Short reports

Intestinal haemorrhage in Turner’s syndrome

D M BURGE, A W MIDDLETON, R KAMATH, AND B J FASHER

Royal Alexandra Hospital for Children, Camperdown, New South Wales, Australia

Summary

A 13-year-old girl with Turner’s syndrome and bleeding from intestinal venous ectasia is reported. The various types of vascular anomaly of the bowel associated with Turner’s syndrome are discussed. Awareness of these anomalies may help prevent unnecessary laparotomy in children with this syndrome.

Gastrointestinal bleeding in patients with Turner’s syndrome was first reported by Lisser et al. Since then, about 30 cases have been described and it has been estimated that the incidence of gastrointestinal haemorrhage in Turner’s syndrome is about 7%. Three types of vascular anomaly of the gastrointestinal tract have been described—namely haemangioma, telangiectasis, and venous ectasia.

Case report

A 13-year-old girl was admitted to this hospital for investigation as she had been passing dark red blood rectally for 48 hours. There were no associated symptoms apart from some mild central abdominal discomfort. This was her fourth episode of gastrointestinal bleeding. The first episode had occurred at age 20 weeks and had lasted for 48 hours. No treatment was required on that occasion.

Further episodes of bleeding took place at 9 years and 12 years of age, each episode lasting for two days. On the third occasion, blood loss was more pronounced and required blood transfusion. At that time, gastroscopy, sigmoidoscopy, and a barium meal were carried out at a local hospital and were all normal.

On examination on this admission, she was noticed to be a small child and both her height and weight were below the 3rd centile for her age, her height age being 9 years and 2 months and her weight age being 10 years. She showed no signs of secondary sexual development but, apart from slightly wide spacing of her nipples, showed no features characteristic of Turner’s syndrome.

Anthropometric measurements showed an upper segment: lower segment ratio of 1·08.

She had no signs of haemorrhagic shock on admission, but her haemoglobin level was 10·4 g/dl and her blood film showed changes consistent with recent blood loss. She had mild central abdominal discomfort but no abnormal physical signs on abdominal examination. On rectal examination, dark blood was present in the rectum.

Further investigations included a pertechnetate scan of the abdomen with pentagastrin enhancement, which failed to show any evidence of ectopic gastric mucosa, and a small-bowel barium examination which was normal. As part of the investigation of her short stature, a blood sample was taken for chromosomal analysis.

Although she still continued to lose a small amount of blood during the next few days blood transfusion was not necessary. However, as this was her fourth intestinal bleed, an explorative laparotomy was performed. At operation, many prominent distended veins were noted on the serosal surface of the small and large-bowel in the distribution of the superior mesenteric vessels. These prominent serosal vessels appeared segmentally along the bowel arising at intervals from the mesentery, each dividing on the antimesenteric serosal surface to run longitudinally, proximally, and distally for about 30 cm.

There were intervening segments of about 10 cm of apparently normal bowel between each abnormal vein system. In addition to these main veins, smaller prominent vessels arose from the mesentery to run circumferentially around the serosa. Veins within the bowel mesentery appeared normal, as did the remainder of the portal venous system and the liver and spleen. In view of the nature of this venous abnormality, no resection was attempted. There was nothing to suggest the exact site of origin of the blood loss.

Examination of the pelvic contents at operation showed a small uterus and streak gonads, while the chromosomal analysis demonstrated a 45XO karyotype. No further gastrointestinal blood loss occurred and the child was discharged 2 weeks later.
Discussion

Three different descriptions have been applied to vascular lesions of the gastrointestinal tract in Turner's syndrome—namely haemangioma, telangiectasis, and venous ectasia. However, reviewing the way these lesions are described by various authors, the terminology seems to overlap greatly, and it may well be that there is one underlying abnormality—namely ectasia of capillaries, venules, and veins. Depending on the preponderance of these three findings, this abnormality may present as a variety of macroscopic appearances.

Diagnosis of this abnormality without resort to laparotomy may be difficult. Telangiectasis has been reported as visible via the sigmoidoscope but this is unlikely to be helpful in patients such as ours with macroscopically visible ectasia only in the distribution of the superior mesenteric vessels. Laparoscopy or culdoscopy would seem to be the most helpful investigation.

Most patients so far reported have each had their first haemorrhage before 20 years of age. In the 20 cases reported by Passarge most bleeds were self-limiting but 3 were life-threatening and 2 patients, notably both young children, died.

In paediatric practice, a cause for recurrent gastrointestinal blood loss can often be found without resort to surgery. However, explorative laparotomy is occasionally required. Awareness of the condition discussed here may help prevent unnecessary laparotomy in children with known or suspected Turner's syndrome. Having established the diagnosis, subsequent haemorrhages, which are likely to occur, can generally be managed conservatively.

References


Correspondence to D M Burge FRCS, Department of Paediatric Surgery, Royal Alexandra Hospital for Children, Camperdown 2050, Sydney, New South Wales, Australia.

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Synovial haemangioma presenting as monarticular arthritis of the knee

W L HAWLEY AND BARBARA M ANSELL

Juvenile Rheumatism Unit, Canadian Red Cross Memorial Hospital, Taplow

SUMMARY Two children with haemangioma of the synovial membrane presenting as swelling of a knee joint are described; in one patient this was associated with epiphyseal overgrowth. This condition should be considered if bloody synovial fluid is obtained and clotting studies are normal.

Disease in a single joint in a child requires full appraisal as quickly as possible. Since the decline in tuberculosis, persistent synovitis in one knee is often attributed to juvenile chronic arthritis. There are however a number of conditions, each requiring surgical treatment, which should be excluded. The purpose of this paper is to draw attention to synovial haemangioma.

Case reports

Case 1. A 14-month-old white boy developed sudden swelling of the right knee with no preceding trauma. He was admitted to a hospital where aspiration of the joint under general anaesthesia produced 24 ml of frank blood. A plaster cylinder was applied to the leg and left on for 3 weeks. When the plaster was removed the knee was still swollen and there was a 30° flexion deformity, so further investigations for evidence of infection, a bleeding tendency, or juvenile arthritis, were instituted. These included erythrocyte sedimentation rate (ESR), prothrombin and kaolin-cephalin clotting time (KCCT), antinuclear antibody, and slit-lamp examination of the eyes. All results were normal, as was the x-ray film.
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D M Burge, A W Middleton, R Kamath and B J Fasher

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