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Intestinal Hemangioma Causing Episodic Bleeding and Iron Deficiency Anemia

Sung Hye Park, Jin Suk Suh, Je G. Chi,
Jeong Kee Seo* and Kwi Won Park**

Departments of Pathology, Pediatrics and Pediatric Surgery***
Seoul National University Children's Hospital, and
College of Medicine, Seoul National University
Seoul 110-744, Korea

= Abstract =Hemangiomas of the intestine are extremely rare. The clinical manifestations are intestinal obstruction, hemorrhage, pain and/or intussusception. Overt blood loss is the most common presenting symptom in children.

Presented case is a 6 year old girl with a jejunal hemangioma who has suffered from chronic iron deficiency amemia during 4 years due to recurrent gastrointestinal bleeding. Melena was not easily noticed because her black stool color was thought to be due to varidus syrup (iron preparation). Unil the 4th admission, all workups except angiography failed to disclose the source of bleeding. Angiography showed contrast puddling in late venous phase at the area of the jejunoileal junction. Exploratory laparotomy was carried out to reveal a reddish discoloration in a segment of small intestine, 170 cm proximal from the ileocecal valve, and cavernous hemangioma was confirmed.

Key words: ***Hemangioma, Intestine, Anemia, GI bleeding***

INTRODUCTION

Hemangiomas of the intestinal tract are extremely rare, but have been observed in all age groups (Nader and Margolin, 1966). The individual clinician or pathologist is likely to see only a single case in their lifetime, and this may account for a reluctance to report them (Mellish, 1971). However, these simple lesions are important because of variable clinical manifestations and diagnostic difficulty unless phleboliths are present. The manifestations are those of intestinal obstruction, hemorrhage, pain or intussusception; only rarely is there a plapable mass (Nader and Margolin, 1966 and Hamilton, 1987). They are sometimes multiple and may be associated with hemangiomas elsewhere. Though it is often fatal due to massive bleeding, it is practically always curable (Marine and Lattomus, 1958 and Hyun *et al.*, 1969). Its preoperative recognition is therefore of importance.

We have experienced recently a case of cavernous hemangioma of the jejunum in a 6 year old girl. Severe chronic hypochromic microcytic anemia with occult blood loss was the presenting sign. Resection of hemangioma at laparotomy resulted in cure of this condition. The purpose of this article is to report a case dealing with this unusual condition and to stress the importance of recognizing this entity as one of the causes of gastrointestinal bleeding in childhood.

CASE REPORT

A 6 year old girl was admitted to Seoul National University Children's Hospital (SNUCH) on April 12, 1989 for further evaluation of known

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iron deficiency anemia.

This patient was born to a 31 year old mother via normal fullterm spontaneous delivery. The birth weight was 4.0 kg. No contributory family history was elicited. She had been healthy until 2 years of her age when developed sudden pallor and lethargy. She was admitted to a hospital on July 1985 for the first time where the diagnosis of iron deficiency anemia was made. Hemoglobin at that time was 4.7 gm/dl. Transfusion and varidus syrup were prescribed, and the pallor improved over 3 months afterwards. On September 1986, she became pallid again and anorexic. she was brought to SNUCH Pediatric Outpatient Clinic and was placed on varidus syrup to improve her hemoglobin level from 4.7 gm/dl to 7.8 gm/dl.

The second admission was February 22, 1988 for diagnostic endoscopy under general anesthesia. Two days prior to admission, she had became suddenly pale and lethargic. Blood pressure was 90/60 mmHg, pulse 140/min, and respiration 30/min. Physical examination revealed a pale lethargic girl who showed essentially normal physical findings except pale conjunctiva. The hemoglobin was 5.6 gm/dl, hematocrit 16.5 %, WBC 8000/mm 3 (poly 38 %, lymph 48 %, mono 12 %, eosino 2 %). Platelet 276 K/mm 3 and reticulocyte 15.0 %. Stool ocult blood was positive. Upper GI series revealed prominent rugal pattern of gastric mucosa and jejunum along with flocculation of barium in proximal jejunum. No evidence of ulcer or intrinsic lesion was seen. Colon study and Meckel's scan were normal. Endoscopy showed no abnormality. Packed red cells were given and the patient was discharged after 11 days hospitalization. The bleeding source was not recognized this admission.

Her therd admission November 14 to 23, 1988 was due to palpitation and sudden aggravation of pallor. All studies were repeated, but the etiology of the iron deficiency anemia was not clarified. On March 19, 1989, she was brought to emergency room because of dizziness.

On April 12, 1989, she admitted a 4th time. On this admission, hemoglobin was 6.6 gm/dl, hematocrit 22.2 %, WBC 7500/mm 3, platelet 317K/mm 3, MCH 23.9 pg, MCV 80.,4 fl and MCHC 29.79 gm/dl. Melena was noticed to the clinician, but was not thought significant because her black stool color was thought to be due to varidus syrup (iron regimen). Angiography done on April 17, showed contrast puddling in jejunoileal junction (Fig. 1). On April 19, 1989 exploratory laparotomy was done and revealed a reddish discoloration in a segment of small intestine, 170 cm proximal from the ileocecal valve.

Pathological examination: Grossly, the serosa of the involved portion showed purplish disoloration. the lumen was partly narrowed by protruding mass, measuring 3 × 5 cm. On section the wall of the jejunum was thickened by sponge-like mass containing fresh blood and was involving the entire thickness of the bowel wall (Fig. 2). Microscopically there were numerous dilated vascular channels that share the common wall and lined by a single layer of endothelial cells (Fig. 3, 4).

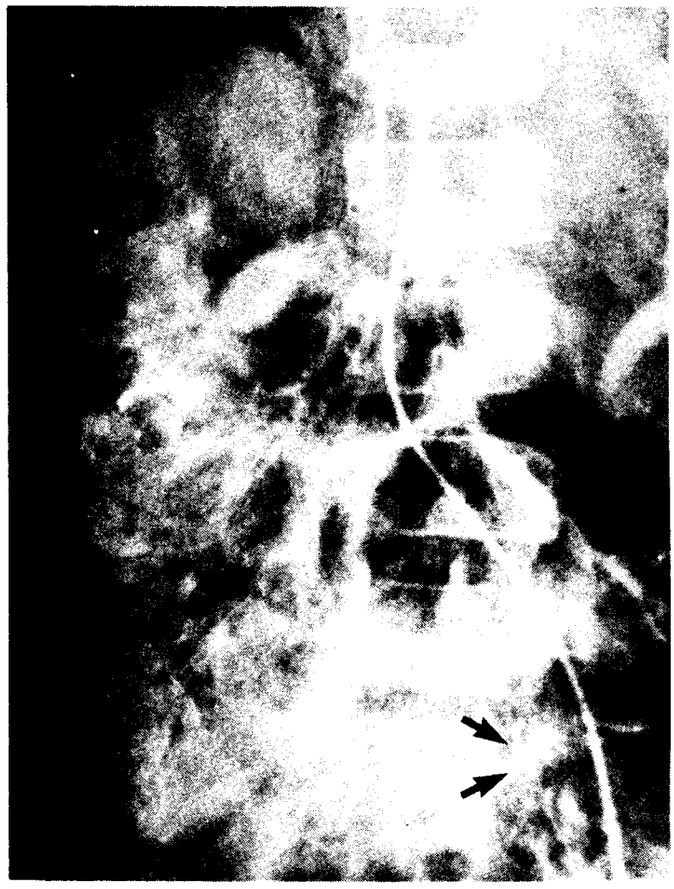

Fig. 1. The angiography shows puddlings sign (arrows) in a segment of small intestine, 170 cm proximal from the ileocecal valve.

Fig. 2. Microscopic finding shows cavernous hemangioma involving the intestinal mucosa. (H & E, ×100)

DISCUSSION

Hemangioma occurring in the gastrointestinal tract is not often considered in connection with gastrointestinal bleeding of unknown etiology (Brayton, 1964). The intestinal hemangiomas may be classified into 4 categories (Rissier, 1960) 1. Multiple phlebactasis, 2. Cavernous hemangiomas, 3. Hemangioma simplex, and 4. angiomatosis with gastrointestinal localization, representing widespread vascular dysplasia, such as Osler-Rendu-Weber disease (Bruusgaard, 1974). Among these, cavernous hemangiomas are probably the commonest type and consist of dilated venules and venous sinuses which may replace the intestinal wall from mucosa to serosa. They are frequently found in the lage intestine (rectum and sigmoid) and cause either hemorrhage or obstruction. The polypoid cavernous type are similar in structure but with a polypoid configuration.

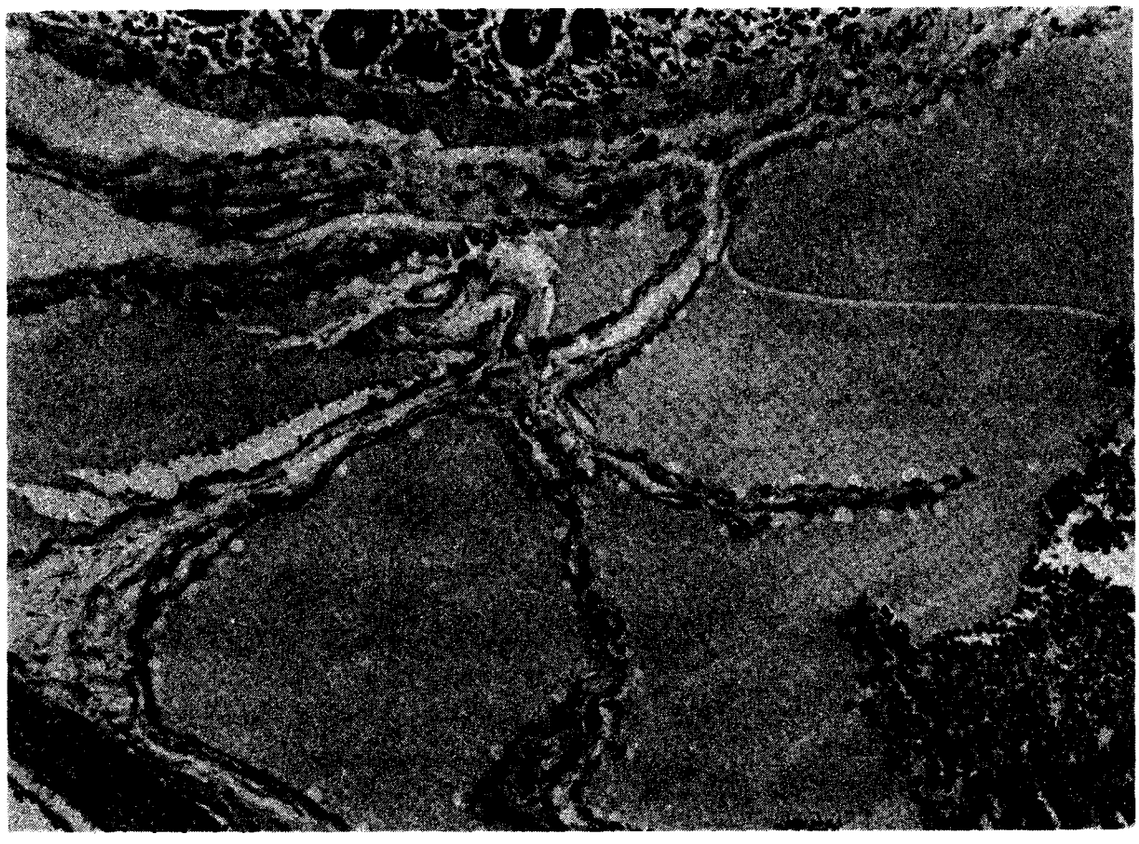

Fig. 3. Cavernous hemangioma involving submucosa. The tumor mass consists of interconnecting vascular channels showing common wall and lined by a single layer of endothelium. The lumen contains protein-rich plasma. (H & E, × 100)

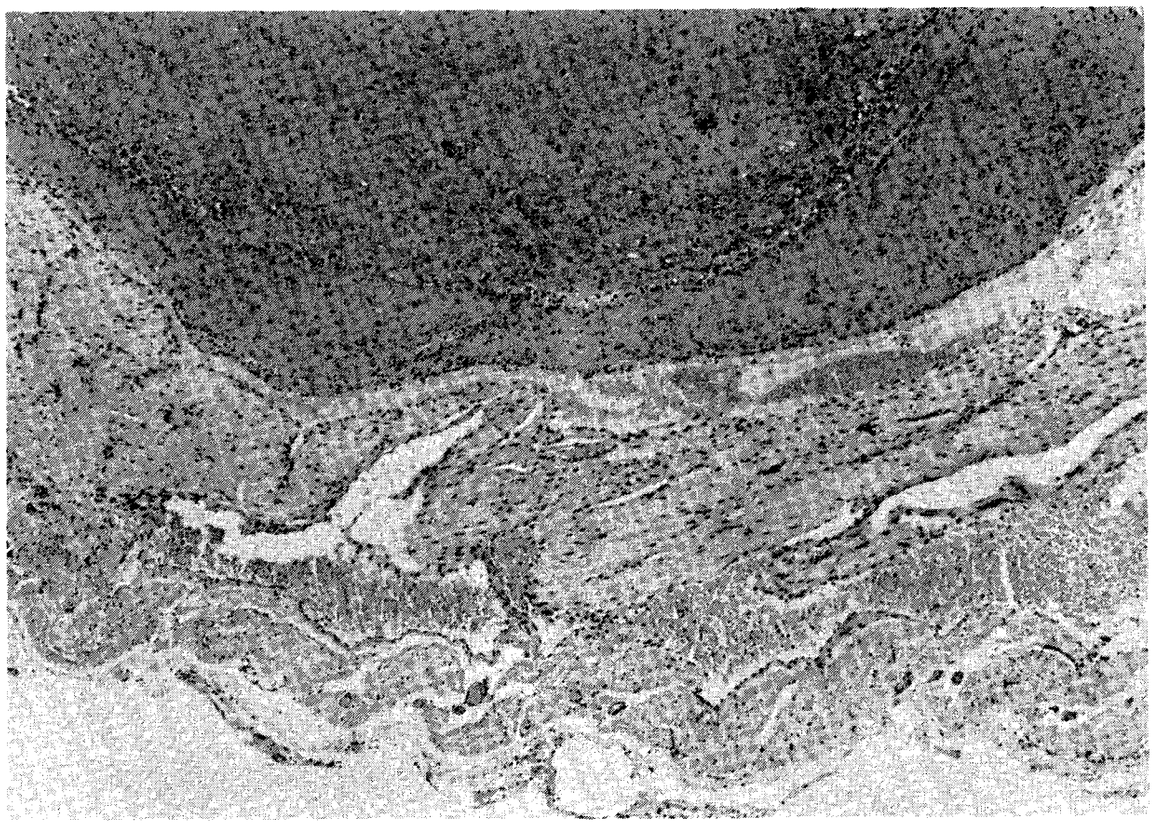

Fig. 4. Cavernous hemangioma involving muscularis propria and subserosa. The upper half shows an organizing thrombus in the submucosa. (H & E, × 40)

The cardinal complications of intestinal hemangiomas are gastrointestinal bleeding and intestinal obstruction due to intussusception. (Weinstein *et al.*, 1963, Hyun *et al.*, 1969). Nader and Margolin collected a total of 47 cases of intestinal hemangiomas causing gastrointestinal symptoms in the pediatric age group from the English literature. Thirty-four of these had gastrointestinal bleeding as a presenting symptom, whereas obstruction or intussusception was found in six, dyspepsia in one and "severe anemia" due to chronic occult blood loss as in this case under discussion in three, and asymptomatic intestinal hemangiomas were found in the large intestine in 26 cases and in the small bowel in 21 instances. Among small bowel lesions, 15 had intestinal bleeding, 6 had intestinal obstruction and 1 had both bleeding and obstruction. Hemangiomas of the skin were found in 22 of the 40 cases in the series.

The characteristic radiologic findings of multiple, relatively smooth-bordered intramural extramucosal filling defects projecting into the bowel lumen, accompanied by phleboliths within the defects, virtually confirm a diagnosis of vascular malformation (Hellstrom *et al.*, 1955, Hollingsworth, 1951, and Marine and Lattomus, 1958). Among the reported cases, X-ray changes of filling defects or phleboliths or both were present. The occurrence of phleboliths seems to be more frequent in adults (Scott and Brand, 1957, Grieco and Bartone, 1967). It is interesting that our case did not reveal phleboliths and all workups except angiography failed to reveal the bleeding source. Delay of making diagnosis in our case was partly due to the fact that the melena history was masked by the prologned taking iron orally and the occult nature of the bleeding. Although intestinal hemangiomas are rare, it should be considered as a possible cause of gastrointestinal bleeding and intestinal obstruction. It is important that intestinal hemangioma should be included in the differential diagnosis of iron deficiency anemia due to chronic blood loss or acute gastrointestinal hemorrhage in childhood.

For treatment of intestinal hemangioma, drastic surgical means seem to offer the only possible cure. Sclerosing injections and radiation therapy have not been successuful in the management of these cases.

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= 국문초록 =

간헐성 장출혈과 철결핍 빈혈로 나타난 소장의 혈관종

서울대학교 의과대학 병리학교실, 소아과학교실* 및 외과학교실**

박성혜 · 서진석 · 지제근 · 서정기* · 박귀원**

장관의 혈관종은 대단히 드물다. 그러나 발생하는 경우에는 흔히 장폐쇄, 장출혈, 복통등으로 나타나기 때문에 의사들은 위와같이 원인모를 장증세가 있을때에는 장의 혈관종을 의심하여야 한다.

본에는 6세의 여아로서 3년여를 간헐성 장출혈에 의한 철결핍성 빈혈로 고생하다가 결국 개복수술 결과 공장의 해면성 혈관종이 발견되어 절제된 증례로서 다시한번 조기 진단의 중요성이 강조된 예이다. 혈관종은 그 크기가 3 × 5 cm로서 회맹장판에서 170 cm근위에 위치하였으며 부분적으로 내강돌출을 야기하면서 공장의 전층을 침범하고 있었다. 조직학적으로는 전형적 해면성 혈관종으로 주병변은 점막하에 있었으나 점막, 근층 및 장막에 동시에 분포하고 있었다.

환아는 절제술로 근치되었으며 기타 동반된 병변은 관찰되지 않았다.