原文題目(出處)：	Seven-year follow-up of Peutz-Jeghers syndrome. Case Rep Dent Volume 2016, Article ID 6052181.
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內文：

Peutz-Jeghers syndrome:

A rare inherited autosomal dominant disease with an incidence of 1 in 12–30,000 live births, characterized by mucocutaneous pigmentation and multiple hamartomatous polyps in the gastrointestinal tract. PJS patients have a marked increase of risk of developing cancer.

A germline mutation in STK11, a tumour suppressor gene localized to chromosome 19p13.2–13.3, is an underlying abnormality.

WHO diagnosis:

1. Three or more polyps, with histological features of PJS.
2. A family history of PJS with any number of polyps
3. A family history of PJS with characteristic mucocutaneous pigmentation.
4. Characteristic mucocutaneous pigmentation with any number of polyps

Case report

A 57-year-old Iranian female patient with a chief complaint of pigmentation in oral cavity for more than 15 years

PI

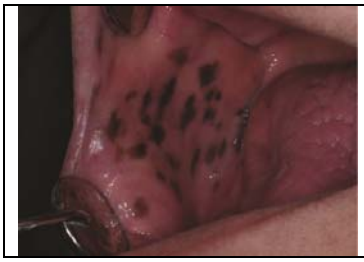
She was previously healthy and took no regular medications. She had no abdominal pain, nausea, vomiting, diarrhea, or history of changing in bowel habits or any significant loss of weight or appetite.

Family history

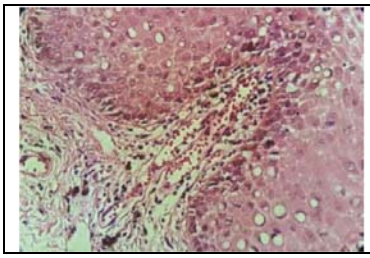
1. Her brother had PJS with perioral pigmentation and intestinal polyps compatible with Peutz-Jeghers polyp on histology and without any signs and symptoms of GI diseases
2. Her children do not have any sign or symptom of this syndrome

Objective finding

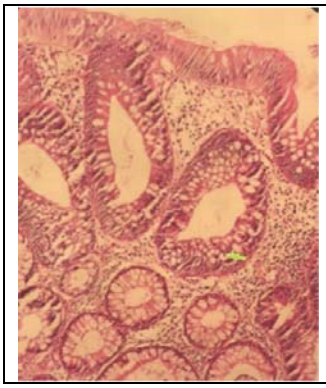
1.

	The patient had scattered dark brown to black macules, most of them measuring <0.5cm on the buccal and labial mucosa. These pigmentations were asymptomatic and well circumscribed and did not fade under pressure. There were not any similar macules on the tongue, lips, perioral skin, nostril, hands, and feet.

2.

	Biopsy of pigmentation on buccal mucosa was performed. Histopathologically there is evidence of increased basilar melanin with melanin incontinence into the submucosa. The patient with primary diagnosis of PJS was referred to gastroenterologists for more diagnostic assessments.

3. Colonoscopy: there was only one polyp in the rectum measuring 0.5 cm in diameter, without any features of invasions or intussusceptions
4. In the past 7 years
   (1) Did not observe any significant change in oral pigmentations
   (2) Did not have any sign or symptom of GI diseases except mild constipation, but endoscopy of GI showed the increase of the number of polyps every year.
   (3) In the last colonoscopy there were more than 100 polyps with the size of <1cm in the large bowel and rectum; no features of invasion and intussusceptions were noticed
5.

	Pathological examination of the polyps confirmed a denomatous and hamartomatous polyps (tubular type), with low grade dysplasia (H&E staining).

Assessment:
Peutz-Jeghers syndrome
Treatment plan:
In the case we reported that our patient even with diffuse gastrointestinal polyposis does not have any complaint of abdomen pain, bloody stool, or other symptoms. So no particular medication or surgery was administrated while in periodic follow-up.
Discussion
The presented case in this report was of particular interest since there are few reported cases of PJS with long-term follow-up. We were able to visit the patient annually and trace her medical findings. Another important issue in this report was a different course of the disease. Her igmentation showed little change but there was adramatic increase in the development of colorectal polyps over 7 years.

General	Our case
Male-to-female ratio of PJS is 1:1	Female
Average age at the time of diagnosis is 23 years in men and 26 years in women	57
Pigmentations of PJS are located around of mouth cheek, nostril, periorbital, ears, and overall orifices of body	Located intraorally in buccal and labial mucosa
Hamartomatous polyps commonly involve the jejunum and patients usually have complained of GI diseases like abdominal pain, diarrhea or bleeding, and bowel obstruction including intussusception	Colon was the most common site of these polyps and she did not have any sign and symptom of GI disturbance even after 7 years
The treatment for mucocutaneous pigmentations of PJS was usually considered unnecessary; but in some patients, the appearance of brown discoloration, especially on the skin, can	The pigmentations of our patient were intraoral and the face skin was not involved so we did not treat them.

be bother some, so laser therapy can be a choice treatment	

Mozaffar et al. reported a PJS without mucocutaneous pigmentation. So PJS may have a variable spectrum of manifestations in mucocutaneous pigmentation. The probable explanation is novel mutations in contributing genes

Matini reported three cases of PJS without clinical symptoms like abdominal pain and acute or chronic rectal blood loss and with no evidence of gastrointestinal or extragastrointestinal malignancy during 14 years of follow-up

The opinions for treatment of polyps:

1. Some consider these polyps as premalignant lesions , which should be paid considerable attention and removed by surgery or endoscopy
2. Others found out that rate of cancer in these polyps is very low , so periodic endoscopic surveillance is enough

Conclusion

1. Clinical manifestations of PJS maybe varied and the clinical course of this disease is not predictable
2. It is recommended that any patient presenting with multiple pigmentation or incomprehensible discoloration in oral cavity should be investigated for systemic diseases like Peutz-Jeghers syndrome

題號	題目
1	關於 Peutz-Jeghers syndrome 下列何者錯誤? (A) Mutation of the STK11 gene which encodes for a serine/threonine kinase, is responsible for most cases of Peutz-Jeghers syndrome (B) The intestinal polyps usually affect jejunum and ileum (C) The skin lesions of Peutz-Jeghers syndrome usually develop late in adult (D) the melanin pigment appears to be retained in the melanocytes rather than being transferred to adjacent keratinocytes
答案(C)	出處：Oral and Maxillofacial Pathology third edition P753
題號	題目
2	下列何者較不常見 recurrent aphthous ulceration ? (A) Behçet's syndrome (B) HIV (C) Peutz-Jeghers syndrome (D) MAGIC syndrome (mouth and genital ulcers and Interstitial chondritis)
答案(C)	出處：Oral and Maxillofacial Pathology third edition p332