A case of confluent and reticular papillomatosis responding to systemic isotretinoin treatment

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Abstract
Confluent and reticular papillomatosis is a rarely seen dermatosis. It is characterised by the joining together of brown, hyperkeratotic papules and plaques in a reticular pattern. It is generally seen in females in the 10-35 years age range and the etiology is not fully known. The case is here presented of a 40-year old female, diagnosed with confluent and reticular papillomatosis who responded well to isotretinoin treatment.

Keywords: Confluent and reticular papillomatosis, Isotretinoin, Gougerot Carteaud disease

Introduction
Confluent and reticular papillomatosis was first defined by the French dermatologists, Gougerot and Carteaud in 1927 [1]. It is a rarely seen dermatosis which is characterised by a reticular appearance formed of hyperpigmented, smooth or verrucous papules spreading peripherally. It has a tendency to be located in sebaceous regions in particular [2]. Typical onset is stimulated by the pubertal period and it is seen in females 2.5-fold more than in males [3]. Although the majority of cases are sporadic, occasionally familial cases have been reported [4]. As the etiology has not yet been able to be fully clarified, topical agents (keratinolytics, vitamin D analogues, antifungals) and systemic treatments (oral minocycline, isotretinoin, antifungal agents) are used in the treatment of the disease. The case is here presented of a case of confluent and reticular papillomatosis successfully treated with oral isotretinoin.

Case
A 40-year old female presented at our polyclinic because of red and brown stains which had formed on the anterior surface of her neck and chest. From the anamnesis it was learned that the complaint had been ongoing for 10 years and there had been no improvement despite several courses of oral and topical antifungal treatments. In the dermatological examination, there were red-brown lesions, which were squamous maculopapules in some places, of reticular character between the breasts and more evidently on the neck and back midline (Figure 1). The Body Mass Index (BMI) of the patient was 34.5. Apart from obesity, there was no systemic disease. No abnormalities were determined in the full blood count and biochemical tests. Examination with potassium hydroxide of the scraping preparate taken from the lesion was negative. No reflex was obtained in the Wood light examination. A biopsy was taken from the patient with initial diagnoses of pityriasis versicolor, acanthosis nigricans and confluent and reticular papillomatosis (CRP). The histopathology report was consistent with CRP, an orthokeratotic epidermis with acanthosis and papillomatosis plus mild perivascular lymphocytic infiltrate in the superficial dermis (Figure 2). Treatment was started of 1mg/kg/day oral isotretinoin. At the 3-month follow-up examination, elevated cholesterol levels were determined in the patient so the isotretinoin dose was changed to 0.5mg/kg/day. At the 6-month follow-up examination, the lesions were observed to have completely recovered (Figure 3).

Figure 1. Hyperpigmented, hyperkeratotic, patches and plaques with scale on intermammary area
Figure 2. An orthokeratotic epidermis with acanthosis and papillomatosis plus mild perivascular lymphocytic infiltrate in the superficial dermis (H&E, x200).

Figure 3. At the 6-month follow-up resolution

Discussion

CRP is a rarely seen dermatosis, the etiology of which is not fully known. Impaired keratinization, abnormal response to an infectious agent ( pityrosporum, dietzia), photosensitivity, amyloidosis, genetic predisposition and endocrinopathies (diabetes mellitus, obesity, Cushing disease, thyroid dysfunction) have been held responsible in the pathogenesis [5]. In the current case, apart from mild obesity (BMI 34.5), the other etiological factors examined were normal.

CRP may be confused with many other diseases. In the differential diagnosis, acanthosis nigricans, prurigo pigmentosa, erythema dyschromium persistans, contact dermatitis, amyloidosis, dyskeratosis congenita, epidermal nerves syndrome, erythrokeratoderma variabilis, micosis fungoides, Darier disease, pityriasis rubra pilaris and tinea versicolor should be considered. Tinea versicolor, as the most common condition with which it is confused can be differentiated by examination with potassium hydroxide [5,6]. Davis et al defined CRP diagnostic criteria as brown flaky patches, some of which show reticular or papillomatous structure, involvement of the upper trunk and neck, not showing fungal structures, unresponsive to antifungal treatment and with an excellent response to minocycline [7]. The current case had 4 of these criteria and minocycline treatment was not attempted.

When the histopathology of CRP is examined, mild hyperkeratosis, papillomatosis, acanthosis and hypogranulosis are seen [1]. In the current case, in accordance with the literature, an orthokeratotic epidermis with acanthosis and papillomatosis plus mild perivascular lymphocytic infiltrate in the superficial dermis, were observed.

In literature, treatment has generally been reported as intense local or systemic antibiotics alone or combined with keratolytics such as topical retinoic acids. With the consideration that the effect mechanism of antibiotics is the antimicrobial effects, it has been suggested that these are associated with anti-inflammatory, immunomodulator and anti-proliferative effects [5,7,8]. In addition, as keratinocyte proliferation has a place in the etiopathogenesis, high and low doses of oral isotretinoin, etretinate, topical tazarotene and topical retinoic acids have been used because of the antiproliferative effects and have been found to be effective. In the current patient, systemic isotretinoin treatment was started in respect of being useful for the nodulocystic acne and despite decreasing the medication, full recovery was obtained and no recurrence was observed.

In conclusion, CRP is a rarely seen disease which may accompany obesity. An excellent response can be obtained with systemic isotretinoin, which is a readily available agent.

References