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## **Acne Fulminans: Management for the First Contact Physician**

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### **ABSTRACT**

Acne fulminans or acne malignant is a rare skin pathology that presents in the form of painful, ulcerative, disfiguring acne, usually accompanied by systemic symptoms such as fever or polyarthralgia. Arriving even in serious cases to cause bone injuries and alterations in laboratory tests. This condition is usually resistant to the antibiotics usually administered, which is why it is so important to have a multidisciplinary team that is in coordination for the care of affected patients.

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### **INTRODUCTION**

Acne fulminans or acne malignant manifests as a rare but severe form of acne vulgaris. What makes it different is its painful, ulcerative and even hemorrhagic form. They can manifest with systemic symptoms such as fever and polyarthralgia, in very advanced cases it causes bone lesions and changes in laboratory values. A very common way of confusing it is the classic presentation of acne conglobata. Both presentations are multiresistant to systemic treatments and their management is complex.<sup>1</sup>

Acne fulminans is a rare disorder and therefore its etiology is poorly understood. Although it is associated with high doses of isotretinoin, it is related to patients suffering from severe acne. An important factor is that men are more commonly affected due to high testosterone levels, however the role of testosterone in its pathogenesis has not been well established. Some of the theories regarding the pathogenesis is the use of anabolic steroids which are known to make infection by propionibacterium acnes more conducive. There have been studies that talk about the relationship that the immune system has in the development of severe cases of this entity.<sup>1,2</sup>

Acne fulminans contains a genetic component, there are different syndromic etiologies that may be related to the pathogenesis of this entity, such as:<sup>2</sup>

- Synovitis syndrome, accompanied by acne, pustulosis, hyperostosis and osteitis (SAPHO).

-Pyogenic arthritis syndrome, pyoderma gangrenosum with acne (PAPA)

-Pyoderma gangrenosum, acne and hidradenitis suppurativa (PASH) syndrome

-Pyogenic arthritis, pyoderma gangrenosum, acne and hidradenitis suppurativa syndrome (PAPASH).

There are few reported cases, young men are usually the group most affected by the factors mentioned above, as well as a history of having suffered from acne, although fortunately its presence has been decreasing over time.<sup>3</sup>

The clinical presentation is usually classic, of sudden onset and with ulcerative acne, they come to complain of systemic symptoms (fever, arthralgia) resistant to antibiotics due to the past history of antibiotic use for the treatment of acne vulgaris. Therefore, bacterial resistance is much higher.<sup>3,4</sup>

Acne fulminans is a disease with a clinical presentation very similar to that of acne conglobata, however, acne fulminans presents with numerous nodules on the trunk, painful, ulcerated, hemorrhagic and with the presence of scabs. Systemic symptoms usually occur because while the conglobata presents with painful splenomegaly and erythema, the fulminant fever, bone pain, and joint pain are more common (the pelvis, hip, and knees being more frequent).<sup>4,5</sup>

Because the disorder may be associated with systemic symptoms suggestive of infection, a blood count may reveal such data as increased white blood cell counts, elevated neutrophils, anemia of chronic inflammatory disease, increased acute-phase reactants such as the rate of erythrocyte sedimentation rate and C-reactive protein and even altered liver function tests in very severe cases.<sup>4,5</sup>

Due to bone and joint pain, imaging studies are often performed to rule out other pathologies. In initial cases, only reactive changes can be seen, but in advanced cases, lytic

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lesions will be present in almost half of the cases. they can be assimilated to osteomyelitis lesions but the cultures of these tend to be negative.<sup>5,6</sup>

### TREATMENT / MANAGEMENT

The indicated treatment for acne fulminans is a combination of corticosteroids and isotretinoin, the doses will be based on the weight of the affected patient, for example, corticosteroids will be started at high doses (0.5 to 1mg/kg/day) for at least 2 weeks and even 4 weeks if the patient presents systemic symptoms, treatment with corticosteroids will be suspended until complete healing of the wounds. Once this step is reached, isotretinoin will be added at a dose of 0.1mg/kg/day for at least 4 weeks, the drug will be de-escalated until a minimum maintenance dose of 120mg/kg is reached.<sup>6,7</sup>

In patients who remain adequately adhered to treatment, relapses rarely occur, but in the event that it happens, the accumulated dose of isotretinoin will be repeated for months, since the initial dose is low.<sup>6,7</sup>

The use of high-potency topical corticosteroids has shown greater efficacy when curing skin lesions caused by exaggerated inflammatory reaction.<sup>6,7</sup>

Acne fulminans does not usually respond to the first-line antibiotics traditionally used to treat acne, even if they are used the response is slow and incomplete.<sup>7</sup>

### OTHER ALTERNATIVE TREATMENTS

The use of pulsed dye laser therapy has had moderate results when treating the lesions, but since it generates multiple adverse effects and pain, adjuvant therapy is required for the complete remission of the lesions.<sup>7,8</sup>

Some rare but effective combinations are prednisone with dapsona or with the combination of cyclosporine.<sup>8</sup>

The treatment must be done jointly by the dermatologist and the internist, care must be taken with the use of retinoids since these agents are teratogenic in many cases (special care in women of childbearing age). If a woman takes isotretinoin she should avoid getting pregnant for at least a month after stopping treatment.<sup>9</sup>

### CONCLUSIONS

This pathology can cause a serious impact on the face of patients. The primary care physician should know to refer to a dermatologist as soon as possible. The self-esteem of these patients is severely affected by poor aesthetics, their quality of life is affected and they tend to avoid going out, becoming withdrawn and isolated. That is why psychosocial counseling should be offered at the first care appointments. Suicidal ideation is very common in these patients, therefore, joint management with psychiatry is key to the success of the treatment of these patients.

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