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CASE REPORT

A 44-year-old woman who had experienced intermittent melena for several months was admitted for evaluation. She had experienced profuse melena for 4 h on the day she visited the emergency room. She had also suffered from dizziness and anemia for several years. Physical examination demonstrated no abdominal pain, tenderness, or mass. Laboratory tests indicated iron deficiency anemia: hemoglobin, 8.1 mg/dL; hematocrit, 23.6%; red blood cells, 2.76 × 106/μL; and serum iron, 55 μg/dL. Colonoscopy showed melenic stool coating the colonic wall, with no colitis or mucosal ulceration. Upper GI endoscopy showed mild gastritis and no active bleeding. Abdominal computed tomography (CT) revealed a 3.6 × 2.0 cm, highly vascularized, strong early arterial enhancing soft tissue mass in the jejunum (Fig. 1A, B). Luminal leakage of contrast material was suspected to be secondary
Angiography for tumor embolization was performed with the presumed diagnosis of AVM or hemangioma of the jejunum. Superior mesenteric arteriogram showed a tortuously enlarged arterial structure, which was tangled in the jejunum and was forming a nidus of AVM. Early-drained veins were also present (Fig. 1C). The angiographic findings were most consistent with an AVM, and the distal jejunal branch of the superior mesenteric artery was selectively embolized with gelfoam particles and two microcoils. Post-embolization angiography showed that the main feeding arteries were occluded. However, some small feeding arteries remained. The patient subsequently underwent an emergency operation. Surgeons found tortuous branches of the superior mesenteric artery feeding the subserosal, hemorrhagic, protruding mass in the jejunum.

The resected segment of the jejunum showed a subserosal, protruding, solid mass with a short stalk measuring 4.5 × 3.6 cm. The mucosa showed ulceration with fresh hemorrhage, corresponding to recent bleeding. Cut sections of the mass demonstrated the typical appearance of a GIST, except for prominent congested vessels. Microscopically, most of the subserosal mass showed an interlacing fascicular arrangement of spindle cell proliferation (Fig. 2A, D). The tumor cells were diffusely positive for KIT immunostaining (Fig. 2B). The average number of mitotic figures was 1/50 high power fields, and there was no cytologic atypia. Histological and immunohistochemical findings were consistent with a spindle cell type GIST of low risk category. A complex vascular malformation consisting of large, tortuous, and irregularly connected arteries and veins was noted predominantly in the submucosal area. It extended into the muscularis propria and even crossed the underlying GIST (Fig. 2A). An elastic stain highlighted internal elastic fibers in most dilated vessels, confirming their arterial character (Fig. 2C). The histologic findings of the vascular lesions were consistent with an AVM. Gelfoam, which was used for transarterial embolization, was noted within the arterial structures of the vascular lesion, in the lumen of intratumoral vessels, and on the ulcerated mucosal surface.

The patient’s postoperative course was uneventful. Hemoglobin and hematocrit returned to the normal range on follow-up laboratory tests.
DISCUSSION

The terms AVM, angiodysplasia (vascular ectasia), and Dieulafoy’s lesion describe distinct entities in the GI tract. AVMs tend to occur in younger patients and are characterized by thick-walled vessels, including arteries, in the submucosa, frequently extending into the proper muscle. Conversely, angiodysplasia is a proliferation of small ectatic vessels and predominantly occurs in elderly patients. Dieulafoy’s lesion is defined as a persistent artery and most frequently occurs in the stomach. To make a diagnosis of AVM in the GI tract, the consideration of both angiographic and histologic findings is mandatory.

Even though GI tract AVMs have most frequently occurred in the jejunum, there are currently fewer than 30 angiographically and histologically proven cases of AVM in the jejunum. In our case, the arterial feeder was identified by angiography, and the arterial nature of the malformed vessels was confirmed by histologic examination. Since the principal features of the vascular lesion in this case were arteries accompanied by mucosal bleeding, it was necessary to differentiate it from Dieulafoy’s lesion. A similar numbers (approximately 45 cases) of Dieulafoy’s lesions have been reported in the jejunum. Dieulafoy’s vascular malformation is characterized by a persistent large artery limited to the submucosal layer immediately under the mucosa. This was not seen in our case. Instead, the vascular proliferation in this case consisted of a complex mixture of arteries and veins extending into the proper muscle layer. Therefore the overall features of our case are consistent with an AVM.

The most interesting feature of our case was the simultaneous development of an AVM and an underlying GIST at the same site in the jejunum. Grossly, the lesion was more likely a solid mass rather than a vascular lesion. However, radiological-
ly, this lesion presented as a vascular lesion rather than as a highly vascular solid tumor. The findings indicate that the AVM extended into the underlying tumor, and there was no distinct border between the two lesions. To the best of our knowledge, there have been only two case reports of GIST associated with a vascular malformation. Tomita et al. reported a case of GIST associated with angiodysplasia in the jejunum. Tomita et al. reported a case of GIST associated with angiodysplasia in the jejunum. That case also presented as a hypervascular mass on angiography and was preoperatively embolized. Vascular changes consistent with angiodysplasia were found mainly within the GIST, and they extended partially into the submucosa. Vats et al. reported a gastric GIST associated with Dieulafoy’s vascular malformation that was diagnosed endoscopically. Our case is characterized by the extraordinary simultaneous occurrence of a GIST and a histologically and angiographically proven AVM in the jejunum.

Despite marked improvement in diagnostic modalities, small intestinal AVMs are still diagnostically challenging. In our case, the AVM was the cause of bleeding, and the concomitant GIST may have contributed to the symptomatology of the AVM and made it more easily seen on imaging. We believe preoperative angiography and embolization were proper management in this patient.

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REFERENCES