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CASE REPORT

DIAGNOSTIC QUANDARY WITH LAUGIER-HUNZIKER SYNDROME

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ABSTRACT

Laugier-Hunziker syndrome being a rare pigmentation disorder chiefly involving lips and nails has so far been coincidentally diagnosed. In Nepal, Laugier-Hunziker syndrome has not been reviewed very well, and no case has been reported in the literature till date. To the best of our knowledge, the current report is the only one in the Nepalese scientific community but addressing the age of the patient it stands with a diagnostic quandary.

INTRODUCTION

Laugier-Hunziker syndrome or Idiopathic lenticular mucocutaneous syndrome as it was known in the past, has so far been recognized as a benign disease of unknown etiology with no systemic involvement.[1,2] Initially it was reported as a mucocutaneous pigmented disorder with involvement of lips and oral mucosa but later it was found to be associated with melanonychia as well.[2] It's benign clinical presentation and resemblance to other more severe mucocutaneous pigmented lesions makes it significant to be recognized instantly so as to avoid any undue apprehension to the patient.[1]Here we are presenting an exquisite case report of an 8 year old male patient with clinical findings similar to that seen in Laugier-Hunziker syndrome which creates a state of dilemma with confirmatory diagnosis.

CASE REPORT

An 8 year old male patient, resident of Nepal presented with features closely resembling that of Laugier-Hunziker syndrome. The patient was accompanied by his parents with a concern related to malocclusion. On examination, numerous irregular hyperpigmented macules of brown color with grayish hue, having indistinct margins and ranging in size from few millimeters to around 0.4 - 0.5 cm were noted involving both the lips and also present bilaterally over the buccal mucosa (Figure 1).

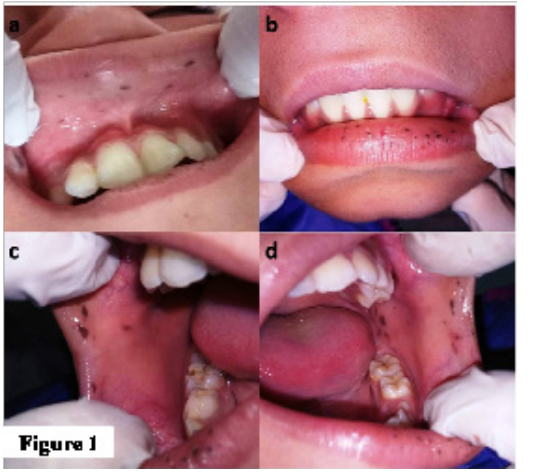

Figure 1: Multiple, discrete, well defined grayish brown colored macules seen at different sites within the oral mucosa

During the full body skin examination, he was found to have similar hyperpigmented macules around the fingernails.

Remarkable feature was the presence of homogenous pigmentation over his fingernails and toenails, extending and involving even the nail folds thereby exhibiting a pseudo-Hutchinson's sign (Figure 2).

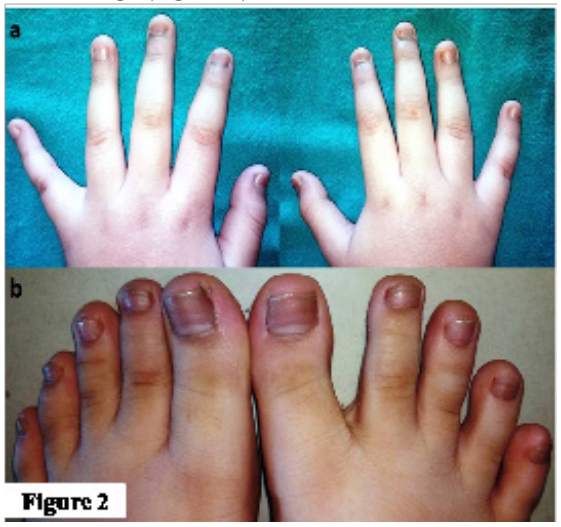

Figure 2: Homogenous pigmentation involving all the fingers and toe nails and nail folds

A diagnosis of exclusion was made on basis of clinical presentation as Laugier-Hunziker syndrome and an incisional biopsy was done to rule it out from any other serious pigmented disorder. On histopathological examination the basal cell layer showed hyperpigmentation along with the presence of melanin incontinence. Cellular atypia or increase in number of melanocytes was not evident; thereby confirming the benign nature of the lesion (Figure 3). Taking into account the clinical and histopathological reports, a final confirmatory diagnosis as Laugier-Hunziker syndrome was made. The patient was further assured and informed that there is no obligation for any treatmentat present but owning to the age of the patient his parents were advised to maintain regular clinical follow-up to assure that the patient doesn't develop intestinal polyposis later in life, thereby ruling out any possibility of Peutz-Jeghers syndrome.

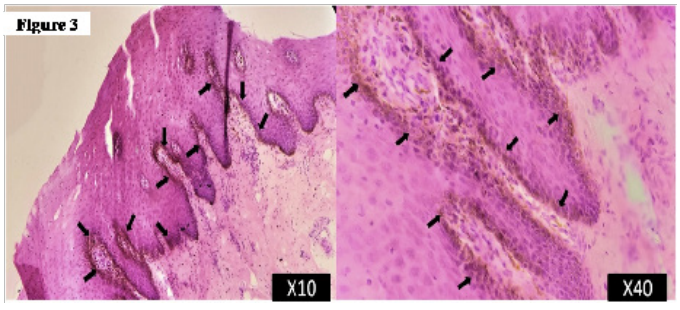

Figure 3: Histopathological image shows hyperpigmentation restricted to basal cell layer with melanin incontinence, without cellular atypia

DISCUSSION

After reviewing the literature we found that, no more than 250 cases of Laugier-Hunziker syndrome have been reported and published so far.Including our case here, majority of the other case reports also proclaimed that it was an accidental finding and the lesions were progressive yet benignin nature.[3-6] Most authors have stated that Laugier-Hunziker syndrome manifests frequently in young adults and middle age group people and has a female preponderance, contrary to that the present case was seen in an 8 year old male patient.[4-6]

The clinical differentials for this include Peutz-Jeghers syndrome, LAMB syndrome, LEPORD syndrome, McCune Albright syndrome, Addison disease and Gardner syndrome, which were ruled out based on the history and clinical examination.[5]Out of these, Peutz-Jeghers syndrome is an autosomal dominant disorder which closely shares its clinical and histological features with Laugier-Hunziker syndrome making the confirmatory diagnosis questionable at times. However, there are certain differentiating features between the two like positive family history, early onset, presence of pigmentation over face, hands, nails, feet, along with intestinal polyposis which are suggestive of Peutz-Jeghers syndrome while Laugier-Hunziker syndrome shows both oral and nail pigmentation.

Though rarely Laugier-Hunziker syndrome has been reported in such a young patient and acknowledging the fact that the median age for gastrointestinal presentations in Peutz-Jeghers syndrome is 11-13 years[7-9] complete exclusion of Peutz-Jeghers syndrome or other pigmentary disorders could not be done. Due to the absence of clinical signs towards Peutz-Jeghers syndrome we did not find it reasonable to subject the patient to vast invasive investigations or molecular genetic studies at the moment.

Considering the clinical presentation in our patient showing hallmark features like no family history and both oral and nail pigmentations; for now it has been considered to be a case of Laugier-Hunziker syndrome Class 1 Type IV and patient has been advised follow-up for regular clinical screening.[10]

CONCLUSION

Our patient presented with a rare syndrome at a very early age, which certainly is not much known by general dentists. However the dentists must be aware of Laugier-Hunziker syndrome as a benign pigmentation disorder to avoid misdiagnosis and also to avoid the non essential trials on patient. Thorough clinical examination and history can help in its early diagnosis and differentiation from other more concerning disorders.

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