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CASE REPORT

Acne fulminans successfully treated with prednisone and dapsone *

Acne fulminans tratada com prednisona associada à dapsona

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Abstract: Acne fulminans is a rare condition and the most severe form of acne. It involves the sudden onset of febrile and multisystemic symptoms, with poor response to ordinary therapy in patients who previously had mild to moderate acne. It is characterized by hemorrhagic ulcerative crusting lesions on the face, chest and upper back. The authors report a case of acne fulminans that was successfully treated with oral prednisone and dapsone.
Keywords: Acne vulgaris; Arthritis; Dapsone; Prednisone

Resumo: A acne fulminans é uma condição rara e a mais severa forma de acne. Manifesta-se com um quadro agudo, febril e multissistêmico, resistente à terapêutica convencional em doentes com antecedente de acne leve ou moderada. As lesões são caracteristicamente úlcero-hemorrágicas e acometem preferencialmente tórax e face. Os autores relatam um caso de acne fulminans com excelente resposta terapêutica ao tratamento empregado.
Palavras-chave: Acne vulgar; Artrite; Dapsona; Prednisona

INTRODUCTION

Acne *fulminans*, also known as acne maligna, is a rare and severe form of acne *vulgaris* that usually affects young adults, predominantly Caucasian adolescent males, aged 13-16 years. It is characterized by the sudden onset of hemorrhagic ulcerative crusting acne on the face, chest and upper back, associated with systemic symptoms such as fever, weight loss, arthralgia and myalgia in patients with a preceding history of mild to moderate acne. [1-3] The authors report a case of an adolescent male with acne *fulminans* presenting with severe lesions that was successfully treated with oral prednisone and dapsone.

CASE REPORT

A 15-year-old man, who had been previously diagnosed as having Grade I acne *vulgaris*, suddenly developed severe worsening of the acne lesions on the neck, arms, chest and upper back. Associated with cutaneous lesions, he also reported fever, chills, arthralgia and myalgia, whose intensity did not allow locomotion and spontaneous movements in bed.

At physical examination, he presented papules, pustules and crusts on the face, neck and upper limbs, and hemorrhagic ulcerations with purulent, thick, yellow adherent crust were scattered on the chest, upper back and shoulders, as shown in figure 1. Laboratory

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Figure 1: Acne fulminans. Hemorrhagic ulcerations with purulent crust scattered on the back and neck

tests revealed abnormal white blood cell counts (23.2 x 10^3/mm^3; segments 84.0%, band forms 1.0%, lymphocytes 13.0%, monocytes 2.0%) and hemoglobin (12.5 g/dl). The pelvic radiography did not reveal any abnormality.

After 21 days using oral prednisone 1 mg/kg daily and dapsone (diaminodiphenylsulfone) 100 mg daily, ulcerative lesions began to noticeably decrease and healed with scarring (Figure 2).

DISCUSSION

Acne *fulminans* was originally described in 1959 as acne conglobata with septicemia by Burns and Colville, who described a 16-year-old youth with acute fever and acne conglobata. [4] After that, other authors also reported patients with acute fever, ulcerative acne and polyarthralgia, as Kelly and Burns (1971), who introduced the term "acute febrile ulcerative conglobate acne with polyarthralgia".[5] In 1975, Plewig and Kligman separated this affection from acne conglobata and started to use the term acne *fulminans,* emphasizing the sudden onset and severity of this disease. [6]

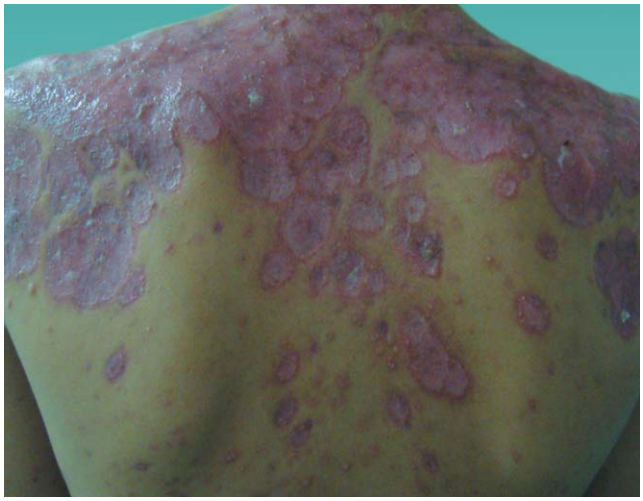

Figure 2: Patient after 21 days using oral prednisone 1mg/kg daily and dapsone 100 mg daily

The aetiology of acne *fulminans* remains unclear, but infection, genetic and immunological causes have been postulated. One of the proposed theories is that the development of acne *fulminans* is related to an explosive immunologically mediated type III and/or type IV hypersensitivity reaction to *Propionibacterium acnes* antigens.[7] Precipitation of this disease by the initiation of isotretinoin therapy has also been suspected after the description of some cases. It is postulated that istotretinoin induces fragility of the pilosebaceous duct epithelium and allows massive contact of *P. acnes* antigen with the immune system.[2,8]

High blood levels of testosterone may also play an important role on the pathogenesis of acne *fulminans,* since there are exacerbation reports during testosterone therapy. [9,10] Another theory is that acne *fulminans* could have an autoimmune aetiology.[1,2,11] Genetic factors are also considered important, as the occurrence of acne *fulminans* in monozygotic twins has already been reported.[12]

Acne *fulminans* is an extremely rare condition. According to Karvonen (1993), approximately 100 patients were described. [8] Patients typically have mild or moderate acne *vulgaris* before the onset of acute symptoms. The dermatological manifestations are highly inflammatory, painful, ulcerative lesions covered with haemorrhagical crusts, most often on the upper chest and back. The face is usually less severely involved than the trunk.[3]

In contrast with acne *vulgaris*, the systemic symptoms of sudden onset are common in acne *fulminans*.[3] These manifestations include fever, myalgia, polyarthralgia, osteolytic bone lesions, malaise, fatigue, anemia, leucocytosis, splenomegaly, and hepatomegaly.[3,13] Osteoarticular involvement is quite characteristic, and it may involve large joints such as iliosacral, ankles, shoulders and knee joints. As reported in this case, patients may demonstrate a bent-over posture because polyarthritis may make walking painful.[3]

Acne *fulminans* is easily identified because of its systemic features, although we also have to consider two important differential diagnosis: acne conglobata and rosacea *fulminans*. Acne conglobata usually is found in similar locations, occurring on most of the trunk and upper limbs, but rarely affecting the face. However, it develops in older age, has a more chronic course without an explosive onset, presents inflammatory cystic lesions and multiple comedones, and is not accompanied by any systemic symptoms.[1-3,13] On the

Table 1: Differential diagnosis of acne *fulminans*, rosacea fulminans and acne conglobata

	Acne *fulminans*	Rosacea fulminans	Acne conglobata
Gender	Men	Women	Men
Age	Adolescence (13-16 years)	Postadolescence	20 – 25 years
Onset	Sudden	Sudden	Slow
Location	Face, neck, chest and back	Face	Trunk and upper limbs. Facial lesions are rare
Clinical features	Hemorrhagic ulcerations	Comedones are rare	Nodules, inflammatory cysts, polyporous comedones
Systemic symptoms	Very common	Often none	None

Adapted source: Wakabayashi et al., 2011[1] and Jansen T, Plewig G, 1998[13]

other hand, rosacea *fulminans* is also characterized by a sudden onset, but it affects more often postadolescent women, usually after a period of stress and without a preceding history of acne. There are no systemic symptoms, the lesions are localized to central areas of the face, and comedones are rare.[1] In table 1, we summarized some characteristics of these diseases that are important to establish the differential diagnosis.[1]

The treatment of acne *fulminans* is difficult and there are many reports of different treatments. It is important to realize that acne *fulminans* does not respond to the conventional treatment for severe acne, and the use of antibiotics is ineffective. Due the severe systemic symptoms, bed rest and hospitalization may be required.[1-3,13] In addition to general supportive care, systemic corticosteroids are the mainstay of the therapy. Seukeran and Cunliffe[14] concluded that the preferable treatment for acne *fulminans* is oral prednisolone 0,5-1 mg/kg daily for 4-6 weeks with oral isotretinoin being added to the regimen in the fourth week, initially at 0.5 mg/kg daily and increasing gradually. According to them, this protocol led to faster control of systemic features as well as clearance of acne when compared with other protocols.

However, we have to consider that the use of isotretinoin is forbidden in some countries (in Japan, for example), and that some authors reported the risk of this drug precipitating acne *fulminans.* [1,8] In this context, other options such as dapsone (diaminodiphenylsulfone) and infliximab may be important. The efficacy of dapsone to treat acne *fulminans* is well established in some reports. The initial dose of dapsone is 50 mg daily, which can be increased to 100 or 150 mg daily.[1-3,8,15] ❑

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