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Abstract: Acne fulminans is a rare and severe variant of acne. In Brazilian medical journals, cases are infrequently reported, confirming its rarity. We followed five young male patients with this severe variant of cutaneous lesions, accompanied by also severe systemic symptoms: fever, anorexia, weight loss, and arthralgia. All had a good response to corticosteroids (prednisone), but had significant scarring.

Keywords: Acne fulminans; Arthralgia; Corticosteroid; Isotretinoin; Prednisone

INTRODUCTION

Although rare, acne fulminans is a diagnosis that needs to be made early due to the severity of the clinical picture and the serious resulting scarring. Cases are sporadically reported in the literature, confirming its rarity, and that is also evident when reviewing cases published in Brazil. ¹⁻⁶ The biggest number of cases in the Brazilian literature was published by Hartmann and Plewig, but the patients were from the Munich Polyclinic, in Germany. ⁷ That study was published in Brazil as a tribute to Professor Braun-Falco's 60th birthday, full professor of Munich.

The initial clinical presentation does not allow predicting the subsequent severity. Initial lesions are the common ones — comedones and small papules and pustules, and the case can be classified as acne grade I or II. However, the lesions suddenly start to multiply, become intensely inflammatory, with the formation of nodules and abscesses, and progress to necrosis and catastrophic healing. The trunk is strongly affected, especially the back, but also the shoulders and face. For the diagnosis of acne fulminans, the systemic clinical features are essential, which are florid with the severity parallel to the cutaneous involvement. There is high fever, bone and joint pains, weakness, anorexia and weight loss. Laboratory tests will show blood count abnormalities, with leukocytosis and neutrophilia, and elevated erythrocyte sedimentation rate. The acne lesions are initially sterile; culture from early lesions and blood cultures are negative.

Of interest, between severe nodular acne, acne grade IV and acne fulminans, there is an intensely inflammatory variant that needs to be treated with corticosteroids, but does not present with

systemic symptoms. This means that there is a clinical spectrum that begins with nodular acne grade IV, evolves to an inflammatory and disfiguring variant with no systemic symptoms, and ends in the most severe form, acne fulminans, with systemic symptoms.

Our cases, put together over five decades of practice, are five. They were presented by invitation, at the 18.ª Reunião dos Dermatologistas do Estado de São Paulo (Radesp), in Santos, 2013. They are very descriptive and it is of interest making them known.

CASES REPORT

First, we will briefly describe the five cases. At the end, in chart 1, we present a summary of the main clinical and laboratory data

Case 1. Fifteen-year-old male teenager, with comedonal acne for almost 6 months that worsened over the last 2 weeks (Figure 1a). He had daily fever of 38°C, besides joint, lumbar spine and knee pain. We diagnosed acne grade III and started oral (tetracycline) and topical treatment (10% benzoyl peroxide). The patient returned after 3 weeks, with severe worsening of the skin lesions (Figure 1b). The systemic symptoms had also worsened: daily fever (above 39°C), anorexia, weight loss of 6kg and generalized joint pain. We started prednisone (40mg/day), with good results.

Case 2. Fourteen-year-old male teenager with moderate acne since he was 13 years old. Four weeks before the consultation he suddenly and progressively worsened, with innumerable inflammatory lesions, mainly on the trunk (Figure 2). There was high fever (38°C) and joint pain. On examination, his temperature was

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37°C and there was edema and pain in both knees and ankles. Along with the usual acne treatment, prednisone (40mg/day) was introduced, with good results.

Case 3. The patient was 21 years old and did not have acne as a teenager. His lesions started 5 months before the consultation and rapidly progressed to the formation of generalized suppurative nodules (Figure 3). The clinical picture was followed by high fever (above 38°C) and intense hip joint pain, impairing his mobility. He had already taken antibiotics, with no success. We prescribed prednisone 60 mg per day and usual acne treatment, with clinical control. Final scarring was extremely unsightly.

Case 4. Fourteen-year-old male patient, with acne for only 30 days. Since the start, the lesions became very intense, nodular and suppurative (Figure 4). He also had daily fever above 39°C and

CHART 1: Acne fulminans — overview

Gender: male; age (years): 14, 14, 14, 18, 21

Fever (°C): 39, 38, 38, 39, 39

Arthralgia: knees (3), ankles (2), spine (1), shoulders (1), hip (1)

Leukocytosis: 12200 to 23000 with neutrophilia (from 71% to 80%)

Culture: positive for Staphylococcus sp. (2), negative (3)

joint pain in both shoulders, that impaired movement. Due to the pain, he was admitted into hospital and was under the care of an infectious disease specialist, to investigate possible septicemia. When called upon to evaluate the patient, we established the diagnosis of acne fulminans and started treatment with prednisone (40 mg/day). There was a rapid improvement.

Case 5. The patient was 18 years of age and had acne since 15. Four months before the consultation, his acne worsened suddenly and was accompanied by high fever (above 39°C). There was loss of appetite and weight (5kg). Intense pain in both knees was also present. We prescribed prednisone (40mg/day), then the dose was increased to 60mg/day, and he progressed with improvement and recurrences. The lesions caused disfiguring scarring, what led to depression and attempted suicide (Figure 5). This patient was lost for follow-up.

DISCUSSION

The severity of clinical symptoms is probably not due to an increased virulence of *Propionibacterium acnes*, otherwise there would be bacterial septicemia. There is no septicemia, even in the most severe cases. The current explanation is that *P. acnes* starts to behave as a superantigen, triggering an exaggerated and disorganized antibody response, resulting in the alarming inflammatory symptoms. The targets are pilosebaceous follicles and musculoskeletal structures.

The fact that all patients were male in early adolescence highlights the role of male hormones. Likewise, it points to the fact





Figure 1 A and B:

Case 1 — The young man presented at the first consultation and started usual acne treatment; two weeks later there was marked worsening, fever and arthralgia





FIGURE **2 A** AND **B**: Case 2 — Extremely necrotizing lesions on the back



FIGURE 3: Case 3 — Ordinary acne until he was 21 years old, when there was a marked worsening with high fever and joint pain



FIGURE 5: Case 5 — Another case with intensely necrotizing lesions





Figure 4: Case 4-14-year-old teenager. All the symptoms developed within 30 days

that somewhat older people take anabolic steroids for muscle building when enrolling in gymnasiums and subsequently develop acne fulminans. The role played by this hormone in the etiopathogenesis may not be central, but adjuvants.

It is necessary to use corticosteroids. The preference is for prednisone, in initial doses that range from 40mg/day to 60mg/day. The improvement of symptoms occurs during the first week of treatment, reverting the clinical manifestations. Corticosteroids must be associated to local and systemic routine measures. Special attention must be given to the association with antibiotics (tetracycline, minocycline, or doxycycline).

A special reference needs to be made of isotretinoin use, which is useful when the cutaneous and systemic manifestations are already under control. It is important to remember that there are reports of cases of acne fulminans in the medical literature that were triggered by this drug. \square

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