A 37 year-old male presented with a 5-year history of recurrent erythematous, demarcated plaques of the right malar region that underwent progressive suppuration. The lesions evolved into a superficial ulcer with atrophic scarring. Despite treatment with numerous antibiotics, the lesions did not improve. Skin biopsy revealed a non-specific granulomatous folliculocentric reaction. Specific stains and tissue cultures were negative for infectious agents. An exclusion diagnosis of superficial granulomatous pyoderma was made and the patient was treated with use of cyclosporine combined with prednisolone with good response; the lesion healed within 3 months.

A 37 year-old male was admitted to our dermatology outpatient clinic with a 5-year history of recurrent erythematous, demarcated plaques of the right malar region that underwent progressive suppuration. The lesions evolved into a superficial ulcer which healed with atrophic scarring (Fig. 1). He denied any antecedent trauma, the lesions had developed de novo. Apart from these lesions the patient had no other complaints. He had a free medical history and no disease had been detected during clinical examination.

Because of the purulent nature of the lesions, attempts were made to culture specific organisms. The lesional swabs were negative for infectious agents. Cultures for acid-fast bacilli and leishmania, as well as routine cultures proved sterile. No cocci, bacilli, fungi or parasites were seen with Ziehl-Neelsen, periodic acid Schiff and Giemsa stains. Laboratory evaluation included renal and liver serum markers, hepatitis serology, protein electrophoresis, cryoglobulins, rheumatoid factor, antinuclear, antineutrophil cytoplasmic and antiphospholipid antibodies, syphilis serology and urine analysis, which were all within normal limits. Other diagnostic tests such as chest X-ray and endoscopic examinations were performed to exclude the presence of an underlying systemic disease.

In spite of a lack of laboratory evidence of infection, because the lesions were apparently purulent, trials of antibiotics were given as in deep impetigo (ecthyma). Patient was treated by several dermatologists with cefadroxil, ofloxacin, fusidic acid, cefradine, amoxicillin-clavulanate, minocycline, roxithromycin, azithromycin, ciprofloxacin, doxycycline and clindamycin intermittently for 5 years but all failed to alter the clinical appearance of the lesion. Itraconazole and rifampicin for possible deep mycosis or cutaneous tuberculosis had been given as well.
A skin biopsy specimen from the lesion was taken. The findings were compatible with a non-specific granulomatous folliculocentric reaction. Specific stains and tissue cultures were negative for infectious agents. An exclusion diagnosis of superficial granulomatous pyoderma (SGP) was made.

The patient was admitted and treatment with oral corticosteroid (prednisolone 50 mg daily) in combination with cyclosporine (approximately 3 mg/kg daily) and topically pimecrolimus cream was initiated. Four weeks later, his examination at a follow-up visit revealed marked improvement (Fig. 2). After three months of treatment the lesions were completely healed. The dose of prednisolone was tapered down gradually and withdrawn, while cyclosporine was reduced to 100 mg daily. The patient was followed up for 2 years while continuing cyclosporine at 100 mg daily (Fig. 3) with no relapse.

Superficial granulomatous pyoderma (SGP), also known as vegetative pyoderma gangrenosum, first described by Winkelmann et al., is an unusual, chronic variant of pyoderma gangrenosum. It presents as a superficial ulcer that is not painful and often lacks the typical violaceous undermined border. It is often a solitary lesion, especially on the trunk, but uncommonly, as in our case, presents on the face. Unlike patients with classical pyoderma gangrenosum, many patients with superficial granulomatous pyoderma do not have an associated systemic disease. However, Langan and Powell reported that 18% of patients with SGP may have additional diseases including diabetes mellitus, rheumatoid arthritis, chronic lymphocytic leukemia, polycythemia vera, paraproteinememia and sarcoidosis.

Suppurative granulomatous infections should be considered as the main histological differential diagnosis of SGP, such as those caused by mycobacteria or deep-seated fungi. SGP is an exclusion diagnosis, therefore other causes of cutaneous ulceration need to be ruled out. No specific laboratory or histopathological features exist. There should be particular effort to find an infective etiology as the treatment involves potent immunosuppressive therapy.

Treatment should be started as soon as possible to avoid disease progression and permanent disfigurement. Treatment modalities for SGP include the use of antibiotics, dapsone, mycophenolate mofetil, oral corticosteroids and cyclosporine. Lachapelle et al. have suggested that cyclosporine should be a first-line treatment for this condition. This case confirms that oral cyclosporine is a useful treatment for SGP. The lesion in our patient healed within 3 months of treatment with cyclosporine in combination with prednisolone.

REFERENCES