Peutz-Jeghers syndrome: intriguing suggestion of gastrointestinal cancer prevention from surveillance.
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Source
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Abstract

BACKGROUND:
Peutz-Jeghers syndrome is characterized by GI polyps and mucocutaneous pigmentation and carries an increased risk of GI cancer. GI polyps may bleed or cause intussusception. Luminal GI surveillance is recommended, but there are few data detailing outcomes from GI surveillance in Peutz-Jeghers syndrome.

OBJECTIVE:
This study aimed to assess outcomes from GI surveillance in patients with Peutz-Jeghers syndrome.

DESIGN:
This study is a retrospective review, using hospital and registry notes and endoscopy and histology reports.

SETTING:
The investigation was conducted at a tertiary referral center.

PATIENTS:
All patients with Peutz-Jeghers syndrome who were followed up at St Mark's hospital were included.
MAIN OUTCOME MEASURES:
The primary outcomes measured were surveillance procedures performed, complications, and long-term outcomes.

RESULTS:
Sixty-three patients from 48 pedigrees were included; the median age when patients were first seen was 20 years (range, 3-59). Only baseline investigations were performed in 12 patients. The remaining patients were followed up for 683 patient years, a median of 10 years (range, 2-41). Seven hundred seventy-six procedures were performed to assess the GI tract. These led to 5 double-balloon enteroscopies, 1 push enteroscopy, and 71 surgical procedures. Of the surgical procedures, 20 were performed as a result of baseline investigations, 12 arose from investigations of symptoms, and 39 were due to surveillance of asymptomatic patients. No emergency surgical interventions were performed. No luminal GI cancers were diagnosed. Of the 2461 polypectomies performed, 6 polyps contained atypia or dysplasia. Six complications arose from endoscopy or surgical intervention, requiring 5 laparotomies to manage these complications.

CONCLUSION:
GI surveillance in Peutz-Jeghers syndrome is relatively safe and avoids the need for emergency surgery for small-bowel polyps. The lack of GI cancers may reflect that surveillance and polypectomy have prevented cancer from developing, although the detection of neoplasia or dysplasia is uncommon.

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