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Original Research Paper

Dermatology

LAUGIER HUNZIKER SYNDROME- A RARE CASE REPORT

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ABSTRACT Laugier Hunziker syndrome is a rare acquired disorder characterized by longitudinal melanonychia which can involve few or all the nails and pigmented lesions over the oral cavity. This is considered to be a benign condition with no malignant potential. Hence proper examination is required to differentiate it from other conditions in which invasive treatment is warranted. Here, we report a 39-year-old female who presented with pigmented macules and patches over the lower lip, angles of mouth and gingiva of the upper incisors and canine with longitudinal melanonychia.

KEYWORDS :

INTRODUCTION:

Laugier Hunziker syndrome is portrayed by pigmented lesions which classically occur in the oral mucosa with melanonychia striata.1 Other sites where the pigmentation can be seen are the palms and soles, neck and trunk.2 The differential diagnoses to be thought are Peutz -Jeghers syndrome, Addison's disease and other syndromes associated with lentigines. Proper diagnosis of this condition is required as this condition requires no treatment apart from cosmetic purposes, whereas the others require invasive investigations and early treatment.

CASE REPORT:

We report a 39-year-old female who presented with pigmented lesions over the oral cavity for 5 years. The patient had noticed it insidiously and they were slowly progressive and attained the present size and number. After history taking and proper examination, the patient was found to be having no other lesions elsewhere in the body and complaints apart from the oral pigmentation. The family history was not significant. The patient had initially consulted a dentist and was referred to a medical gastro-enterologist to rule out Peutz- Jeghers syndrome.

The vital parameters and systemic examination were normal. The cutaneous examination showed pigmented macules and patches involving the lower lip, angles of the mouth and gingiva over the upper incisors and canine. Longitudinal melanonychia was present in the finger nails. There were no other lesions in the body. The routine investigations were normal. The patient had already performed investigations like ultrasound abdomen, upper GI endoscopy and colonoscopy, the results of which were normal.

The diagnosis of Laugier Hunziker syndrome was made and the patient was explained about the condition and reassured. The patient did not opt for Laser therapy as she was not bothered for cosmetic reasons.

FIGURE 1

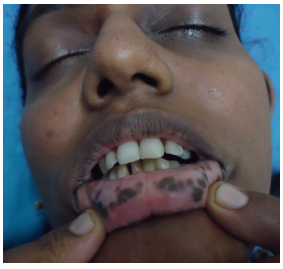


FIGURE 2

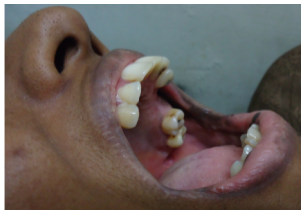


FIGURE 3

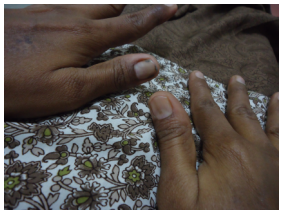
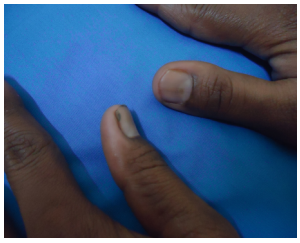


FIGURE 4



FIGURE

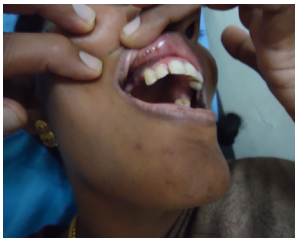


FIGURE 6

DISCUSSION:

Laugier Hunziker syndrome is a benign acquired disorder of hyperpigmentation. The condition can occur in a sporadic fashion most commonly or sometimes it can be inherited in an autosomal dominant manner. The etiology of this condition is unknown[3] but it has been proposed that there is an alteration in the function of melanocytes which show increased melanosome number and subsequent transfer to the basal cells.

The condition begins in the early or middle adulthood. The condition manifests as asymptomatic hyperpigmented macules which usually measure less than 5mm in diameter and may coalesce to form patches as seen in our case. The lesions have a well to ill-defined margins. The common locations are the lips, buccal mucosa and hard palate. The other areas in the oral cavity which can be involved are the soft palate, tongue, gingiva and floor of the mouth. Rarely, the disorder can also affect other sites like conjunctiva, trunk, genitals and perioral area.

The nail changes are seen in approximately 60% of the cases and present as longitudinal melanonychia. The nail finding can be present in few or all the nails. Pseudo- Hutchinson sign can be seen in few cases.

The histopathology of the pigmented macules shows epithelial acanthosis with pigmentation confined to the basal cells. The

melanocytes are normal in number, morphology and distribution. Pigment laden macrophages can be seen in the papillary dermis.[4]

The important differential diagnoses are Peutz- Jeghers syndrome and Addison's disease. Peutz Jeghers syndrome is associated with pigmentation around nose and mouth and associated with hamartomatous gastrointestinal polyposes which have a high risk of turning into malignancy. Addison's disease is characterized by diffuse hyperpigmentation of the skin, creases, knuckles and mucous membranes with systemic features like hypotension, dehydration and abdominal pain.

The other less common differentials are LEOPARD syndrome, McCune Albright syndrome, Gardner syndrome and heavy metal exposure.

Treatment includes reassurance of the patient and laser treatment can be offered for cosmetic purposes. Lasers like Q switched Nd Yag laser and Q switched Alexandrite laser [5] can be offered for the pigmented lesions with sun protection measures without which the chance of recurrence is high.

CONCLUSION:

Laugier Hunziker syndrome is a rare benign pigmented condition commonly involving oral mucosa and must be differentiated from other conditions mentioned above. This syndrome is not confined to a particular race and can occur universally. The recognition of this condition is necessary to avoid unnecessary invasive investigations and patient's agony.

LEGENDS TO FIGURE

FIGURE 1& 2: Pigmented macules over the lower lip
FIGURE 3: Pigmented patches over the angle of mouth
FIGURE 4 & 5: Longitudinal melanonychia on the thumb nail
FIGURE 6: Pigmented patches over the gingiva of upper incisors and canine

CONFLICTS OF INTEREST: None

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