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Case Report

Diagnostic challenges and treatment difficulties in a patient with excoriated acne conglobata

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Abstract

Acne conglobata is a rare and severe form of acne vulgaris, characterized by the presence of comedones, papules, pustules, nodules and sometimes hematic or meliceric crusts. Acne excoriée is a form of self-inflicted skin condition in which the patient picks on imaginary or real acne lesions.

We report the case of a 16 year old Caucasian female patient from the urban area who addressed our dermatology department for erythematous, edematous plaques covered by pustules and crusts, located on the face. The anamnesis revealed that during the last weeks she had had a depressive mood after ending a relationship with her boyfriend and started scratching and picking on the lesions.

The patient's depressive mood prior to the worsening of the disease was probably aggravated by the condition. This might have determined the picking of the skin which could have impeded the response to standard treatment. The self-excoriative behavior could also be regarded as an appeal for help.

Keywords: acne conglobata, acne excoriée, self-inflicted, azithromycin

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Introduction

Acne conglobata is a rare, severe form of acne vulgaris characterized by the presence of comedones, papules, pustules, nodules and sometimes hematic or meliceric crusts, located on the face, trunk, neck, arms and buttocks. Males are affected more frequently than females. Acne excoriée is a form of self-inflicted skin condition in which the patient picks on imaginary or real acne lesions. It is more frequent in girls and young women. Patients with this disorder generally openly admit the self-inflicted nature of the lesions but are unable to stop picking their skin. Therefore, acne excoriée is considered a form of psychocutaneous disease (1-3).

Case Report

We report the case of a 16 year old Caucasian female patient from the urban area who addressed our dermatology department for erythematous, edematous plaques covered by pustules and crusts, located on the face. The lesions were painful. The patient asserted that she had a mild form of acne for three years before presentation. In the last three weeks the disease worsened and the patient applied topical isotretinoin with no clinical improvement. The anamnesis revealed that during the last weeks she had a depressive mood after ending a relationship with her boyfriend and started scratching and picking on the lesions. The physical examination revealed an afebrile patient and was within normal range. The personal and family histories were unremarkable.

Clinical examination noticed erythematous, edematous plaques located on the forehead, cheeks and chin (Figure 1). The lesions were covered by pustules, erosions and meliceric crusts. Multiple comedones were observed, especially on the nose and few nodules were present on the cheeks. The other areas of the body were not affected.

Laboratory findings revealed an increased erythrocyte sedimentation rate. Bacterial cultures were performed from some of the pustules and were inconclusive. Endocrinology consult showed no abnormalities.

The differential diagnosis included acne conglobata, acne excoriée, acne fulminans, pyoderma faciale and impetigo. Based on the clinical and laboratory findings the patient was diagnosed with acne conglobata. She started treatment with systemic isotretinoin and corticosteroids with no significant improvement. Meanwhile, she continued picking and scratching the lesions despite the repeated recommendations to stop this behavior, the patient accusing an uncontrollable urge to manipulate the skin.

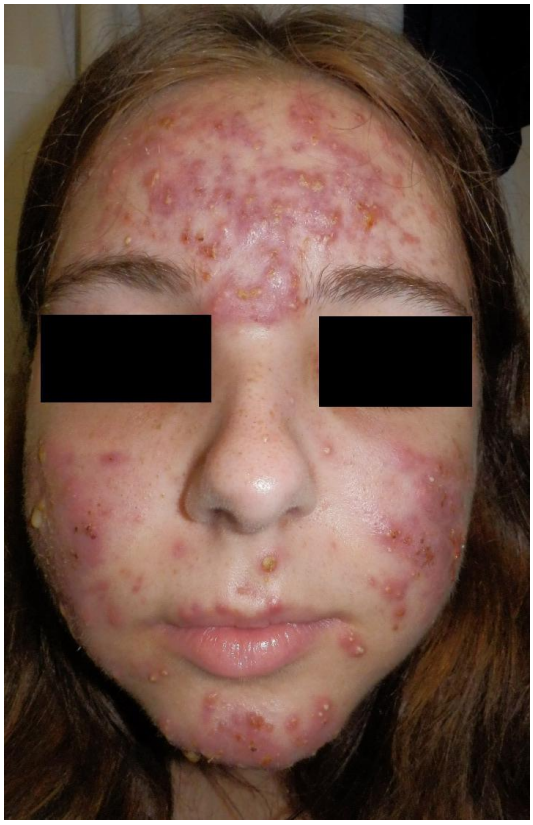

Figure 1. Clinical presentation before treatment

A psychiatric consultation was recommended but the criteria for a specific psychiatric disorder were not met. Given the poor response to treatment, the patient was

referred to the infectious disease department. Together with the infectious disease specialist we hypothesized that the repeated injury of the acne lesions might have led to superinfection which could explain the lack of response to conventional treatment. The retinoid and corticosteroid therapies were stopped and the patient started treatment with azithromycin 500 mg orally daily and topical fusidic acid.

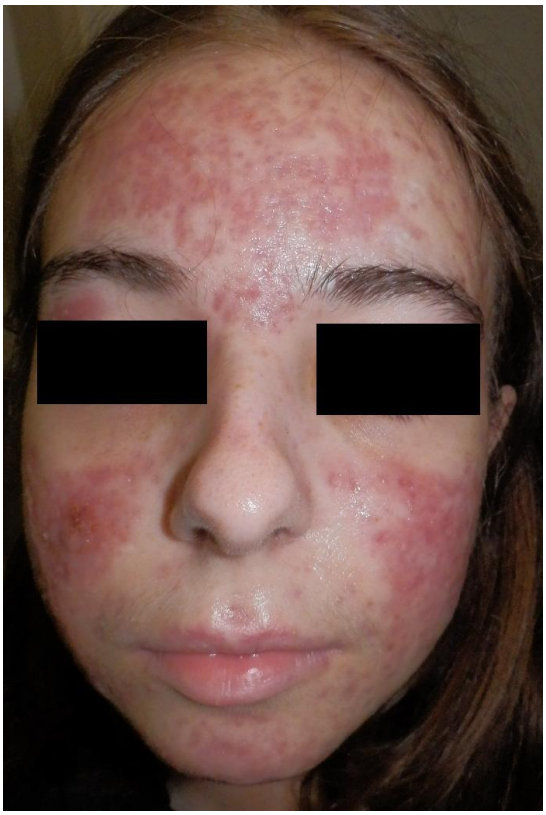

Figure 2. Clinical presentation after treatment

After two weeks of treatment the clinical improvement was obvious. Complete resolution of the inflammatory lesions was obtained after 3 months of treatment with azithromycin. The disease healed leaving significant atrophic scars and postinflammatory erythema (Figure 2). After 3 months of azithromycin, the patient was switched to hormonal treatment with antiandrogens. She was recommended psychiatric counseling but she refused. The patient remains under our supervision.

Discussion

Acne is a multifactorial disorder of the pilosebaceous unit characterized by the occurrence of comedones, papules, pustules and sometimes nodules. It is a ubiquitous disorder most often affecting males and females during puberty. It can however occur at any age. It is one of the dermatological disorders most frequently seen in practice. Acne conglobata is a rare and severe form of acne which affects males more frequently than females. Apart from the typical acne lesions, patients also present nodules, abscesses, draining sinuses, cysts and fistulated comedones which are located on the face, chest, back and buttocks. The lesions are often painful and the systemic signs and symptoms are usually absent. The onset of the disease is generally sudden, the affliction occurring in patients with no acne or mild forms of acne (3-5).

Acne excoriée (sin. acne des jeunesfilles) is a psychocutaneous disorder characterized by excessive picking or scratching of imaginary or real acne lesions. Psychocutaneous disorders are classified as primary psychiatric disorders, in which patient's behavior leads to perceived or real dermatologic conditions and primary dermatological disorders in which the dermatological disease determines depression, anxiety and an impaired quality of life. Acne excoriée is a subset of neurotic excoriations, a primary psychiatric condition in which patients produce skin lesions through repetitive excoriation of the skin and admit their role in the disease (1, 5, 6).

Acne excoriée was first described in 1898 by Brocq (7). Girls and young women are primarily affected but it can occasionally occur in males. Patients with acne excoriée describe an uncontrollable urge to manipulate their lesions. Therefore, comorbid psychiatric disorders such as obsessive-compulsive disorder, body dysmorphic disorder, depression or anxiety must be suspected in those patients. Some authors consider acne

excoriée a protective device to conceal personal failure or an appeal for help coming from persons with immature personality who lack coping mechanisms (8-11).

Clinically, acne excoriée presents as erosions covered by crusts as well as excoriations, ulcerations and scars which are often disfiguring (2, 8). Studies showed that the severity of acne does not correlate with the intensity of self-excoriation in young females. Therefore, most females with acne excoriée have mild forms of acne. Personality traits however seem to be important in those patients, perfectionists or compulsive persons having a higher risk of developing this disorder (12).

A study performed by Gupta et al. in males with acne showed that the self-excoriative behavior is correlated with depression and anxiety but not with body dysmorphic disorder and obsessive-compulsive disorder. The group of authors also showed that in males, the severity of acne is correlated with the self-excoriative behavior but the severity of the behavior is usually not high enough to determine acne excoriée (11).

In the case we are reporting the patient presented comedones, erythematous, edematous plaques covered by pustules and crusts as well as a few nodules. The lesions were confined to the face and were painful. The patient presented mild acne which suddenly aggravated shortly after the patient broke up with her boyfriend. The patient admitted that she scratched and picked on the lesions and asserted that she could not resist the urge to do so.

The clinical aspect of the lesions suggests the diagnosis of acne conglobata. The occurrence of the disease in a female patient, the presence of very severe lesions on the face in the absence of any lesions elsewhere on the body as well as the poor response to conventional therapies compelled us to exclude other possible disorders. The differential diagnosis included acne fulminans, rosacea fulminans, acne excoriée and impetigo.

Acne fulminans, also known as acne maligna, is a rare and severe form of acne characterized by pustules, severe, painful, hemorrhagic nodules covered by crusts affecting the face, trunk and, rarely, the thighs. The disease has a sudden onset and generally affects young males. It is associated with fever, chills, loss of appetite, weight loss, anorexia, polyarthralgia, musculoskeletal pain and hepatosplenomegaly. The lesions heal with severe scarring (13-15). In the case we are reporting the patient presented severe lesions on her face with a sudden onset. She did not however present any systemic symptoms. The diagnosis of acne fulminans was therefore excluded.

Rosacea fulminans, also known as pyoderma faciale, is a rare and very severe form of rosacea affecting young women. It is characterized by the sudden onset of erythematous plaques with pustules and nodules predominantly located on the face. Unlike acne, the patients do not present comedones. Systemic symptoms are absent. It heals with scarring (3, 16). The age and gender of the patient, the onset and location of the lesions could suggest the diagnosis of rosacea fulminans. The presence of comedones and the poor response to oral prednisone however helped us exclude the diagnosis of rosacea fulminans.

Impetigo is a superficial skin infection caused by Staphylococcus aureus or Streptococcus pyogenes. Bullous impetigo occurs in newborns and infants. Non-bullous impetigo can affects both children and adults. It clinically presents as vesicles or pustules which evolve to plaques covered by meliceric crusts most often located on the face (1, 17). In the case we are presenting the patient's lesions were not characteristic for impetigo. We do however suspect impetiginization of the acne lesions secondary to continuous scratching but also to the prednisone treatment. The lack of response to the

retinoid and corticotherapy and prompt response to antibiotic also support this theory.

Acne excoriée is a self-inflicted disorder most often affecting girls and young women. It is characterized by erosions and ulcerations covered by crusts and scars from healed lesions. Patients presenting this disorder scratch imaginary or real acne lesions. The disease is often associated with psychiatric disorders.

Our patient admitted that she was scratching the lesions and asserted that she could not control the urge to do so. The lesions however were too severe to be produced by skin picking alone. The patient had a depressive mood because she had recently broken up with her boyfriend. The psychiatric examination did not reveal a psychiatric disorder. The self-excoriative behavior might have been an appeal for help or attention of a girl who was under a lot of stress.

According to the European evidence based guideline for the treatment of acne, oral isotretinoin is strongly recommended as monotherpy for the treatment of severe nodular and conglobate acne. Other treatment options, with moderate strength of recommendation, include systemic antibiotics, namely doxycycline and lymecycline, in combination with the fixed dose combination of adapalene and benzoyl peroxide or with azelaic acid. There is also little evidence that hormonal antiandrogens in combination with topical treatment could be beneficial in patients with severe nodular or conglobate acne (18).

We initially treated our patient with oral isotretinoin and oral corticosteroids and observed no clinical improvement. We therefore hypothesized that the patient's self-excoriative behavior might determine a superinfection of the lesions and, together with the specialist in infectious diseases decided to start azithromycin treatment as the patient's bacterial examinations were irrelevant. The clinical improvement was observed after two weeks of treatment and complete resolution was obtained after three months. At that point we also noticed that our patient's mood had improved and she claimed that she only rarely picked on the lesions.

Azithromycin is a macrolide antimicrobial with a wide spectrum of activity. Even though it is not the standard treatment for acne, several studies showed that it could be a good alternative to regularly used antibiotics. Rafiei and Yaghoobi performed a study on 290 patients with moderate to severe acne in which they aimed to determine the efficacy of azithromycin versus tetracycline and concluded that azithromycin is a safe and effective alternative in the treatment of acne (19). Babaeinejad et al compared the efficacy of azithromycin and doxycycline in a randomized, double-blind clinical trial which included 100 patients with moderate acne. The authors concluded that azithromycin is as effective as doxycycline in patients younger than 18 years but doxycycline is more effective in patients older than 18 years (20).

Conclusions

We report a case of acne conglobata in which the patient's self-excoriative behavior altered the usual course of disease. This case had several particularities. The patient had very severe lesions on the face but no lesions in other areas usually affected by acne. The patient's depressive mood prior to the worsening of the disease was probably aggravated by the condition. This might have determined the picking of the skin which could have impeded the response to standard treatment. The self-excoriative behavior could also be regarded as an appeal for help. The lack of response to isotretinoin, the first line treatment for acne conglobata and rapid response to azithromycin is another particularity. Anti-anxiety or anti-depressant drugs should probably be considered in the future, for similar patients.

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