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Rosacea Fulminans: two case reports and review of the literature

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Rosacea Fulminans is a rare and severe inflammatory dermatosis which affects predominantly childbearing women. It is characterized by sudden onset and it usually localizes exclusively on the centrofacial areas, presenting with numerous fluctuant inflammatory nodules and papules which may coalesce. Treatment with isotretinoin in combination with topical and systemic corticosteroids is successful. Clearance of lesions may be obtained under systemic treatment with no or minimal scarring outcomes.

Due to rare incidence its pathophysiological mechanisms, diagnosis and management remain controversial. We report two cases of Rosacea Fulminans arised in otherwise healthy people and completely healed after treatment. Our aim is to share our experience about this disease in order to increase knowledge about its diagnosis, management and its treatment. We also make a review of the literature of this peculiar dermatosis.

Keywords: rosacea fulminans; isotretinoin; centrofacial area; childbearing; dermatology.

Introduction

Rosacea Fulminans (RF) is a rare acute inflammatory dermatosis characterized by abrupt onset of severe inflammatory lesions on erythematous and edematous skin confined to the centrofacial area. It was originally classified as an acne conglobata variant and then categorized as a Pyoderma Faciale by O'Leary and Kierland in 1940 supposing an infectious cause of the pilosebaceous units (1), even if a clear infective agent was never demonstrated.

Afterwards, Plewig proposed renaming of this condition and introduced for the first time the term - currently used- of Rosacea Fulminans in 1992, emphasizing the non-bacterial origin of the disease (2) and susteining it was not a variant of acne conglobata but an unique entity.

Only few reports about RF can be found in all the literature since its first description. We have found a total of 144 cases described and female sex was the gender most commonly affected (92% Vs 9%).

Due to infrequent reporting in the literature, this condition remain controversial regarding pathophysiology and especially treatment.

We report two cases of Rosacea Fulminans arised in otherwise healthy people and completely healed after treatment.

The aim of our report is to share our experience about this disease in order to increase knowledge about its diagnosis, management and its treatment.

We also make a review of the literature.

Case Reports

A 17-year-old otherwise healthy caucasian man was admitted to our department due to sudden onset of inflammatory cutaneous eruption on the face since about two months.

Patient had previously been affected by acne vulgaris. Dermatological examination revealed large coalescing pustules, nodules and cystic structures on a background of widespread erythema localized on the cheeks, chin and forehead. Large sinuses with spontaneous discarge of purulent material were also present.

A 33-year-old Asian childbearing woman referred to us due to arising of facial eruption since about 5 weeks before characterized by very large and coalescing nodules on the forehead, cheeks and chin with large and interconnecting sinuses draining purulent and ematic mixed material (*figure 1*). She reported a history of eythematotelangiectatic rosacea which was into clinical remission since long time.

No comedones or extrafacial involvement were noticed in both cases. No systemic symptoms or fever were referred by our patients.

Swab microbiological cultures were negative as well as routine biochemistry finding were within normal ranges including blood count, erythrocyte sedimentation rate and C-reactive protein.

Diagnosis of Rosacea Fulminans was made on the basis of the history and clinical examination in both patients.

We prescribed oral treatment with low-dosage of metilprednisolone (16 md/daily) associated to antibiotic therapy such as azithromycin pulsed therapy (500mg daily for three consecutive days a week).

After about ten days, low-dosage oral isotretinoin was added and it was maintained at very low-dosage (0.3 mg/kg/day) for about 3 months. Systemic corticosteroid was gradually tapered and stopped within one month while azithromycin treatment was prolonged for 8 weeks in pulsed therapy.

No topical medication was associated except for mosturizing cream.

Oral contraceptive was administered to the childbearing woman due to potencial teratogenic effects of isotretinoin and it was stopped after one month by the end of treatment.

A rapid improvement of skin lesions was obtained in both patients. Reduction and desappearance of papulo-pustular component were showed at the end of treatment.

Post-inflammatory hyperpigmentation healed in few months without scarring outcomes (*figure 2*).

Materials and Methods

A computerized literature search in PubMed was performed from all years available through January 2019 using the following search terminology: “rosacea fulminans” OR “pyoderma faciale” OR “rosacea conglobata”. Only articles in english were considered. Articles in other languages were excluded.

Discussion

Rosacea Fulminans is a rare and severe inflammatory dermatosis arising in centrofacial regions. It usually affects females between theirs 20s - 40s. Only few cases have been described in children or in man, as our case do it.

To date only 144 cases of RF have been reported in all the literature: 92% of patients were female while men amounted to 8%. Average age of onset was of 31.3 years.

Patients are usually healthy people with medical history of facial dermatosis: flushing or erythematotelangiectatic rosacea have been reported in about 80% of cases in contrast to vulgaris acne in less of 50% of cases.

Clinically, sudden erythematous papules, pustules and nodules tipically arise on centrofacial regions with a tendency to coalesce into plaques and interconnecting sinuses draining purulent material. Forehead, nose, chin and cheeks are frequently the most centrofacial regions affected as shown in about 87% of reported cases in literature. No sign of acne is present in Rosacea Fulminas and lack of comedones represents an important diagnostic criteria.

Extrafacial involvement is not common: neck, back and decollete have been involved in 13% of cases. Ocular involvement is very rare (less of 4%).

Fever or other systemic symptoms are tipically absent in most of the cases.

Remission with no or minimal scarring is usually obtained under specific treatment. Recurrence is very rare (20% of the cases).

Despite broad knowledge about clinical signs of Rosacea Fulminans, its etiopathogenesis is nowadays unclear and its remains an open issue.

Its probably represent an acute inflammatory immuno-mediated response to specific unknown triggers. Emotional or stressful events or hormonal alterations, such as menopause, pregrancy or oral contraceptive intake, often precede the onset of RF and they could act as trigger inducing a possible neuro-vascular dysregulation that would be at the basis of pathogenic mechanisms of the disease as well as happen in other form of rosacea (3-4).

Although not certain, these theories based on hormonal triggers could be justify why disease is more common in young women during pregnancy or postpartum period (7-15), while it rarely arises in men (16-26). In all the literature, it has been reported until 11/144 (10%) cases of RF arised during pregnancy.

Cases of Rosacea Fulminans have also been reported following specific drugs intake such as azathioprine, high dose vitamins B6 or B12 and interferon which could have a role as triggering factor (27-30).

Make a diagnosis of RF is not so complicated; it is exclusively based on clinical signs and medical history.

In 1994 Plewig et all.(5-6) examined a clinical series of patients affected by RF and they set out clinical features of Rosacea Fulminans in order to helping make the diagnosis.

Nowadays, these clinical criteria are still used by most of clinicians. Both our patients perfectly fell within these criteria.

Histological examination is not diriment and rarely performed. It shows a superficial and deep dermal inflammatory infiltrate of neutrophils, lymphocytes and eosinophils with a perivascular and periadnexal distribution. Superficial accumulations of neutrophils are present in pustules. In later stage, giant cells and epithelioid granuloma are seen. Occasionally, cutaneous biopsy could be indicated to rule out granulomatous diseases.

All laboratory parameters are usually within normal ranges. C- reactive protein or erythrocyte sedimentation rate can be rarely increased but they remains aspecific. Swabs microbiological cultures are negative unless otherwise bacterial overinfection.

Management of RF is not yet standardized.

In 2017, the National Rosacea Society (NRS) Expert Committee proposed an update standard classification of Rosacea in order to provide clearer instruments for diagnosis and treatment of rosacea but Rosacea Fulminans was excluded (31). So, do not exists any guideline regarding treatment of this rare condition and few reported cases from literature probably not allows a standard treatment shared by all scientific comunity.

Systemic treatment is needed to achieve clinical remission with no or minimal scarring. Risk of scarring is more high if untreated.

Unlike other variants of rosacea, RF is the only one in which use of topical and systemic steroid is indicated.

We believe that a better therapeutical approach for RF is that Plewig and co-authors (5) proposed as first.

Their therapeutic regimen was based on administration of a short course of topical and systemic corticosteroids for 10-14 days (prednisone – dosage 0.5-1.0 mg/kg/daily) and after that oral isotretinoin was introducted at a dosage of about 0.2-0.5 mg/kg/daily.

Oral corticosteroid needed to gradually tapering in about 2 or 3 weeks while oral isotretinoin can be maintained for 2-4 months. For chilbearing women oral contraceptives (especially antiandrogen type) were also prescrived.

In our opinion, antibiothic therapy should be always associated, especially in severe cases.

When oral corticosteroids cannot be used, isotretinoin can be administered alone in association to potent topical corticosteroids achieving good clinical response (32).

Dapsone, administered alone (23,33-34) or in combination with tetracyclines, has been documented in few refractory cases at dosage of 50-100 mg daily. It was used for neuthophilic infiltrate showed in skin biopsies (35).

When RF occurs in pregnancy therapeutical approaches became a challenge since when all validated effective treatments are controindicated during pregnancy.

Tetracyclines can induce impaired bone growth and Metronidazolo is not recommended before the second trimester. Dapsone may cause neonatal haemolysis. Topical antibiotic or steroid therapy can be administered but they lead to poor improvements as reported in literature (10-11).

In these special condition, we suggest to introduce oral antibiotic therapy in association with high potency topical corticosteroids.

Macrolide antibiotic with their immunomodulatory effects are the choise in these kind of patients and they can be used throughout the pregnancy or nursing. Clarithromycin and Azithomycin are better than Erytromycin which it is often associated with resistance in some strains of Propionobacteria (12).

Use of systemic steroids always has to be carefully valutated during pregnancy and they should be reserved only for selected cases. Their administration is justified only if the benefits outweight the risks because of risk of intrauterine growth retardation, maternal diabetes mellitus and hypertension.

Conclusion

Many question remains uncertain about Rosacea Fulminans, especially its pathogenetic mechanisms and its possible trigger factors.

Nowadays, so few cases are reported in literature, especially in male sex. It could be related with a misdiagnose with acne fulminans that it is usually more common in male. So it is important to differentiated by these two conditions keeping in mind that Acne Fulminans is characterized by fever and maleise, polyarthralgias, comedonesd and extrafacial lesions while Rf doesn't.

So, more attention is required to better recognize this dermatosis and in our opinion clinical criteria by Plewig et all (2) represent a valid instrument for this aim.

Further studies and more cases are required to improve management and therapeutic approaches for this rare dermatosis.

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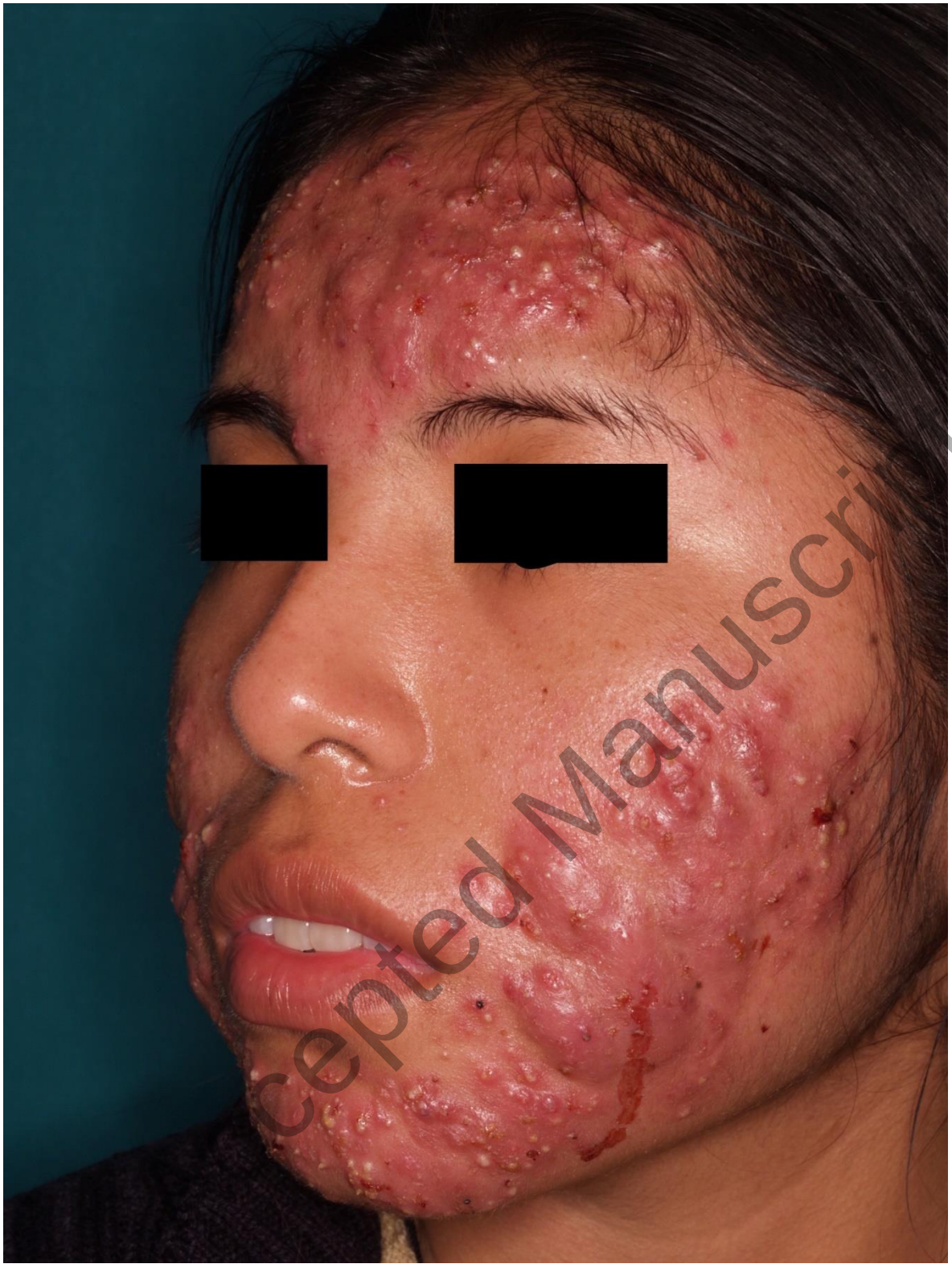
Figure 1. Multiple large and coalescing nodules with large and interconnecting sinuses draining purulent and ematic mixed material on childbearing woman‘s centrofacial area.



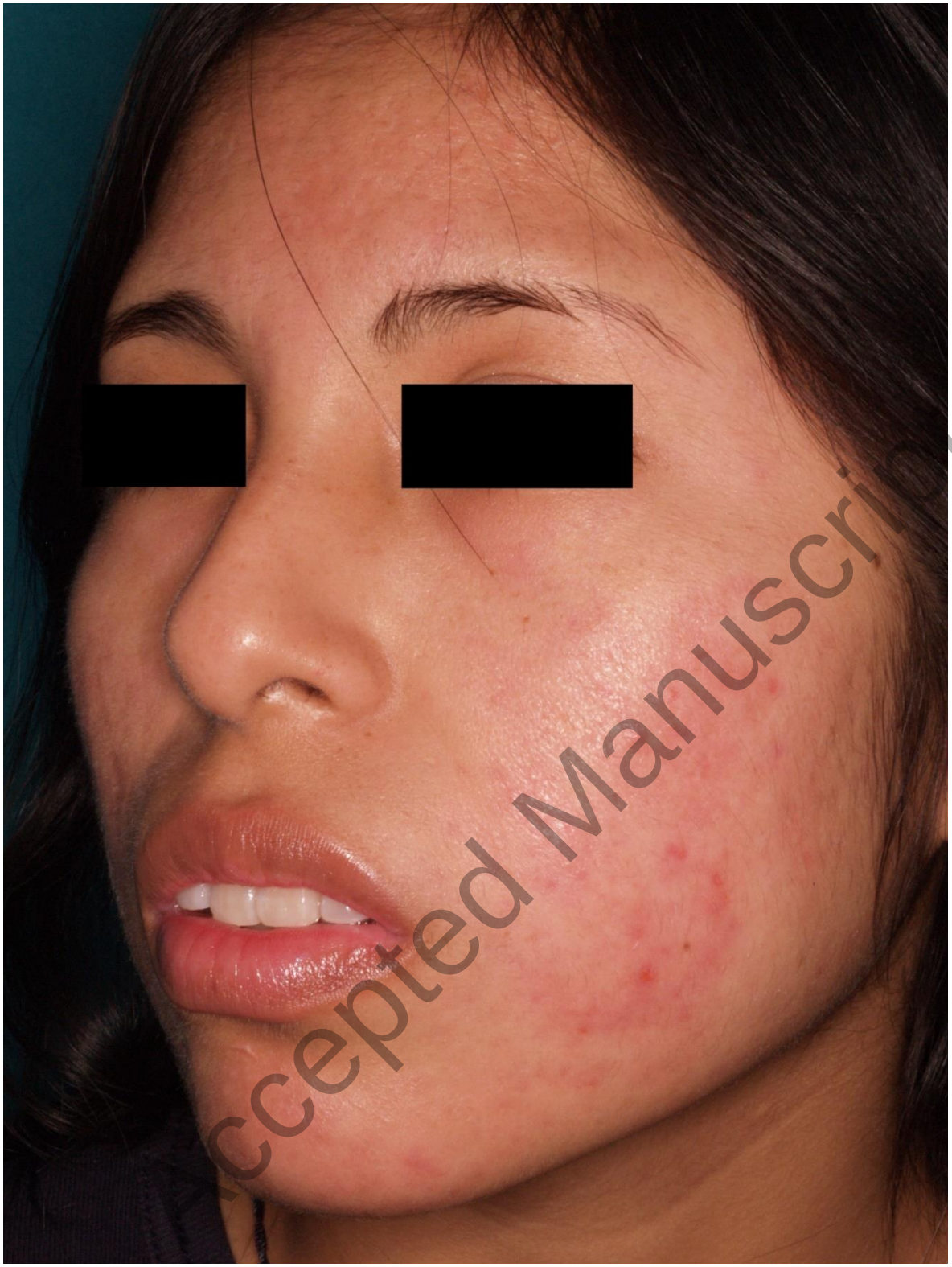


Figure 2. Six-month follow-up: complete remission with no scarring outcomes.