ID Design Press, Skopje, Republic of Macedonia
Open Access Macedonian Journal of Medical Sciences. 2018 Aug 20; 6(8):1438-1441.
https://doi.org/10.3889/oamjms.2018.267
eISSN: 1857-9655
Case Report

A Rare Dermatologic Disease in Pregnancy: Rosacea Fulminans- Case Report and Review of the Literature

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Citation: Demir O, Tas IS, Gunay B, Gungor Ugurlucan F. A Rare Dermatologic Disease in Pregnancy: Rosacea Fulminans- Case Report and Review of the Literature. Open Access Maced J Med Sci. 2018 Aug 20; 6(8):1438-1441. https://doi.org/10.3889/oamjms.2018.267

Keywords: Rosacea; Pregnancy; Rare disease

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Received: 07-Apr-2018; **Revised:** 26-May-2018; **Accepted:** 30-May-2018; **Online first:** 04-Aug-2018

Funding: This research did not receive any financial support

Competing Interests: The authors have declared that no competing interests exist

Abstract

BACKGROUND: Rosacea is a common, chronic disorder that can present with a variety of cutaneous or ocular manifestations. Skin involvement primarily affects the central face, with findings such as persistent centrofacial redness, papules, pustules, flushing, telangiectasia, and phymatous skin changes. The pathways that lead to the development of rosacea are not well understood. The relationship of pyoderma faciale (also known as rosacea fulminans) to rosacea also is uncertain. We aimed to write this article with the aim of showing how a pregnant patient who has been aggravated by the degree of lesions on the face during the first trimester of pregnancy is treated and to show what is in the literature in this issue.

CASE REPORT: A 22-year-old woman complained of painful erythema, papules and pustules on the face. She had fever and malaise during the sixth week of her first pregnancy and a history of the mild eruption and seborrhea before her pregnancy with flaring over the preceding 4 weeks. Dermatologic examination revealed red erythema of all involved facial areas; the lesions consisted of papules, pustules and nodules. The case was diagnosed as rosacea fulminans (pyoderma faciale) by these findings. In the literature, there are some effective therapeutic options such as retinoids, tetracyclines, antiandrogenic contraceptives, and dapsone and these were not used because they are contraindicated in pregnancy. Amoxicillin-clavulanic acid 1 gr/day, wet compresses, and a fusidic acid cream were started. After the activity of the disease had been suppressed for 10 days, antibiotic was stopped, and the other treatment options were applied topically for the next month. One month after cessation of treatment, the lesions had disappeared with only mild erythema remaining. There was minimally flushing on the face and no telangiectasia.

CONCLUSION: In conclusion, there is no substantial evidence as to the mechanism by which pregnancy may trigger this conditioner whether the gender of the fetus influences the development of rosacea fulminans, but is generally accepted that hormonal changes in pregnancy play an important role. The pathogenesis of rosacea fulminans remains uncertain, but it is obvious that the further basic and clinical research is required to optimise the management of this rare facial dermatosis.

Introduction

Rosacea is a common, chronic disorder that can present with a variety of cutaneous or ocular manifestations.

Skin involvement primarily affects the central face, with findings such as persistent centrofacial redness, papules, pustules, flushing, telangiectasia, and phymatous skin changes. The pathways that lead to the development of rosacea are not well understood [1] [2].

Proposed contributing factors include abnormalities in innate immunity, inflammatory reactions to cutaneous microorganisms, ultraviolet damage, and vascular dysfunction.

The relationship of pyoderma faciale (also known as rosacea fulminans) to rosacea also is uncertain [3]. Patients present with intensely inflammatory, purulent facial plaques and nodules with draining sinuses on a background of erythema. A history of rosacea may or may not be present. Young women are most commonly affected.

Because pregnancy is also a process in which

the immune system is weakened, cases of rosacea exacerbated in pregnancy, namely rosacea fulminans, have been reported in the literature.

We aimed to write this article to show how a pregnant patient who has been aggravated by the degree of lesions on the face during the first trimester of pregnancy is treated and to show what is in the literature in this issue.

Case Report

A 22-year-old woman complained of painful erythema, papules and pustules on the face. She had fever and malaise during the sixth week of her first pregnancy and a history of the mild eruption and seborrhea before her pregnancy with flaring over the preceding 4 weeks. Dermatologic examination revealed red erythema of all involved facial areas; the lesions consisted of papules, pustules and nodules (Figure 1A, 1B).

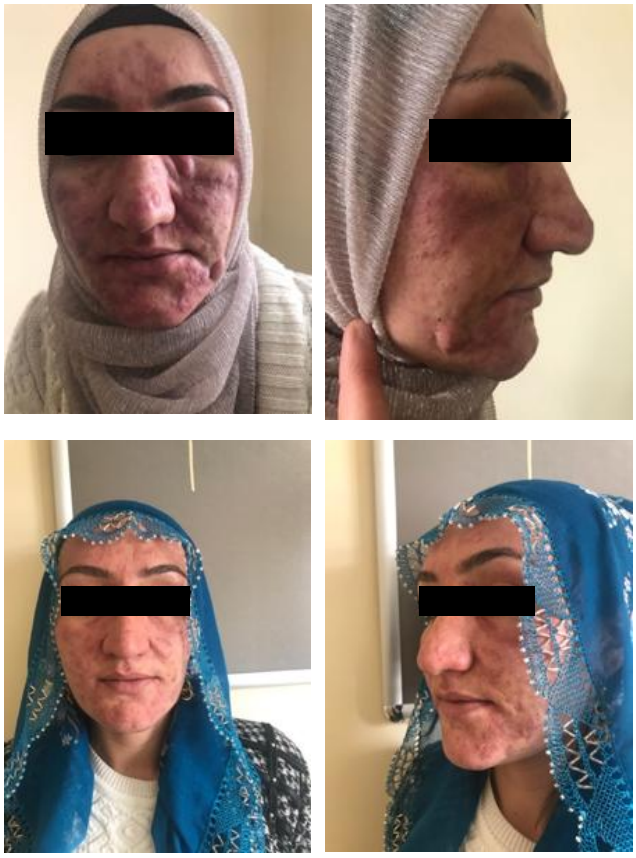

Figure 1: Patient with rosacea fulminans (pyoderma faciale). A, B = before the treatment; C, D = after the treatment

There were no comedons or telangiectasia and no pathologic findings upon systemic examination. Laboratory studies revealed a white blood cell count of 14600/mm, haemoglobin level of 13.8 g/dl, CRP level of 6 mg/dl. Cultures for bacterial pathogens were negative. Hormonal tests were not performed. The case was diagnosed as rosacea fulminans (pyoderma faciale) by these findings. In the literature, there are some effective therapeutic options such as retinoids, tetracyclines, antiandrogenic contraceptives, and dapsone and these were not used because they are contraindicated in pregnancy. Amoxicillin-clavulanic acid 1 gr/day, wet compresses, and a fusidic acid cream were started. After the activity of the disease had been suppressed for 10 days, antibiotic was stopped, and the other treatment options were applied topically for the next month.

One month after cessation of treatment, the lesions had disappeared with only mild erythema remaining. There was minimally flushing on the face and no telangiectasia (Figure 1C, 1D).

Acknowledgement

To use the pictures for scientific purposes, permission was obtained from the patient.

Discussion

In 2002, the National Rosacea Society assembled an expert committee to develop a standard classification system for rosacea.

The committee established four distinct subtypes of rosacea: erythematotelangiectatic, papulopustular, phymatous, and ocular rosacea [3]. Since then, increasing knowledge of the pathophysiology of rosacea has favoured a view of rosacea as a consistent multivariate disease process with multiple clinical manifestations rather than distinct subtypes of disease [4].

Following recommendations from the Global ROSacea COnsensus (ROSCO) Panel supporting the use of phenotype-based, rather than a subtype-based, approach to the diagnosis and classification of rosacea, the National Rosacea Society expert committee released an update supporting a similar approach [4] [5].

The cutaneous histopathologic findings in rosacea are nonspecific, and skin biopsies are rarely indicated. When the diagnosis is uncertain, biopsies may be performed to rule out other disorders or to provide support for a diagnosis of granulomatous rosacea. No serologic studies are useful for diagnosis.

Specimens from erythematous facial skin exhibiting centrofacial erythema and telangiectasias usually show dilation of superficial blood vessels and

a low-grade perivascular, lymphohistiocytic, inflammatory infiltrate with occasional plasma cells. Solar elastosis is often present. Histopathologic examination of papular lesions usually reveals prominent perivascular and perifollicular inflammatory infiltrates in the superficial and mid-dermis composed of lymphocytes, neutrophils, and plasma cells. Superficial accumulations of neutrophils are present in pustules. In contrast to acne vulgaris, inflammation often is more perivascular and extends well beyond the follicle [6].

Rosacea fulminans was first described by O'Leary and Kierland in 1940 as ‘pyoderma faciale [7]. Plewig et al., Regarded the condition as an extreme form of rosacea and suggested that it be renamed ‘rosacea fulminans’ in their report of twenty cases had been reported in the literature [8].

Rosacea fulminans is characterised by rapid onset, a fulminating course, strict localisation to the face, absence of comedones and absence of acneiform lesions on the chest or back. The characteristic lesions are superficial papulopustules and nodules combined with cyanotic erythema.

The pathogenesis of rosacea fulminans is unknown, but severe emotional trauma (such as the death of a family member, divorce or an accident) is often blamed [7] [8].

Dermatologic conditions that may be exacerbated perimenstrually include acne vulgaris, rosacea, psoriasis, atopic eczema and urticaria. Hormonal changes resulting in increased cutaneous vascularity, seborrhea, and dermal oedema during the perimenstrual period resemble those of pregnancy and may be related to eruption or exacerbation of these diseases [9].

Massa and Su reported that 6 of 29 rosacea fulminans cases were associated with pregnancy [10]. In the series published by Plewig et al., 3 of 20 patients had developed the disease during pregnancy [8]. Haugstvedt and Bjerke reported a flare-up of rosacea during pregnancy in 1998 [11].

In 2004, Lewis and colleagues first described a case of rosacea fulminans in pregnancy, and treatment options were mentioned for the first time in this publication [12].

As we did not perform hormonal tests in our patient and there was no history of oral contraceptive use, it is not possible to state that pregnancy or oral contraceptive us may have been a triggering factor for rosacea fulminans in our patient. However, we agree with Plewig et al., [7]. That pregnancy can be considered an exacerbating factor for rosacea fulminans, and that hormonal factors may be a trigger for rosacea fulminans during pregnancy and in females taking oral contraceptive pills.

In 2006, Ferahbaş and colleagues from Turkey, they issued a rosacea fulminans cases occurring in pregnancy and compiled the literature up to that time. They have treated this case with systemic corticosteroids and have continued to treat with topical metronidazole cream and have reported successful treatment [13].

In 2008, Cisse and colleagues reported that rosacea fulminans developed during the early period of a pregnancy obtained with IVF. In this case, hormonal treatments in IVF treatment have been shown to be the major cause of rosacea fulminans development [14].

In 2010, Jarrett and his colleagues identified three cases of rosacea fulminans in pregnancy and explained how this dermatological problem affected pregnancy outcomes [15].

In 2011, Morais e Silva FA and her colleagues published a case of rosacea fulminans in pregnancy that caused ocular perforation, and it states how serious the consequences may be [16]. In 2011, Fuentelsaz and colleagues treated rosacea fulminans case of pregnancy with azithromycin and achieved success. And they stated that the use of systemic corticosteroids is not an untreatable treatment [17]. In 2014, Wollina U. made a better comprehension of the aetiology of rosacea fulminans and compiled recent publications on management [18]. Then, in 2015, Haenen CC and her colleagues treated a patient who had rosacea fulminans in pregnancy with erythromycin and was successful [19].

When both the treatment options are given in the literature, and the treatments mentioned in the dermatology books are compiled, early and aggressive treatment is recommended, yet pregnancy poses a therapeutic dilemma; the recognised effective treatments, including retinoids, tetracycline antibiotics, antiandrogenic contraceptives and dapsone are all contraindicated during pregnancy [12].

Isotretinoin has been associated with congenital anomalies; tetracyclines are associated with discolouration of the teeth and impaired bone growth, dapsone may cause neonatal haemolysis [12].

Metronidazole is not recommended before the second trimester and although erythromycin is considered safe it is not always effective because of resistance in some bacteria [12].

Erythromycin administration is usually a safe option in pregnancy, but it has been shown in the literature that erythromycin administration is more effective in cases with comedones and bacterial reoccurrence [12].

In our case, erythromycin was not administered because of the low incidence of comedones, as we have already mentioned, and the other options were directed.

The use of systemic steroids can only be justified if the benefits outweigh the risks of

intrauterine growth retardation, maternal diabetes mellitus and hypertension [12].

For these reasons, we do not consider the use of systemic steroids in our case. Since topical medicines are generally known not to damage the fetus during pregnancy, they have been tried in the treatment of our case.

After taking other options that were not available in pregnancy, we also gave the patient topical fusidic acid and systemic penicillin antibiotics and found that there was a significant improvement.

In conclusion, there is no substantial evidence as to the mechanism by which pregnancy may trigger this conditioner whether the gender of the fetus influences the development of rosacea fulminans, but is generally accepted that hormonal changes in pregnancy play an important role. In pregnancy, the use of topical drying compounds, surgical drainage, and topical and systemic antibiotics and corticosteroids have been reported, but the best practice remains inconclusive.

We suggest a multidisciplinary approach with early diagnosis, shared obstetric and dermatology care and provision of emotional support to the mother. The pathogenesis of rosacea fulminans remains uncertain, but it is obvious that the further basic and clinical research is required to optimise the management of this rare facial dermatosis.

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