PEUTZ - JEGHERS SENDROMLU BİR HASTADA İNCE BARSAK TIKANMASI

SMALL BOWEL OBSTRUCTION IN A PATIENT WITH PEUTZ - JEGHERS SYNDROME

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Özet

Peutz-Jeghers sendromu gastrointestinal sistemde polipler, ağız etrafında ve vucudun diğer yerlerinde pigmentasyonlarla karakterize nadir görülen bir hastalıktır. Hastalarda en çok görülen semptom, intestinal obstrüksiyon nedeni ile oluşan karın ağrılarıdır. Obstrüksiyona ise polipler veya invajinasyon neden olur. Sık görülmemesi nedeni ile ince barsak obstrüksiyonu gelişmiş Peutz-Jegher sendromlu bir hastayı rapor etmeyi uygun gördük.

Anahtar kelimeler: *Peutz-Jegher sendromu, Polipozis*

Summary

Peutz-Jeghers syndrome is a rare disease characterizated by polyps of the gastrointestinal tract and pigmentation of the mouth and other parts of the body. In the patients the most common symptom is abdominal pain often caused by intestinal obstruction. The obstruction is usually due to a polyp or intussesception. Because of its rareness we find it interesting to report a patient with small bowel obstruction with Peutz-Jeghers syndrome.

Key words: *Peutz-Jeghers syndrome, Polypozis*

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Introduction

Peutz-Jeghers (P-J) syndrome is characterizated by mucocutaneous melanin hyperpigmentation and hamartomatous polyps found throughout the upper and lower gastrointestinal tract. This inheritable form of polyposis is transmitted by outosomal dominant, Mendelian genetics(3-5). The polyps are found most frequently in the small bowel, particularly in the jejenum, but they also may occur in the stomach, colon and rectum. The major morbidity results from intussesception, obstruction and bleeding(3).

Case report

A 15- year- old female was presented to General Surgery Department of Atatürk University Medical Faculty with the complaints of abdominal pain and distention. The pain was upper abdomen in location, crampy in character. She didn't have any previous medical problems and was on no medication. A palpable mass was found on phisycal examination. This mass was extending from right hypocondrium to hypogastrium in palpation. There was a polyp in anal canal on rectal examination. There was cutaneous pigmentations around the lips and buccal mucosa (Figure-1). Screening laboratory tests including complete blood count and urinanalysis were normal. There were air - fluid levels at plain abdominal x - ray film. Abdominal ultrasonography was performed. There were dilatations at jejenum and ileum segments. Free fluid was seen in the abdominal cavity. There was a big mass 15x15 cm. in dimension extending from hypocondrium to epigastrium. Sonographically the mass was reported as an intussesception. The patient was prepared for an emergency operation with the diagnosis of mechanical intestinal obstruction and operated on. There found to be a jejunojejunal intussesception. After desinvagination, gangren was determined in this segment which was resected. The cause of intussesception was a polyp. There were a lot of polyps 1-1,5 cm in size could be palpated in jejenum, ileum and colon. There weren't any palpable polyp in the stomach and duedonum. Jejuno-jejunostomy was performed and the operation was complated. Histologic examination revealed hamartomatous polyps. The patient was externed twelve days after operation. Endoscopic, ultrasonographic, tomographic and mamographic follow up program was made up the patient and she was called for control one month later. The patient's family invited for endoscopic research. One month later, the patient came for control. Colonoscopy and endoscopic polypectomy was performed to patient in our department (Figure2).

Discussion

In P-J syndrome, the most common and one of the most difficult to manage symptom is abdominal pain, often due to intestinal obstruction. The obstruction is usually due to a polyp or intussesception(3). There was abdominal pain and palpable mass in the abdomen in our case. The pain was crampy in character. The patient was suffering

Figure 1. *Cutaneous Pigmentations Around the Lips.*

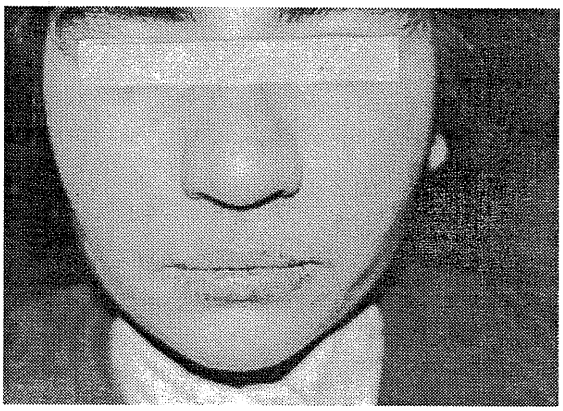
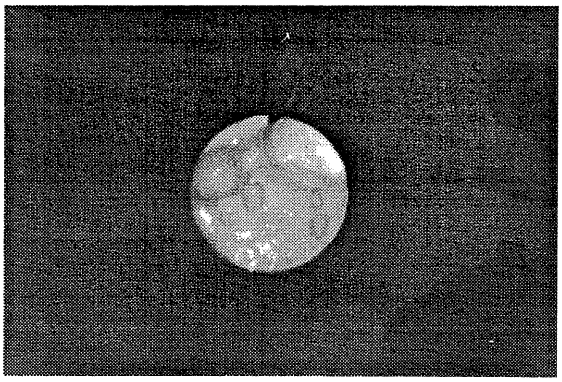


Figure 2. *Polyps Were Demonstrated By Colonoscopy in the Colon*



from nausea and vomiting. There was a jejuno-jejunal intussesception caused by a polyp in our patient. The classical mucosal pigmentation occurs primarily on the face and upper body, but may also be seen occure on any mucosal surface. Cutaneous pigmentation usually is noted at the birth or infancy; the skin changes may actually disappear after puberty. They consist of clusters of black or dark brown frecklelike spots from 1 to 2 mm. in diameter on and around the lips, buccal mucosa, fingers and toes (3,4). There were cutaneous pigmentations around the lips and buccal mucosa in our case. The fingers and toes were normal. P - J syndrome has been reconized as a benign hamartomatous disorder. However, it recently has been reported in several articles that gastrointestinal or extragastrointestinal malignancies were found to occure in patients with P - J syndrome who were being followed up for long periods of time (4,6). Spigelman at al (5) reviewed 72 individuals and found malignant tumors to have developed in 16 (22 %) with only one survival. They concluded that there is evidence to suggest a hamartoma - carcinoma sequence in this sendrome and suggested that the gene locus may relevant to the development of the malignancy in general. Hizova at al (7) rewieved the clinical courses of eight patient with P-J syndrome who had been followed up for as long as 12 years. Four cases of malignant neoplasm among the eight patient were found (adenocarcinoma cervix of the uterus, gastric cancer, duedonal cancer and pancreatic cancer). It should be realized that cancer may have occured or that they may developed in the near feature, even if young patients, once diagnosis of P - J syndrome is made. Our patient is 15 - year - old. There was no evidence of breast or gynecologic abnormalities. There weren't any pancreatic or gynecologic pathology at abdominal ultrasonography that performed before operation. The findings were justified in the operation. A follow up program was made up for the patient with ultrasonography, computerized tomography, endoscopy and mamography. She was called for the control one month later. We invited the family members for endoscopic research. The authors experience confirms veritable malignant potency in P - J syndrome and suggest that an intensive follow up gastrointestinal and extragastrointestinal sites are needed in patients with this syndrome (2). Additional intensive and systemic evaluations may be needed in patients with P - J syndrome, especially in young patients with symptomatic disease.

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