Journal of Pigmentary Disorders
ISSN: 2376-0427

de Lima Grynszpan et al., Pigmentary Disorders 2015, 2:4
DOI: 10.4172/2376-0427.1000175

Case Report Open Access

Partial Unilateral Lentiginoses

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Abstract

The authors present a case of a rare pigmentation disorder, partial unilateral lentiginosis (PUL), which is characterized by multiple lentigines located in only one side of the body with histopathology of lentigine.

Keywords: Lentigines; Partial unilateral lentiginosis; Pigmentation

Introduction

Partial unilateral lentiginosis (PUL) is a rare pigmentation disorder characterized by multiple lentigines located in only one side of the body, which histopathologically show typical finding of lentigine [1,2]. It is characterized by small, well delimited, hyperchromic macules, distributed in a limited area of the body, which can affect one or more dermatomes and stop at the median line. PUL comprises a group of small macules which appear on a skin of normal aspect and with histological findings of lentigo simplex [3]. If the distribution of the lesion is limited to a single dermatome, it may be confused with nevus spilus, also known as lentiginous nevus or zosteriform nevus [4,5]. Nevus spilus manifests clinically as a grouping of numerous lenticular elements, brownish macule or papule on a uniform hyperpigmented background, with diameters between 1 and 20 cm subject to appear both in childhood as in adulthood [5]. In histopathology, the lenticular elements correspond to junctional melanocytic nevus, or compound or Spitz' nevus and, in the hyperpigmented background, an increase of the epidermal melanin without melanocyte proliferation is observed [6,7]. Nevus spilus is a pigmented nevus accompanied by a melanocytic nevus (nevus over nevus) [8].

In 1904, McKelway was the first author to study a case of a patient with unilateral distribution of lentigines, but Pickering was first to introduce the term partial unilateral lentigines in 1973 [5,9].

PUL does not usually have a familial relationship; the lesions almost always start in childhood without being present at birth. In the majority of cases it affects the upper part of the body rarely affecting the lower portion, thus the denomination of partial. When affecting both parts, it is called complete. It may affect either the right or the left side, and generally begins in the occipital region. It is more common in white skin and rare in black skin. To present, only one case with family history was reported [10]. The term lentiginosis is applied when the lentigines are present in great number with distinct distribution.

Case Report

A 53 year-old black, retired female, born in the State of Bahia, referred spots on hand, neck, lips and trunk, asymptomatic for 30 years, after pregnancy, without any growth, change of color pattern or size. As comorbidities, she presented essential arterial hypertension, seronegative rheumatoid arthritis, and arthrosis in right foot. She had a melanoma in the left nose wing, which had been excised a year before (Figures 1 and 2). She uses subcutaneous methotrexate 0.6 ml and omeprazole, losartan and oral atenolol. No family members have similar lesions.

At dermatologic examination, small brownish macules were observed on the back of the right hand, right cervical region, right upper chest, right lower lip and right forehead up to the medial line,

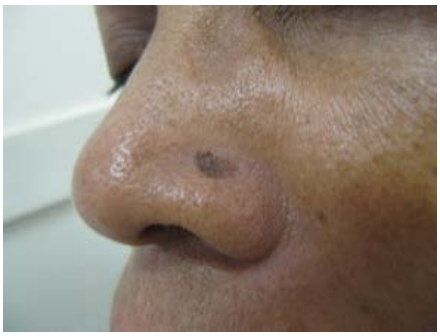

Figure 1: Melanoma in the left nasal wing, contralateral to PUL before its surgical excision a year before.

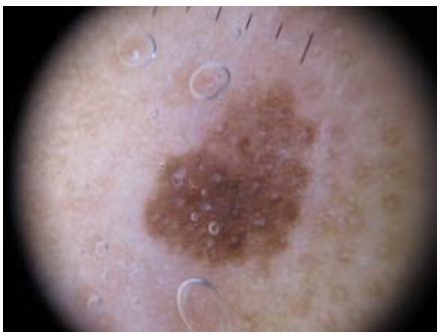

Figure 2: Dermatoscopy of the melanocytic lesion in the left nasal wing before biopsy.

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Received March 16, 2015; **Accepted** April 02, 2015; **Published** April 06, 2015

Citation: de Lima Grynszpan R, Corbellini JPN, Gaiser BM, e-Silva MR (2015) Partial Unilateral Lentiginoses. Pigmentary Disorders 2: 175. doi:10.4172/2376-0427.1000175

respecting the upper right half (Figures 3-7). The left nasal wing presented a surgical scar from the excised melanoma.

Complementary exams

Incisional biopsy of the left nasal wing: atypical focal intraepidermal melanocytic proliferation, associated to hyperpigmentation. A small perivascular mononuclear inflammatory reaction is noted as well as several melanophages in the upper dermis.

CK "pool": diffuse immunoreactivity in the epidermis.

Figure 3: Multiple lentigines in face – right side with UV light highlighting the lentigines.

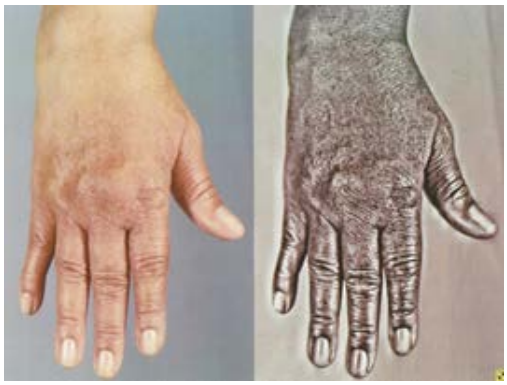

Figure 4: Multiple lentigines on right hand – right side with UV light highlighting the lentigines.

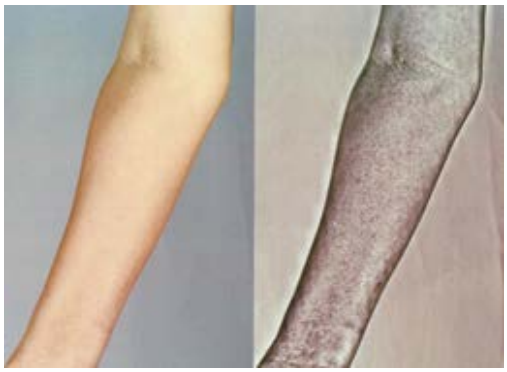

Figure 5: Multiple lentigines on right arm – right side with UV light highlighting the lentigines.

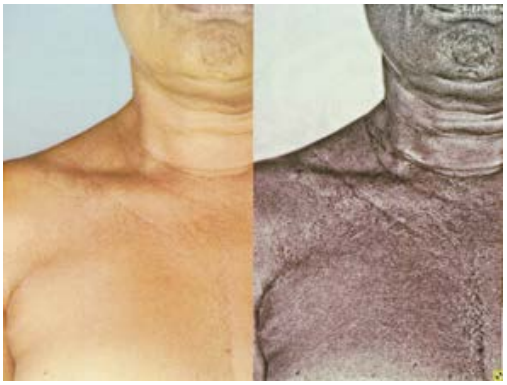

Figure 6: Multiple lentigines on right upper chest and neck – right side with UV light highlighting the lentigines.

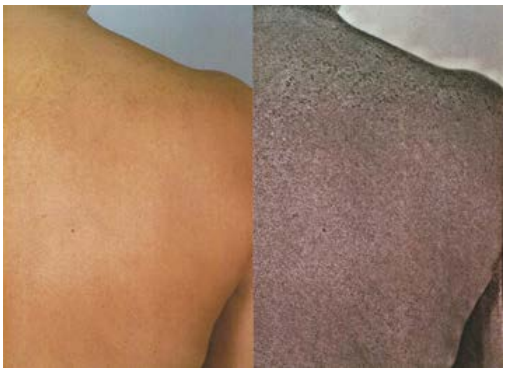

Figure 7: Multiple lentigines on right dorsum – right side with UV light highlighting the lentigines.

S100 and HMB-45: immunoreactivity in cells distributed in the lower epidermal layers without confluence, however apparently in increased numbers.

Excisional biopsy in the left nasal wing: melanoma in situ, Clark's Level 1, observed margins free of malignancy.

Biopsy of dorsum: lentigo solaris.

Colonoscopy: semi-pedunculated polyp with 1.2 cm with reddish surface in the sigmoid. Pathology: tubular adenoma with low degree of atypia (moderate dysplasia).

Opinions were requested from Gastroenterology, which did not find a polyposis syndrome, removed an adenoma with low degree of atypia and indicated a new colonoscopy in three years; and to Ophthalmology, which did not detect Lisch nodule, or hyperpigmentation of the retina.

Discussion

Lentigo simplex is a small well-delimited pigmented macula entailed by the presence of an increased number of isolated melanocytes alongside the dermoepidermal junction, mainly at the sides and bases of elongated and thin interpapillary cones. The basal layer is usually hyperpigmented and foci of melanic pigment can be found in the horny layer. Discrete fibroplasia, melanophages and sparse mononuclear inflammatory reaction are present in the papillary dermis. Despite being stable lesions, it is believed that some lentigines evolve into junctional melanocytes or into compounds with lentiginous

pattern. Lentigo simplex can be induced into the patient by PUVA treatment. When affecting the lips, the Peutz-Jeghers syndrome should be investigated [11].

Solar lentigines are completely different lesions, with broad limits and greasy surface. Histologically, they represent a proliferation of pigmented keratinocytes, without increase in the number of melanocytes. By definition, they occur in the skin with solar aging.

Partial unilateral lentiginosis is characterized by numerous brownish, macules or slightly elevated papules, with sizes varying between 1 to 15 mm in diameter, asymptomatic, over the normal skin [7].

There are reports of PUL associated to a central nervous system disease, such vascular malformation and mental retardation. This way an alteration in the embryonic development before crest closure could be suggested as a cause, however, there is no alteration in the development [12].

Melanocytic cells, as the nervous cells, have the same ectodermal origin. Allegue et al. reported the somatic post-zygotic mutation that would explain the coincidence with segmental neurofibromatosis (NF) [13].

Some authors suggest that PUL could precede neurofibromatosis. Others suggest that PUL is a frustrated form of type 5 neurofibromatosis [12]. The researchers disagree regarding the etiology, admitting the possibility of dealing with mosaicism, partial chimerism or a post-zygotic mutation, the same explanation for segmental neurofibromatosis [13-16].

PUL can be associated to a series of abnormalities, affecting several systems such as cardiovascular, muscle-skeleton, gastrointestinal, auditory and respiratory system. Cutaneous abnormalities also occur, such as vitiligo, café-au-lait stain, cutis marmorata and acanthosis nigricans. It should be mentioned that all these alterations are considered as findings without an established relation with the disease. Several of them may be categorized in separate syndromes such as Leopard, LAMB, NAME, and Peutz-Jeghers syndromes. All have lentigines bilaterally, are autosomal dominant and present multiple abnormalities, in contrast with PUL that presents unilateral distribution without known interaction [2,17-20].

Among the systemic associations, the following were described: ipsilateral rigid talipes cavus, mental retardation and falciform anemia, goiter, since thyroid C cells also have their origin in the neural crest [3,12,21]. PUL has been associated with cutaneous abnormalities, such as nevus depigmentosus and blue nevus [22]. There are exceptional clinical case reports with eyes involvement and nevus of Ota, among others [23-26].

It is convenient to perform complementary exams which should include routine biochemistry, thyroid profile, FSH and LH, and to refer to evaluations by a Neurologist, Oftalmologista, Cardiologist and Gastroenterologist, in order to detect eventual associations [27].

Treatment is indicated for the improvement of the esthetic aspects of the patients. The lentigines can be treated with topical agents, as a combination of tretinoin and hidroquinone in cream, producing a clearing after a few months of application. Laser therapy is also effective, as Q-switched Nd:YAG laser and HGM K1 krypton laser [10]. Long term prognosis is unknown and the risk of malignant transformation is very low [28].

Conclusion

LUP is a rare pigmentation disorder, which should be well known by dermatologists, since it may have association to other diseases and individual lesions may evolve into melanomas, despite its rarity. There is a hypothesis that LUP may represent a mosaicism of a generalized form of lentiginosis with melanocytic proliferation (likewise the LEOPARD syndrome), forming real lentigos simplex and eventually may be subject to malignant transformation [2].

LUP is an uncommon, benign and asymptomatic disorder that may pass unperceived and is often underdiagnosed. It may be associated to systemic and cutaneous abnormalities, and the Dermatologist must recognize it in order to perform an appropriate handling of the patient [27].

There are few cases described in literature and some associated with lentiginous syndromes or segmental neurofibromatosis; therefore requiring the need to investigate its coexistence [29].

It is of vital importance to carry out an exhaustive clinical historical, that includes a complete physical examination and pertinent complementarity exams to reach a correct diagnosis and the appropriate management and follow-up of the patient.

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