The association of pyoderma faciale and erythema nodosum

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Summary

Pyoderma faciale (PF) is a sudden severe eruption of pustules and cystic swellings which may be interconnected by sinuses. It affects mainly adult women. We report the case of a 21-year-old woman presenting with sudden onset of coalescing nodules and abscesses on the face, with mild systemic disturbance. She also had erythema nodosum (EN)-like lesions on the legs. To the best of our knowledge, this is the first reported case of PF associated with EN.

Pyoderma faciale (PF) is a rare disorder originally described by O’Leary and Kierland in 1940.1 Plewig et al.2 have proposed it be renamed ‘rosacea fulminans’ but rosacea is unlikely to be the basis for the condition in every patient. There is still much debate on this issue, and further clinical and basic research is required on this distressing facial dermatosis.3

Report

A 21-year-old woman presented with sudden onset of facial nodules and abscesses in January 2004. She had a 5-year history of erythematous papules and pustules treated with tetracyclines, antiandrogenic compounds, topical tretinoin and clindamycin, with partial to good response.

In November 2003, antiandrogenic compounds (Dianette®; Schering AG, Berlin, Germany) and topical clindamycin were reintroduced. One month later, following a stressful event, painful nodules and cysts appeared on the patient’s face. Oral clindamycin 150 mg twice daily was started with no response. New lesions appeared around the nose and on the forehead. After 20 days, tender erythematous nodules appeared on the patient’s shins, accompanied by abdominal pain. Treatment was changed to doxycycline 100 mg twice daily, but the facial and leg lesions worsened.

When referred to our clinic, the patient was afebrile and in good general health. There were numerous erythematous papules and pustules with inflamed nodules on the chin, nose and forehead (Fig. 1). Some papules and pustules were noted on the back. The

Figure 1 Numerous papules and pustules with inflamed nodules on the patient’s chin, nose and forehead.

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The patient did not experience flushing. Comedones were not common on the face and back. Tender, red and slightly elevated nodules up to 40 mm in diameter were observed on the shins (Fig. 2). In addition, the right ankle was mildly tender and swollen. The patient had no lymphadenopathy or abdominal tenderness, and no organomegaly was detected on examination.

Laboratory studies revealed an erythrocyte sedimentation rate of 51 mm/h [normal range, (NR) 0–25], haemoglobin 12.7 mg/dL (NR 12–16), leucocytes 9400 cells/mm³ (NR 4000–10 000) with 81.7% neutrophils (NR 48–73). Liver, renal and thyroid function tests were within normal limits. Chest X-ray and purified protein derivative test were normal. Direct smear from the facial pustules showed many white blood cells and few Gram-positive cocci.

The clinical impression was PF, thus treatment with oral prednisolone 0.5 mg/kg/day and erythromycin 400 mg four times daily was started. There was significant improvement of the facial lesions after 2 weeks (Fig. 3) and only postinflammatory hyperpigmentation remained on the legs. Erythromycin was discontinued, oral isotretinoin 20 mg twice weekly (about 1 mg/kg/day) was started, and prednisolone was gradually tapered to cessation over approximately 2 weeks. There was some flare of the facial lesions after discontinuation of prednisolone, but erythema nodosum (EN) lesions did not recur on the limbs.

Although PF is also known as rosacea fulminans, it is not yet clear whether this condition is a variant of rosacea or acne vulgaris or a separate entity.4 It is uncommon, and occurs in patients who usually have mild skin disease, which suddenly erupts, producing many pustules and nodules, especially on the face. It mainly affects postadolescent women (aged 20–40 years), often following a period of stress. Comedones are rare, and facial flushing frequently precedes the acute illness. In contrast to acne fulminans, there are usually no systemic symptoms. The reason for the sudden flare is unknown. Rosacea fulminans has been reported in association with Crohn’s disease (CD) and high-dose vitamin B supplements.3

The clinical findings in our patient were in favour of PF: sudden eruption of numerous papules and pustules

Figure 2 Erythema nodosum-like lesions on the shins.

Figure 3 Dramatic improvement of the facial lesions 2 weeks after treatment.
without significant comedones, after a stressful event. She was afebrile and in good general health, and leucocytosis was not detected. She did not have acne conglobata, which is found particularly in males; the lesions usually occur over most of the trunk and upper limbs, and facial lesions are less common. She did not also have any marked systemic disturbance such as fever, polyarthropathy, marked leucocytosis, weight loss, anorexia or general malaise, which are seen in acne fulminans; patients are predominantly young men who quite suddenly develop extensive inflammatory lesions, especially on the trunk.3

Of particular interest is the EN accompanying the PF in our patient. The pathogenesis of EN has not yet been fully clarified, but it is believed to involve an allergic or immune complex-mediated reaction to a wide variety of antigens. The most prevalent triggering factors of EN are infections, sarcoidosis, lymphoma, CD, drug reactions and pregnancy; however, in up to 50% of the cases the aetiology remains unknown.5 In our patient, PF could be considered the trigger of EN. She was using clindamycin before EN presentation, so this drug may have a role.

Although to the best of our knowledge, the association of PF and EN has not been reported previously, EN in the setting of acne fulminans (AF) is well documented in several reports.6–9 Kellett et al.6 suggested that two of their three patients with EN and AF appeared to have had the condition triggered by treatment with isotretinoin. They were found to have circulating immune complexes. Tan et al.,7 reporting a similar case, mentioned that despite continuing isotretinoin, the acne and EN improved together with dapsone treatment, so EN was not related to isotretinoin, but to AF. Williamson et al.8 found an excessive intradermal response to Propionibacterium acnes in their case and proposed the occurrence of an Arthus reaction.

The best treatment for PF is perhaps oral isotretinoin for 4–6 months. To minimize an acute exacerbation, oral prednisolone should be prescribed initially before starting isotretinoin, which can then be taken at the same time.9 Our patient improved with this treatment protocol, but showed some flare of the facial lesions when prednisolone was being tapered.

Because EN is regarded as a nonspecific immunological response to a variety of stimuli, we suggest that PF should be added to the list of possible factors triggering the condition.

References