Case Report

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Laugier-Hunziker Syndrome: A Rare Cause of Oral and Nail Pigmentation

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Abstract

Laugier-Hunziker syndrome is a rare, benign, acquired pigmentary condition. It is a hereditary idiopathic lenticular mucocutaneous pigmentation. The exact etiology and pathogenesis involved in LHS remain still uncertain. It involves the lips, oral mucosa, acral surfaces, nails and perineum. While patients with LHS may manifest pigmentation in all of the aforementioned areas, most present with pigmentation localized to only a few of these anatomical sites.

We report a case of a morrocan patient with an acquired oral pigmentation with homogenous pigmentation of the toenails and longitudinal melanonychia of the fingers nails.

Keywords: Laugier Hunziker syndrome; Melanonychia; Nails; Oral mucosa; Pigmentation

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Introduction

Laugier-Hunziker Syndrome (LHS) is a rare, benign, acquired pigmentary condition [1]. It is a hereditary idiopathic lenticular mucocutaneous pigmentation [2], which mainly affecting lips, oral mucosa, perineum and acral area, frequently associated with longitudinal melanonychia [3].

We here in report a case of a morrocan patient with an acquired oral pigmentation withh omogenous pigmentation of the toenails and longitudinal melanonychia of the fingers nails.

Case Report

We report a 48 years old moroccan women, with no medical history and no familial pigmentary disorder. There was no drug intake.

She had consulted for an pigmentation of the toenails evolving since 2 years. The clinical exam found an homogenous pigmentation of the 10 toenails with a pseudo hutchinson's sign nail (Figure 1). She had also a longitudinal lateral melanonychia of the 2 nd left finger'snail (Figure 2). We had noted also an hyperpigmentation of the oral mucosa and the palate (Figure 3), with a flat pigmentedlesion of the lower back (Figure 4). Cutaneous biopsie of this lesion showed an exaggeration of the basal melanic pigmentation without interface lichenoid reaction. She had no pigmentation in the lips or in the perineum. Cortisol level, blood ionogram and NFS were normal.

Discussion

Laugier-Hunziker syndrome has more frequently been reported in the Asian population and displays a higher incidence in the Chinese population. Cases have also been reported in European regions such as France and Italy [2].The exact etiology and pathogenesis involved in LHS remain still uncertain [1].

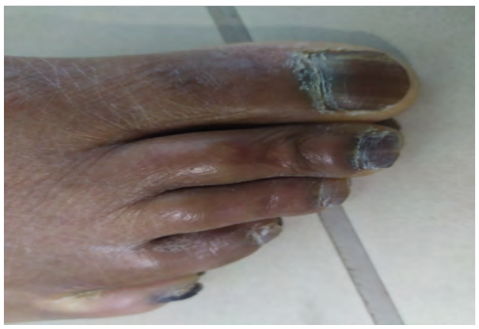

Figure 1: Homogenous pigmentation of toenails.

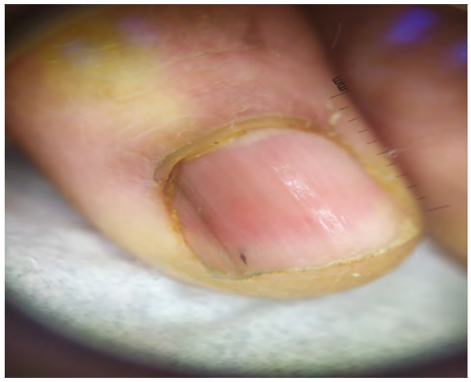

Figure 2: Longitudinal melanonychia of the 2nd left finger'snail.

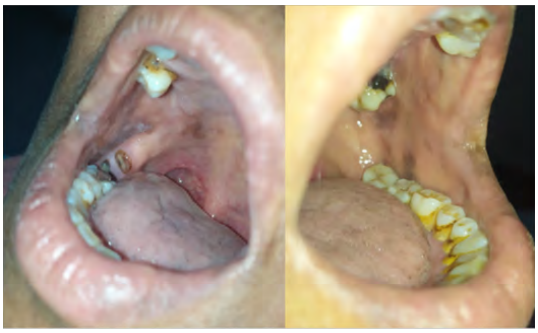

Figure 3: Oral mucosa pigmentation and palate pigmentation.

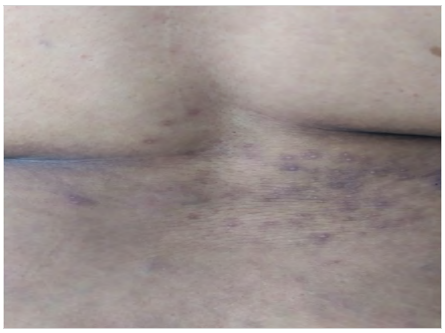

Figure 4: Pigmentation of the lower back.

It is an acquired hyperpigmentation involving the lips, oral mucosa, acral surfaces, nails and perineum. While patients with LHS may manifest pigmentation in all of the aforementioned areas, most present with pigmentation localized to only a few of these anatomical sites[3].

Nail involvement is seen in two-thirds of cases and can be divided into 4 types based on the extent of pigmentation : single 1 to 2 mm longitudinal streaks, which is present in the 2nd left finger'snail of our patient, double 2 to 3 mm longitudinal streaks on the lateral parts, homogenous pigmentation involving radial or ulnarhalf or complete pigmentation [2], which is present in the toenails of our patient .

Initially, there was still no established evidence of systemic complications or increased cancer riskso far [1]. However, recent reports of associated malignancies have suggested cancer screening, particularly in adult cases [2].

LHS can be associated with esophageal melanocytosis, actinic lichen planus, hypocellular marrow, thrombocytopenia, pancreatic cancer, etc [4-6].

These features indicate that the pigmented disorder could be due to increased melanocytic activity rather than to an increased number of melanocytes. The main differential diagnosis of LHS includes Peutz-Jeghers syndrome (PJS), Addison'sdisease, McCune-Albright syndrome, lichen planus and drug-induced pigmentation [1].

The goal of therapy is purely cosmetic in case of Laugier-Hunziker syndrome. Treatment options include cryotherapy, Q-switched Nd:YAG laser, Q-switched Alexandrite laser, erbium: YAG laser, CO2 laser, and diode laser [2,5].

Conclusion

Laugier-Hunziker syndrome is a rare disease and diagnosis may be challenging. Through our case we would like to put the light on a syndrome which can present as an acquired homogenous pigmentation of the nails. His association with malignanciess warrants evaluation in suspected cases.

References

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