Oral Melanosis: A Case Report of Peutz-Jegher's Syndrome in Suva, Fiji

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Abstract: This paper reports on a case of oral melanosis resulting from Peutz-Jeghers Syndrome, a hereditary disease characterized by harmatomatous polyposis and by mucocutaneous melanic pigmentation, in a 14-year-old Fijian girl with a family history of the disease. The patient underwent surgery for treatment of an intestinal obstruction due to a small intestine intussusception. Recognition of the characteristic pigmentation by dentists may lead to early diagnosis of associated harmatomatous polyps or neoplastic disease that may be life threatening.(PHD, 2003; 10 (1), Pages 55-56)

Introduction

Peutz-Jeghers syndrome formerly known as hereditary intestinal polyposis syndrome, was described by Jeghers[1], characterized by familial generalized intestinal polyposis and pigmented spots on the face, oral cavity and sometimes the hands and feet. Peutz-Jeghers syndrome is inherited as an autosomal dominant trait with variable incomplete penetrance.[2,3] This paper presents a case of oral pigmentation of the Peutz-Jeghers syndrome; the aim of the paper is to increase the awareness of dentists about this clinical entity.

Case Report

A 14-year-old Fijian girl from a family of five was admitted into the surgical unit of the Colonial War Memorial Hospital, Suva, Fiji with a history of recurrent spasmodic abdominal pain. Patient also gave the history that the father as well as one of her siblings suffered a similar condition. A diagnosis of Peutz-Jeghers syndrome was made as a result of the clinical observation of pigmented spots on the lips (Figure 1A), oral cavity, hands (Figure 1B) and toes, combined with the signs and symptoms of intestinal obstruction. The intestinal intussuception led to the surgical resection of a segment of small intestine. At surgery, polyps were seen on the resected portion of the intestine. Surgical specimen was submitted for histological examination; the diagnosis of Peutz-Jeghers syndrome was confirmed.

Figure 1A

and oral pigmentation induced by premarin.[4-6] As the number of reports increased, the association of cutaneous and mucosal pigmentation, multiple gastrointestinal harmatomatous polyps with or without nasal polyp or oral papillomas have been well described as Peutz-Jeghers syndrome.[1, 7-10] Mutations in LKBI/STK11 gene have been identified as the cause of Peutz-Jeghers syndrome.[3, 11]

The current opinion is that patients with Peutz-Jeghers syndrome can develop malignancies in the gastrointestinal tract and tumors in other organs as documented by several authors.[2, 11-13] Guldberg P et al[11] in their study, explained that mutations in LKBI/STK11 as manifested in patients with Peutz-Jeghers syndrome could contribute to tumorigenesis in a small fraction of malignant melanomas, contrary to the old belief that the polyps in this syndrome have little or no malignant potential. This case report is in agreement with the literature finding that generalized intestinal polyposis with oral pigmentation (Peutz-Jeghers syndrome) has a familial history and not sex-linked. Up till now, there is paucity of literature on this syndrome in Fiji. In spite of the low frequency of this lesion, the dentist is in a strategic position to detect this syndrome early. Thus early diagnosis of suspected cases of Peutz-Jeghers syndrome will facilitate early referral to the surgical team before the onset of intestinal obstruction.

Figure 1B

pigmentation by dentists will in no doubt lead to early diagnosis of associated harmatomatous polyps or neoplastic disease that may be life threatening. The case review will increase the awareness of dentists about this clinical entity.

Reference

1. Shafer WG, Hine MK, Levy BM. Developmental disturbances of oral and paraoral structures. *In A Textbook of Oral Pathology.* 4th ed. Philadelphia: WB Saunders; 1983. p18-20.
2. Kitagawa S,Townsend BL, Hebert AA. Peutz-Jeghers syndrome. *Dermatol Clin.* 1995; 13(1): 127-133.
3. Boardmen LA, Couch FJ, Schroeder JJ, Thibodeau SN. Genetic heterogeneity in Peutz-Jeghers syndrome. *Hum Mutat.* 2000; 16(1): 23-30.
4. Lamey PJ, Nolen A, Lewis MA, Rademaker M. Oral presentation of the Laugier-Hunziker syndrome. *Br Dent J* 1991; 171(2): 59-60.
5. Satriano RA. Oral manifestations of idiopathic lenticular mucocutaneous pigmentation (Laugier-Hunziker syndrome): a clinical, histopathological and ultrastructural review of 12 cases. *Oral Dis* 1999; 5(1): 80-86.
6. Perusse R, Morency R. Oral pigmentation induced by Premarin. *Cutis* 1991; 48(1): 61-64.
7. Laviada AF, Reyes PA, Palomo CA. Multiple gastrointestinal familial poliposis (PJS). Report of a clinical case. *Rev Gastroenterol Mex* 1975; 40(2): 76-8 3.
8. Cerqua N, Piccirillo E, Spaziani G. Rare manifestation of nasal polyposis in the Peutz-Jeghers syndrome. *Acta Otorhinolaryngol Ital* 1999; 13(4): 333-338.
9. Bermejo FA, Mas Bermejo C, Garcia BC. Peutz-Jeghers syndrome (mucocutaneous melanosis associated with gastrointestinal polyposis). Presentation of 5 cases presenting in the same family. *Av Odontoestomatol.* 1991; 7(1): 11-17.
10. Oshiro T, Maruyama Y, Mima M. Treatment of pigmentation of the lips and oral mucosa in Peutz-Jeghers syndrome using ruby and argon lasers. *Br J Plast Surg.* 1980; 33(3): 346-349.
11. Guldberg P, Thorstraten P, Kirkin AF, Zenthen J. Somatic mutation of the Peutz-Jeghers syndrome gene, LKBI/STKII in malignant melanoma. *Oncogene* 1999; 18(9): 1777- 1780.
12. Rodu B, Martinez MG Jr.Peutz-Jeghers syndrome and cancer. *Oral Surg Oral Med Oral Pathol* 1984;58(5): 584-588.
13. Mullhaupt B, Bauerfeind P, Kurrer MO, Fried M. Anal squamous cell carcinoma in a patient with Peutz Jeghers syndrome. *Dig Dis Sci* 2001; 46(2):273-277.
14. Pennazio M, Rossini FP. Small bowel polyps in Peutz-Jeghers syndrome: management by combined enteroscopy and intraoperative enteroscopy. *Gastrointest-Endosc.* 2000; 51 (3): 304-308.
15. Lin BC, Lien JM, Chen RJ, *et al.* Combined endoscopic and surgical treatment for the polyposis of Peutz-Jeghers syndrome. *Surg Endosc* 2000; 14 (2): 1185-1187.
16. Taiwa K, Matsubara H, Isa T, *et al.* Combined endoscopic and surgical treatment for multiple polyps of the small intestine in Peutz-Jeghers syndrome: a case report. *Surg Laparosc Endosc Percutan Tech,* 2000; 10 (6): 409-411.

Recognition of the characteristic pigmentation by dentists will in no doubt lead to early diagnosis of associated harmatomatous polyps or neoplastic disease that may be life threatening.

It is the patient rather than the case which requires treatment
(Robert T. Morries in Doctors versus Folks)