Primary angiosarcoma of the small intestine: radiological appearances

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Abstract. Angiosarcoma of the small intestine is a rare malignancy that is clinically manifested by gastrointestinal bleeding. In the few cases previously reported discussion mainly focused on the recent contribution of immunocytocchemistry in tumor identification. It seems that the radiologic appearance of the tumor is poorly documented, and its preoperative imaging evaluation ill-recognized. A 75-year-old male with an intestinal angiosarcoma is presented. Emphasis is given on the radiologic features of the tumor and the possibility of its preoperative demonstration when using enteroclysis.

Key words: Angiosarcoma – Small intestine – Enteroclysis – Small-bowel tumor - Sarcoma - Vascular tumors

Introduction

Sarcomas of the small intestine other than leiomyosarcomas are very unusual [1]. Primary angiosarcomas of the small intestine are even rarer. They are generally mentioned, but are not illustrated in books or extensive reviews concerning small-intestinal tumors, although their histopathology usually poses diagnostic difficulties [2].

Despite a variety of available radiologic images of small-intestine tumors [3], scant attention has been given in the radiologic literature concerning small-intestinal angiosarcomas, and we were unable to find radiologic descriptions of such tumors. We report a case of a primary angiosarcoma of the small intestine with emphasis on its radiologic features.

Case report

A 75-year-old male was admitted for evaluation of progressive fatigue and diarrhea. Upon physical examination skin palor was prominent. Routine laboratory studies revealed anemia (Ht = 23.3; hemoglobin = 7.7 g/dl), whereas tests for occult blood in feces were positive. An upper gastrointestinal (GI) tract endoscopy, upper GI tract barium series, and an abdominal CT scan were unremarkable. A barium enema showed no abnormality, except for diverticulosis of the sigmoid colon.

The patient underwent blood transfusions and was discharged. He was readmitted 2 weeks later with repeated episodes of melena, anemia (Ht = 19) and effort dyspnea. A technitium (TC) 99m-RBC scanning showed concentration of the isotope in the hepatic flexure 1 h after injection, whereas the entire colon was outlined 12 h later. The findings were attributed to the possible presence of a hemorrhagic lesion at the right colon. A selective angiography that followed proved unsuccessful in identifying the lesion.

An enteroclysis, using dilute barium suspension 25 % w/v, infused through a 135-cm long nasoduodenal catheter, was then performed. It revealed three small (0.5-1.3 cm in diameter), ill-defined, sessile, polypoid filling defects in a jejunal loop (Fig. 1). The contour of the lesions seemed inseparable from the margins of the valvulae, and the value of these findings was underestimated during the initial study and radiologic report.

At exploratory laparotomy five small bleeding nodules were found, spread over the mucosa of a 30-cm segment of the jejunum, histologically consistent with malignancy on frozen sections. Segmental resection of the involved part of the small intestine, together with a 20-cm long segment of the right colon, was then performed.
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Pathologic findings

Surgical material from the small intestine was available for histologic and immunohistochemical studies. The large-bowel specimen was normal. Gross pathologic examination of the jejunal specimen showed five mainly submucosal, elevated, half-domed, purple-red masses ranging from 0.5 to 1.3 cm in diameter, with ulcerated mucosa (Fig. 2). Microscopically, all nodules exhibited features consistent with epithelioid angiosarcoma. Each tumor was characterized by angiomatoid differentiation, although some of them were composed of solid sheets of large cells with abundant eosinophilic cytoplasm and centrally located round or oval vacuolated nuclei with conspicuous nucleoli (Fig. 3). Immunohistochemical staining confirmed the diagnosis by showing that the tumor cells were positive to Factor VIII-Ra, Vimentin and Ulex europaeus-1 Lectin, but negative to Keratin (Fig. 4).

Follow-up

Five days postoperatively the patient’s hematocrit was raised to 40%. However, 2 months later he was readmitted for rectal bleeding. Multiple cutaneous nodules of red-iodine color were noted on his back. A skin biopsy showed that these corresponded to foci of metastatic angiosarcoma. The patient died 10 days later due to uncontrollable GI bleeding. Autopsy was not performed.

Discussion

Angiosarcomas represent a vasoformative malignant type of soft-tissue tumor that exhibit morphologic and functional properties of endothelial cells [1]. They are reported to account for approximately 1% of all soft-tissue and skin sarcomas [2], whereas intra-abdominal angiosarcomas, except of primary hepatic, splenic, renal, and adrenal origin, are extremely rare [4].

The actual incidence of primary intestinal angiosarcomas remains unknown. This is attributed to existing difficulties in establishing a preoperative diagnosis, combined with inadequate clinical and histologic documentation of most of the here to fore reported cases, and to a rather confusing terminology that is still in use [5].

Gentry et al. [6], reviewing the Mayo Clinic’s experience with 106 vascular tumors of the GI tract (seen in a 20-year period) reported only three small-intestine angiosarcomas. Since then large reviews of primary small-intestine tumors have only sporadically included rare cases of angiosarcomas, and in a more recent cumulatively similar series [1] six additional cases were reported, most of them with poor histologic documentation [5]. However, another recent review of angiosarcomas of the GI tract emphasizes the rarity of these tumors while raising the possibility that a number of them may remain unrecognized [7].

Angiosarcomas of the GI tract are believed to arise de novo rather than from preexisting hemangiomas [8]. No incriminating correlation with environmental carcinogens, reported to be associated with the development of liver or adrenal angiosarcomas [9, 10], has been reported thus far, except for therapeutic pelvic irradiation, which was implicated as a causative factor in two cases with angiosarcoma of the terminal ileum [11, 12], and in two additional cases of intra-abdominal angiosarcomatosis involving the small intestine [4]. In our patient no previous history of irradiation or relevant intoxication was present.

The clinical presentation of patients with intestinal angiosarcomas has been diverse and rather nonspecific [4]. Gastrointestinal bleeding and/or persistent anemia appear to be the most frequently reported clinical manifestations [1, 5, 7], although signs of intestinal obstruction, epigastric pain, a palpable abdominal mass and/or undue tiredness and weakness may also be present. Altered bowel habits have also been reported [4], and this was an additional feature in our patient in the form of recurrent episodes of diarrhea.

Unlike colonic angiosarcomas, which exhibit a strong female preponderance and an age range at diagnosis between 16 and 46 years [13], primary-intestine angiosarcomas are reported to occur between an age range of 64 and 80 years, with a female-to-male ratio of 2:1 [5, 7]. Our case was compatible with this age and sex predilection data.