The Turkish Journal of Pediatrics 28:249 - 254, 1986

A CASE OF NEONATAL GASTRIC HAMARTOMATOUS POLYP*

*Türkan Patıroğlu MD***, Alev Hasanoğlu MD***,
Tahir E. Patıroğlu MD****, Yücel Arıtaş MD*****

Key words: gastric polyp, hamartoma, neonatal

Isolated polyps of the gastrointestinal tract are most frequently seen in the colon. Rarely, the patients have one of the genetic gastrointestinal polyposis syndromes, such as the Gardner syndrome, familial adenomatous colonic polyposis, Turcot syndrome, Peutz-Jeghers syndrome, generalized juvenile polyposis and Cronkhite-Canada syndrome. In evaluating persons with multiple gastrointestinal polyposis, the most important information for purposes of differential diagnosis is the number and distribution of the polyps within the gastrointestinal tract, the pathological features of the polyps and the nature and distribution of any extraintestinal manifestations[1]. The pathological nature of the familial polyps are either hamartomatous or adenomatous[1,2]. Hamartomatous polyps consist of epithelial elements and smooth muscle cells. The best model of the hamartomatous polyps is with the Peutz-Jeghers syndrome[3]. In this disorder, the presence of an autosomal dominant gene causes both gastrointestinal polyposis and abnormal pigmentation. Patients are usually in their middle 20's when the diagnosis is made[1-7]. To date the youngest published case of a hamartomatous polyp is of a 14-week-old male infant[6]. In this paper, we present the case of a neonatal gastric hamartomatous polyp.

Case Report

A six-week-old male infant was admitted to the Pediatric Department of Erciyes University Medical Faculty with the complaints of vomiting and weight loss. It was learned that the vomiting had been going on for three weeks.

On physical examination the patient's weight was found to be 2750 g, height 55 cm, head circumference 35 cm and thorax circumference 33 cm. Extreme loss of

* From the Departments of Pediatrics, Pathology and General Surgery, Erciyes University Medical Faculty, Kayseri.

** Lecturer in Pediatrics, Erciyes University Medical Faculty.

*** Associate Professor of Pediatrics, Erciyes University Medical Faculty.

**** Associate Professor of Pathology, Erciyes University Medical Faculty.

***** Associate Professor of General Surgery, Erciyes University Medical Faculty.

subcutaneous adipose tissue was observed. There was epigastric distention and a solid mass 2 x 2 cm in diameter. Other findings in the physical examination were unremarkable.

Laboratory findings: hemoglobin was 10.40 g/dl, the white blood count 13800/mm^3. Erythrocytes appeared hypochremic and normocytic in the peripheral blood smear. Urine analysis and blood sugar, electrolytes and blood urea nitrogen levels were within normal limits. A filling defect in the antral stomach was demonstrated on the X-ray studies (Figure 1).

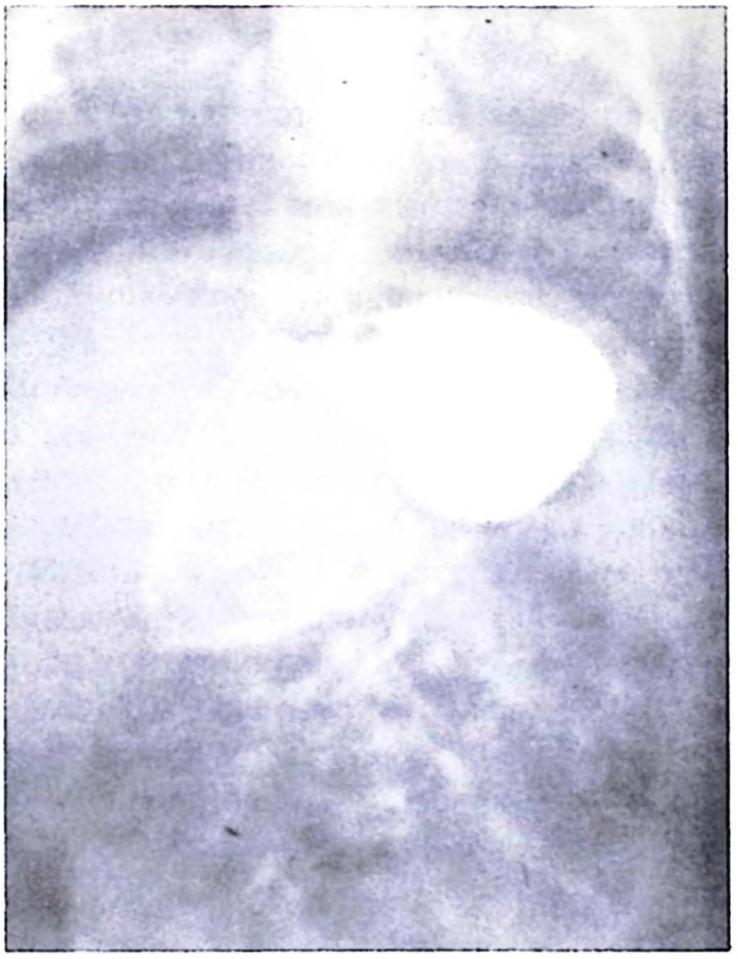

Figure 1

Filling defect in antral stomach on the stomach X-ray

The patient was operated on and a cecil polyp 4 x 5 cm in diameter in the prepyloric region was excised, and pyloroplasty and gastrostomy were performed. Cephazolin, gentamycin and cimetidine were administered in the postoperative period, when the course was complicated by severe and persistent hypoglycemia, electrolyte imbalance and septicemia. The patient died 30 days after the operation.

Pathological examination: the broad-based antral mass was covered with mucosa and measured 4.5 x 4 x 3 cm. On gross observation of the sections, there were mucoid areas and hemorrhage. On microscopic examination the surface mucosa

TABLE I

The Inherited Gastrointestinal Polyposis Syndromes

Syndrome	Gastrointestinal lesions	Pathological nature of polyps	Extraintestinal lesions	Predisposition to cancer
Familial adenomatous colonic polyposis	Polyposis limited to colon and rectum	Adenomatous	None	Marked
Gardner syndrome	Polyps of colon and rectum (rarely small bowel)	Adenomatous	Osseous and soft tissue tumors	Marked
Turcot syndrome	Colonic polyps	Adenomatous	Tumors of brain	Not reported
Generalized juvenile polyposis	Usually colon and rectum, but may involve small bowel and stomach	Hamartomatous	None	Probable
Peutz-Jeghers syndrome	Generalized polyposis, but small bowel is consistent	Hamartomatous	Mucocutaneous pigmentation	About 2-3%
Cronkhite-Canada syndrome	Generalized polyposis	Hamartomatous	Alopecia, cutaneous hyperpigmentation and nail distrophy	Not reported

was seen to be ulcerated and under this there were many dilated glandular structures. These were lined with mucous cells some of which were hyperplastic. Bundles of smooth muscle extended from the muscularis mucosa from the base into the upper part of the polyp and between the glands. With these findings the diagnosis of "hamartomatous polyp" was made (Figure 2).

Postmortem examination: bronchopneumonic infiltration was the only other positive finding. No other polypoid structures were to be found in the gastrointestinal tract either macroscopically or microscopically.

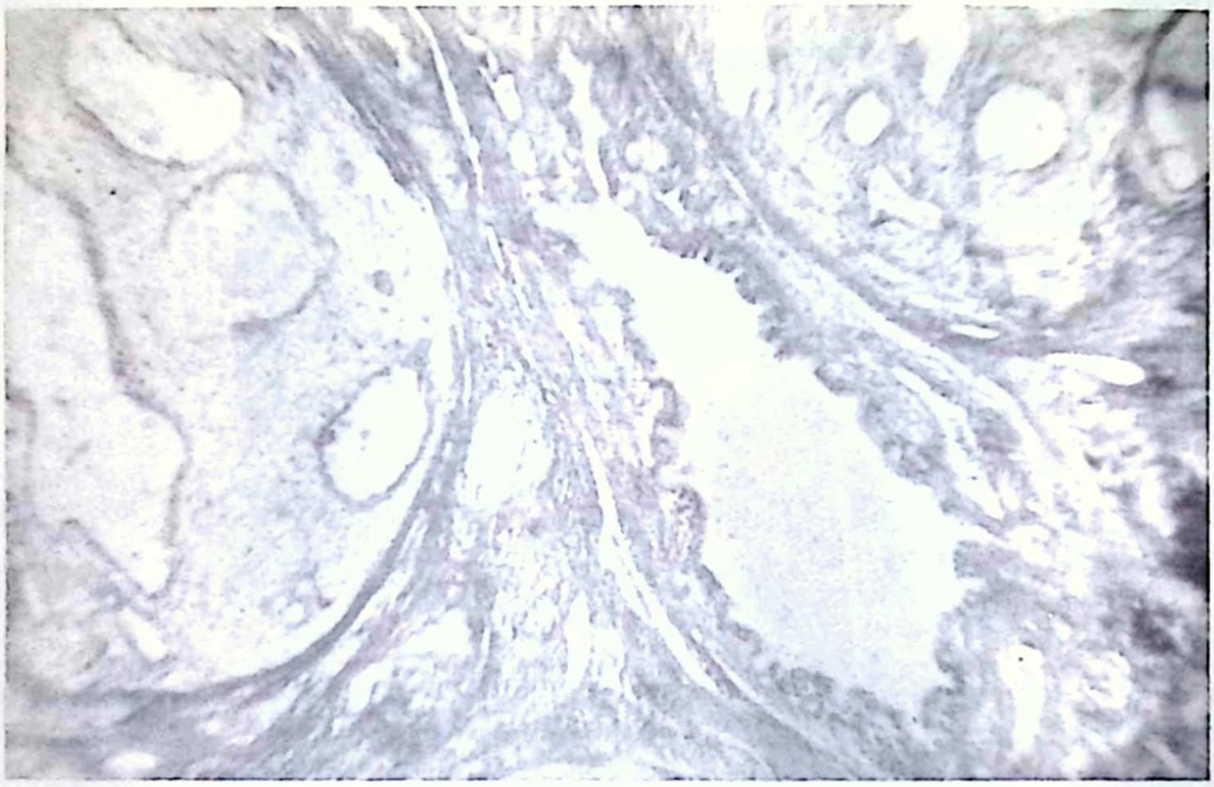

Figure 2

Smooth muscle bundles among glands; some of them were cystic in appearance (HE x 63).

Discussion

Isolated gastrointestinal polyps in childhood, with the exception of the familial polyposis syndromes, are not very significant. These familial polyposis syndromes are summarized in Table I[1,2,6,8-10]. Microscopical evidence in our patient was compatible with the Peutz-Jeghers syndrome one of the familial polyposis syndromes.

This syndrome was defined by Peutz for the first time in 1921, followed by Jeghers in 1929; it is inherited as autosomal dominant[5]. The polyposis is generalized. In a series of 182 cases, involvement of the gastrointestinal tract was as follows: 96% small bowel, 29% colon, 30% rectum, 24% stomach[3]. Polyps may be pedinculated or cecil[3,11]. The large polyps cause intussusception and obstruction[1,11]. Rectal bleeding and recurrent colicky abdominal pain is present in

many patients[1,11]. Hematemesis may occur with gastric or duodenal polyps and many patients are anemic[1]. Bundles of smooth muscle extending from the muscularis mucosa at the base of the polyp into the upper part of the polyp and between the glands are histological characteristics of hamartomatous polyps[3].

The extraintestinal manifestation is abnormal pigmentation that usually appears in infancy or early childhood. These melanotic macules look like freckles but are unusual because of their location on the lips, the buccal mucosa, the eyelids, the digits and more rarely the intestinal mucosa[1,2,7]. The Peutz-Jeghers syndrome usually becomes evident in the second decade; but the case of a 14-week-old infant with a hamartomatous polyp in the rectosigmoid region who developed findings of pubertas precox at two years of age has been reported[6]. Five percent of patients with Peutz-Jeghers polyposis have no abnormal pigmentation and 5% of patients have such pigmentation without polyposis[1]. Abnormal pigmentation may become evident later; indeed the case has been reported of a neonate with abdominal pain and vomiting who developed the typical pigmentation five years later[7]. Burdiek followed up a family with the Peutz-Jeghers syndrome for 27 years; the six and eight-year-old children had abnormal pigmentation but no polyps, but there were polyps in the two-year-old child of the same family[4].

Our patient had no familial history and no abnormal pigmentation. On postmortem examination no polyps were found in other parts of the gastrointestinal tract. Although there was no abnormal pigmentation we think that the gastric polyp in our patient, which was of the hamartomatous type, was a condition of the Peutz-Jeghers syndrome, and that abnormal pigmentation would have developed later if the patient had lived.

Finally, differential diagnosis in cases of gastrointestinal polyps in childhood is very important. If polyps are hamartomatous in nature extraintestinal system findings are useful in differential diagnosis.

Summary

Isolated gastrointestinal tract polyps are frequently seen in the colon. Occasionally the patients have one of the genetic gastrointestinal polyposis syndromes such as the Gardner syndrome, familial adenomatous colonic polyposis, Turcot syndrome, Peutz-Jeghers syndrome, generalized juvenile polyposis or the Cronkhite-Canada syndrome. Pathologically, the familial polyps is either hamartomatous or adenomatous. To date the youngest published case of a hamartomatous polyp is that of a 14-week-old infant. In this paper, we present a case of neonatal gastric hamartomatous polyp, not accompanied by any abnormal pigmentation of the mucosa.

REFERENCES

1. Erbe RW. Inherited gastrointestinal polyposis syndromes. *N Engl J Med* 294:1101, 1976.
2. Burke V, Anderson MC. Other disorders of the large intestine. In Anderson CM, Burke V (eds). *Pediatric Gastroenterology.* London: Blackwell Scientific Publications, 1975.
3. Ming S. *Tumors of the Esophagus and Stomach.* AFIP, Washington DC 20306, 1973, pp 139-140.
4. Burdiek D. Peutz-Jeghers syndrome. A clinicopathologic study of a large family with 27-year follow-up. *Cancer* 15:2139, 1982.
5. Jeghers H, McKusick VA, Katz KH. Generalized intestinal polyposis and melanin spots of the oral mucosa, lips and digits. A syndrome of diagnostic significance. *N Engl J Med* 241:993, 1949.
6. Lamesch AJ. An unusual hamartomatous malformation of the rectosigmoid presenting as an irreducible rectal prolapse and necessitating rectosigmoid resection in a 14-week-old infant. *Dis Colon Rectum* 26:452, 1983.
7. Rosenack U, Caspary W. Das Peutz-Jeghers Syndrom. *Dtsch Med Wochenschr* 108:389, 1983.
8. Gardner EJ. Follow-up study of a family group exhibiting dominant inheritance for a syndrome including intestinal polyps, osteomas, fibromas and epidermal cysts. *Am J Hum Genet* 14:376, 1962.
9. Turcot J, Despries JP, St Pierre F. Malignant tumours of the central nervous system associated with familial polyposis of the colon: report of two cases. *Dis Colon Rectum* 2:465, 1959.
10. Cronkhite LW, Canada WJ. Generalized intestinal polyposis: an unusual syndrome of polyposis, pigmentation, alopecia and enychetrophia. *N Engl J Med* 252:1011, 1955.
11. Weed DA. *Tumors of the Intestines.* AFIP, Washington DC 20305, 1967, p 26.