



## Confluent Hyperpigmented Plaques in a 12-Year-Old Adolescent

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### Case Report

We present the case of a 12-year-old patient of Moroccan origin, without any information regarding the biological parents. From her medical history, we highlight a syndrome of generalized congenital lipodystrophy (Berardinelli-Seip syndrome). This consists of an absence of adipose tissue alongside an alteration in the metabolism of fats and carbohydrates [1]. The patient presented a long time, velvety pigmentation on the neck and armpits that had been classified as acanthosis nigricans related to her genetic disease (Figure 1). However, on her annual clinical assessment, she complained about the appearance of new asymptomatic, hyperpigmented, confluent skin lesions with reticulated edges. It develops progressively in a couple of months, starting with the pectoral region and upper back (Figure 2). No response to any topical depigmentation was observed.

Wood's light examination and direct Potassium Hydroxide (KOH) examination were negative. A biopsy of the cervical region was performed, which revealed epidermal papillomatosis with orthokeratosis hyperkeratosis without acantholysis, associated with basal melanin hyperpigmentation (Figure 3). After a treatment with topical retinoids and oral Josamycin 500 mg/24 h for 3 months, the lesions showed an evident improvement.

### Diagnosis

#### Confluent and Reticulated Papillomatosis (CRP)

CRP is a not very well-known dermatosis that usually begins in adolescence. It is more frequent in women and in dark-skinned patients, as in our case. Some theories have been proposed to explain its etiopathogenesis: Genetic or acquired keratinization abnormalities, photosensitivity or underlying endocrine disorders such as obesity, diabetes or thyroid dysfunction. These ones would cause an abnormal-epithelialization related to the excess of certain hormones [2]. In our patient, due to the genetic lipodystrophy syndrome, we are led to consider the possible relationship with endocrine disorders.

In order to make a diagnosis, Lee et al. [3] defined three main criteria needed, which include: Hyperpigmented plaques in the centre of the chest or back with a reticulated pattern on the edge. Negativity for fungi in the KOH stain or the absence of hypha in the biopsy. Histology showing papillomatosis, hyperkeratosis and acanthosis with hypergranulosis.

As a topical treatment, good results have been observed with selenium sulphide, vitamin D derivatives and topical retinoids [4]. Oral isotretinoin at doses of 1 mg/kg/day to 2 mg/kg/day has also been used with favorable responses [3]. Oral doxycycline presents a success rate of over 70% at doses of 100 mg/day and lead, as in our patient, to rapid resolution of the lesions in 1 or 2 months (Figure 4). It is believed that its antiproliferative and anti-inflammatory action could play an important role [5].

### Differential Diagnosis

- **Acanthosis Nigrans (AN):** It is one of the main differential diagnoses, with a similar histology, but different clinical features. In AN, the lesions are thicker with a velvety hyperpigmentation and are mainly located on the flexures [6] (Figure 1). The onset- aged is earlier and it is more frequently related to endocrine disorders. On rare occasions, as in our case, both entities can coexist.
- **Pitriasis Versicolor:** It is a superficial fungal infection caused by *Malassezia* yeasts. The

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Figure 1: Genetic disease.



Figure 2: Upper back.

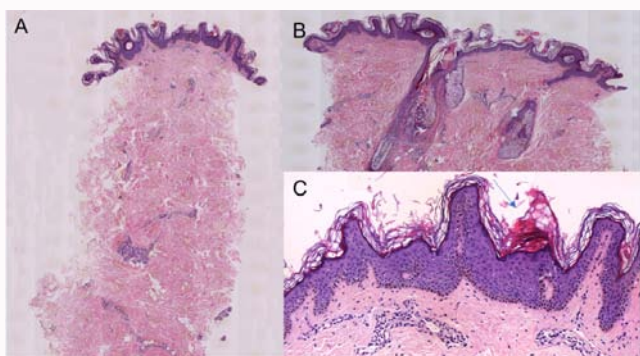


Figure 3: A) Punch biopsy with scarce hypodermis and increased number of collagen fibers. B) Micro abscessing neutrophilic infiltrate in the sebaceous gland, with mild associated infundibular dilatation. C) Basal hyperpigmentation and focal compact hyperkeratosis (arrow).

lesions are dyschromic macules, with furfuraceous scale, that usually affects the trunk and upper extremities [7]. Unlike CRP, it has a favorable response to topical and oral antifungals.

- **Dermatitis Neglecta:** It consists on reticulated hyperpigmented plaques with a dirty appearance that disappear after rubbing with ethyl alcohol [8]. The lesions arise as a result of a sustained accumulation of sebum and microorganisms.



Figure 4: Rapid resolution of the lesions in 1 or 2 months.

- **Macular Amloidosis:** Grayish reticular hyperpigmented macules frequently located on the back. Histology shows Amyloid deposition as amorphous eosinophilic material in the papillary dermis [9].
- **Dowling-Degos:** Genodermatosis characterized by reticular pigmentation in the folds. Histopathologically, the presence of digitiform proliferations in the epidermis and in the wall of the pilosebaceous follicles are characteristics [10].

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