borst-Jadassohn phenomenon

Acantholytic squamous cell carcinoma (SCC) is a rare variant of invasive SCC. We report a case of acantholytic Bowen's disease (SCC *in situ*) histopathologically showing the Borst-Jadassohn phenomenon.

A 77-year-old Japanese woman visited us in November 2016 with an asymptomatic lesion on the left leg, which she had noticed two years previously. Physical examination revealed a dark reddish plaque, 1 cm in diameter, on the malleolus medialis of the left leg (*figure 1A*). An incisional biopsy was taken from the plaque. Haematoxylin and eosin staining showed elongated and thick rete ridges, and islands within the epidermis (*figure 1B*). The islands showed acantholysis and were composed of acantholytic atypical keratinocytes, including clumping, mitotic, and dyskeratotic cells (*figure 1C, D*). The lesion was excised with 5-mm margins. No invasion of the tumour cells into the dermis was found. Immunohistochemical studies revealed that the tumour cells were positive for keratin (K) 5, K6, p53, p63 (*figure 1E*), and epidermal growth factor receptor (EGF-R) (*figure 1F*), but were negative for S-100 protein, K7, carcinoembryonic antigen (CEA), and CAM5.2. The rate of Ki-67 positivity was 50% (*figure 1G*). Scant CD117+ cells were present within tumour nests.

The Borst-Jadassohn phenomenon manifests with well-defined islands of keratinocytes embedded within the epidermis [1-3]. The cells of the islands differ in their appearance from the surrounding keratinocytes. This phenomenon may be seen in benign tumours, such as seborrheic keratosis [2] and poroma, and in malignant tumours *in situ*, such as Bowen's disease [1, 3].

The tumour nests in this case did not show trichilemmal, sebaceous or ductal differentiation. Differential diagnoses included porocarcinoma *in situ*, malignant hidroacanthoma simplex, ectopic extramammary Paget's disease, actinic keratosis, and seborrheic keratosis. Porocarcinoma *in situ* and malignant hidracanthoma simplex commonly express CEA and CD117 [4], and were excluded because of the absence of ductal differentiation and negative immunohistochemistry for CEA and limited expression of CD117. Ectopic extramammary Paget's disease, normally expressing K7 and CEA [5], was excluded because of the lack of expression of K7 and CEA. Actinic keratosis was excluded because of the absence of atypia in the epidermal basal cells and the absence of solar elastosis in the dermis.

A strong expression of EGF-R, Ki-67, p53 and p63 was consistent with both clonal Bowen's disease and clonal seborrheic keratosis; lesions which may show the Borst-Jadassohn phenomenon [1]. The acantholytic variant of seborrheic keratosis may contain dyskeratotic cells [6], however, the presence of clumping cells and multiple mitotic cells, as well as acantholytic atypical keratinocytes, in the epidermis suggested the diagnosis of acantholytic Bowen's disease.

Acantholytic SCC is characterized by atypical keratinocytes associated with loss of cohesion between epidermal or adnexal epithelial cells with formation of clefts [7]. Ogawa *et al*. classified 115 cases of acantholytic invasive SCC and SCC *in situ* into (1) acantholytic SCC, follicular type; (2) acantholytic SCC, follicular pattern; (3) acantholytic actinic keratosis (AK); and (4) acantholytic SCC arising from AK [7]. Ogawa *et al*. defined acantholytic SCC of follicular type as involvement of follicular epithelium but not epidermis, and acantholytic SCC with follicular pattern as involvement of both follicular epithelium and the epidermis [7]. However, acantholytic Bowen's disease was not included in the proposed category of acantholytic SCC and SCC *in situ*. Pai *et al*. reported a case of SCC that was probably derived from an acantholytic variant of Bowen's disease [8]. Previous cases [7, 8] and our case suggest that acantholytic SCC *in situ* is classified under acantholytic AK and acantholytic Bowen's disease, and that both types may progress to SCC.

Our case is a rare type of acantholytic Bowen's disease histopathologically showing the Borst-Jadassohn phenomenon; it emphasizes the importance of histopathological and immunohistochemical examination to diagnose the tumour accurately and suggests that an updated classification of acantholytic SCC and SCC *in situ* is required. ■

Disclosure. *Financial support: none. Conflicts of interest: none*