A case of multiple deep and necrotic ulcers on the face

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Case report

A 31-year-old male presented with painful facial ulcers over bilateral cheeks and the forehead for the past 2 weeks (Figure 1). Recent contact history included superficial chemical peeling with glycolic acid and self-squeezing of his cystic acne. The ulcers were shallow, clean and had a relatively broad base, with particular lesions over the left face arranged in a bizarre linear pattern. Self-induced pyoderma was suspected. However, the ulcers did not improve with antibiotic treatment. One month later, the patient was admitted under the impression of pulmonary tuberculosis and meningitis because of fever, cough, yellowish sputum, headache and nuchal rigidity for 3 weeks.

Routine blood sampling revealed significant leukocytosis (19.2 × 10⁹/L), elevated C-reactive protein levels (149.83 mg/L [1427 nmol/L]), elevated liver functions (glutamate oxaloacetate transaminase, 94 IU/L; glutamic pyruvic transaminase, 121 IU/L), elevated erythrocyte sedimentation rate (43 mm/hr), pyuria and microscopic hematuria. X-ray and computed tomography revealed multisinus sinusitis and lung cavitations in the bilateral lung apex. Bronchoscopy showed inflammatory change over bilateral bronchi.

Further autoimmune investigation demonstrated elevated levels of anti-neutrophil cytoplasmic antibody with antiproteinase 3 specificity (C-ANCA, 502 U/mL) (reference range: <7 U/mL), negative perinuclear(p)-ANCA, and elevated rheumatoid factor (RA factor, 293 IU/mL) (reference range <301U/mL).

During admission, the patient developed pain over the elbow joints, shoulders and thighs. The facial ulcers became more necrotic and deeper with purulent yellowish discharge, with an undermined cavity and purpuric overhanging border; they were associated with severe pain (Figure 2). These symptoms were suggestive of malignant pyoderma.¹⁻³ In addition, new ulcers developed on bilateral shoulders. Aerobic, anaerobic, and mycobacteria wound pan-cultures were negative. Skin biopsy at the edge of the ulcer showed massive destruction of the dermis, many multinucleated giant cells, diffuse lymphocytic infiltration, and extravasation of erythrocytes (Figure 3).

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Figure 1  Painful facial ulcers over bilateral cheeks and the forehead in November 2008. The photos indicates acne excoriée.

Figure 2  (A) In December 2008, the ulcers became deep and necrotic with purulent yellowish discharge, and there was an undermined cavity, purpuric overhanging border and severe pain. (B) Left shoulder and (C) the left side of the face.
Wegener’s granulomatosis (WG) is a rare disease and occurs most often in Caucasians with a slight male predominance. The most prominent feature is necrotic granulomatous vasculitis affecting a wide range of organ systems including lungs, kidneys, musculoskeletal system, the skin, and the central nervous systems. It classically is composed of a triad of granulomatous inflammation of the upper and lower respiratory tracts, and systemic necrotizing small vessel vasculitis. Cytoplasmic ANCA directed against the antigen proteinase 3 occurs in up to 80% of patients with WG.

A list of differential diagnosis must be considered including infectious entities, facial acne related dermatosis, and malignancy. Infectious entities can be excluded by repeated pan-cultures. Previous acne and skin peeling history brought about by disease entities such as acne excoriée and acne fulminans should be taken into consideration; malignancy including natural killer/T cell lymphoma, characterized by ulceronecrotic facial lesions, is also a possibility. Acne excoriée is a psychological disorder with neurotic excoriation of acne lesions. Mild acne may be accompanied by extensive excoriation. Acne fulminans is the abrupt onset of nodular and suppurative acne in association with systemic symptoms. However, in our case, findings from the skin biopsy did not support the diagnosis of acne-related diseases or lymphoma. The natural course of disease was eventually demonstrated as WG.

Skin lesions occur in WG in 40–50% of patients and 8.6% have skin as the first manifestation. Palpable purpura and papulonecrotic lesions are the most common, both with a predilection for the lower extremities. Specific skin findings of WG have a prognostic value because they tend to occur early during the course of the disease, and are related to a much higher prevalence of the severe systemic form of WG. In our case, the patient did not have lesions on the extremities. Instead, there was a bizarre presentation of malignant pyoderma-like multiple ulcerated lesions on bilateral cheeks, glabellas, chin and a small lesion on the shoulder. The oral mucosa showed classic presentation of WG because the gingiva was hyperplastic and periodontitis was present.

Pathological examination of WG mostly shows nonspecific findings such as perivascular lymphocytic infiltrates. Up to 50% of cases demonstrate leukocytoclastic vasculitis or granulomatous inflammation. In our case, there was diffuse necrotizing granuloma with no obvious vasculitis observed.

The term malignant pyoderma was first described in 1968 by Perry et al. The condition is characterized by ulcers with irregular borders, a progressive course, undermined cavities, peripheral purpuric erythema and purulent discharge located on the head and neck region. It is often first regarded as an infection-related disease entity, but antibiotics are not beneficial and pan-cultures reveal negative results. It has been repeatedly shown to be associated with WG. Treatment with cyclophosphamide, methotrexate and prednisolone can significantly improve the otherwise fatal outcome of WG. Appropriate workup should be performed when such skin manifestation is encountered.

References