Vascular hamartomas of the gastrointestinal tract

Case reports

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Summary

Three patients with vascular hamartomas of the gastro-intestinal tract are described. These congenital lesions are distinct from hereditary telangiectasia and acquired lesions including vascular ectasia (angiodysplasia). The classification of these hamartomas is based on the capillary and reticular phases of development of the vascular tree, and the reticular forms are further subdivided into venous, arterial or arteriovenous fistulas, and haemolymphatic hamartomas. The typical features, diagnosis and management are discussed.

Between 1975 and 1980, 5 patients seen at Groote Schuur Hospital, Cape Town, with gastro-intestinal bleeding have been investigated by angiography and found to have a vascular anomaly which was then resected. One of these patients had Rendu-Osler-Weber syndrome and 4 had vascular hamartomas. One patient with a vascular hamartoma was the subject of a previous publication and the other 3 form the basis of this article.

Case reports

Case 1

A 56-year-old man who had had occasional bouts of abdominal colic in the past presented with colicky abdominal pain. He was passing red blood per rectum, and needed 5 units of blood for resuscitation. A barium meal and enema examination, gastroscopy and sigmoidoscopy were negative. Superior mesenteric angiography showed an increase in the number and tortuosity of the vessels supplying the proximal and mid-jejunum. A number of small vascular pools were seen in this area. Venous filling was normal.

At laparotomy the vascular malformation was not seen, and 55 cm of proximal jejunum was resected, as this corresponded with the segment localized angiographically. In addition a small tumour was palpated in the ileum, and this was resected with a small margin of bowel. The patient has since remained asymptomatic.

On microscopic examination the tumour in the ileum was found to be a leiomyoma which had undergone almost complete collagenization, and there was a vascular malformation of the jejunal submucosa comprised of veins and arteries. There were also two distinct nodules in the mesentery which were hamartomas of fat, blood vessels and lymphatics. The resection lines were not free of the submucosal lesion.

Case 2

A 60-year-old woman had presented 11 years previously passing fresh blood per rectum and was treated by haemorrhoidectomy. She remained well until 1979 when she had had three further episodes of bleeding, all requiring transfusion. Barium enema examination and colonoscopy were negative. Superior mesenteric angiography showed the number and tortuosity of vessels from the mid- to distal transverse colon to be increased. No evidence of early venous filling was seen, but venous drainage from that area was dense and prolonged. At surgery an obviously abnormal distal transverse colon was resected, with many large vessels coursing over the bowel and in the mesentery. On histological examination there was found to be a marked increase in the number of veins and arteries in the submucosa and serosa, in keeping with a vascular malformation.

Case 3

A 56-year-old woman presented at her local hospital with haematemesis and melena, needing 9 units of blood before a vagotomy and pyloroplasty was performed for 'gastritis'. Postoperatively the bleeding continued and she needed a further 8 units of blood to maintain her haemoglobin concentration. On transfer to Groote Schuur Hospital, gastroscopy failed to reveal a bleeding point and coeliac and mesenteric angiograms were normal. At laparotomy an obvious vascular malformation at the duodenojejunal flexure, consisting of a plexus of veins coursing over the bowel, was found and resected. Histological examination revealed large and small veins and arteries in a vascular malformation.

Discussion

The diagnosis of the source of gastro-intestinal haemorrhage is usually straightforward if haematemesis has occurred, in which case barium meal examination and gastroscopy will usually provide the diagnosis. With recurrent blood loss, where the clotting profile, barium studies and endoscopy fail to demonstrate a cause, angiography and isotope studies are needed to identify and localize a lesion before resorting to diagnostic laparotomy, which may also fail to demonstrate the lesion. Angiography may identify a lesion as a structural abnormality of the vessels, an arteriovenous shunt or extravasation of blood if active bleeding is fast enough. The vascular abnormalities can be divided into hereditary telangiectasia, congenital vascular
hamartoma or an acquired lesion, including vascular ectasia, with differing angiographic features and natural histories. Vascular hamartomas which have been documented have been superficial, and the head and neck are the commonest sites, followed by the limbs and the trunk. Of the deep forms, pulmonary and pelvic sites are commoner than the gastro-intestinal tract; the latter lesions are said to affect 1 in 14,000 people, to cause 1% of gastro-intestinal haemorrhages and to account for 0.3% of gastro-intestinal tumours and 3% of small-bowel tumours. Ten per cent are associated with extra-intestinal hamartomas, which may be visible and suggest a related cause for undiagnosed gastro-intestinal bleeding.

The classification of bowel vascular hamartomas has been enigmatic, with division into 'flat or polypoid' to distinguish those which are visible on barium examination or produce mechanical effects in the bowel from those which are not visible on barium examination and are unlikely to produce mechanical effects. With the advent of angiography, a more definitive classification is required. Malan, in discussing haemangiomata of the limbs, postulated that haemangiomata are related in structure to the phases of development of the vascular system. If the arrest occurs in the capillary phase, a capillary haemangioma results. Hamartomas resulting from arrest in the reticular phase were subclassified as venous, arterial or arteriovenous fistulas, or as haemolympathic (mixed), according to the structure of the constituent vessels. The arteriovenous fistulas have been further subclassified by Szilagyi et al. as 'microfistulous', with no bruit and no arteriovenous shunting demonstrable on angiography but biochemical evidence of shunting in the venous blood, and 'macrofistulous', which is a more obvious arteriovenous fistula. The amount of blood shunted and the local, regional and systemic sequelae will increase until the capacity of the fistula is reached by dilatation of the feeding vessels. The 3 cases presented represent the arterial, venous and haemolympathic forms.

Hamartomas, which have been present from birth, may remain clinically silent. Bleeding may occur from submucosal vessels at any age, and any shunting will depend on the degree of communication between the arteries and veins in the hamartoma. Rare physical signs are a mass and a bruit. Phleboliths may be seen on plain radiographs with a mass on barium examination and endoscopy may reveal abnormally prominent or numerous submucosal vessels. Radionuclide scanning with technetium-99m-labelled red blood cells or albumin has been used, but the upper jejunal area may be obscured by the renal uptake and accurate localization may be difficult. Angiography may reveal large or numerous arteries, a macrofistulous arteriovenous shunt with early venous filling, and/or dilated veins emptying early or late as determined by the rate of blood flow. Selective catheterization may be needed for accurate localization. Demonstration at surgery depends on the presence of visibly abnormal vessels beneath the serosa or in the mesentery, but these may not correspond exactly to the intramuscular or submucosal vessels, which are left to perpetuate the problem.

Aspects of diagnosis, treatment and classification are illustrated by the cases presented here. In the first case, the lesion demonstrated angiographically could not be seen macroscopically. The Doppler probe, mesenteric transillumination or methylene blue injection might well have been of value here, with in vitro injection of the vessels of the resected specimen with dilute barium, and check radiography to avoid incomplete excision. The haemolympathic hamartoma is unusual, but this has also been reported by Taylor and Torrance. The leiomyoma may have been the source of bleeding (not identified on angiography) and the vascular hamartoma an incidental finding. In the second case the hamartoma was easily demonstrated angiographically and macroscopically, and in the third it was not seen on angiography but easily recognized at surgery. No doubt there is a fourth category of patients with lesions which are not demonstrated angiographically or macroscopically at surgery and who present with recurrent unexplained blood loss. A venous malformation may be difficult to see on angiography, and if submucosal it will not be seen at laparotomy either. These patients need repeated investigation, with isotope studies to identify the presence and possibly the site of loss of blood into the gastro-intestinal tract, angiography and endoscopy. The optimal treatment of these lesions is complete excision of the affected area, although if the lesion is irresectable embolization in the pelvis and stomach has been used in preference to attempted ligation of feeding vessels.

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REFERENCES