Angiosarcoma of the Colon

Report of a Case with Long-Term Survival


A case of angiosarcoma of the large bowel is presented. The tumor occurred in a 16-year-old girl who presented with lower abdominal pain and rectal bleeding. A sigmoid colectomy was performed. Although macroscopic omental and pelvic peritoneal metastases were noted at operation, she did not receive adjuvant therapy and was alive and well more than three years after surgery. The literature on colonic angiosarcoma is also reviewed. [Key words: Angiosarcoma; Colon; Hemangioendothelioma; Hemangiosarcoma]

ANGIOSARCOMA IS A malignant tumor of vascular endothelium. Common sites of occurrence include the skin (with or without lymphedema), breast, deep soft tissue, and liver.1 Gastrointestinal angiosarcomas are exceedingly rare and only two cases, arising primarily from the colon, have been reported.2,3 The paucity of information available on these colonic neoplasms means that little is known about the rate of spread and the optimal mode of treatment.

Report of a Case

A previously well 16-year-old high school girl presented to The Royal Melbourne Hospital with a three-week history of episodic left iliac fossa pain. On the day of admission there were two episodes of painless, bright rectal bleeding, amounting to approximately 50 ml. There were no other gastrointestinal, urinary, or gynecologic symptoms.

Examination revealed a thin teenager in no distress. Her blood pressure was 130/65 mm Hg and pulse 106 beats/min. There was an ill-defined, mildly tender mass in the left iliac fossa. Bright blood was noted in the rectum on digital examination and at sigmoidoscopy, but no mucosal lesion was seen. The physical examination was otherwise normal.

An air-contrast barium enema demonstrated a lesion compressing the midsigmoid colon, but the mucosa appeared intact. On flexible sigmoidoscopy, a 5-cm mass was found protruding into the lumen of the sigmoid colon at 25 cm. It had an ulcerated, blue-red, domelike appearance. Histology of the biopsy specimen revealed only necrotic tissue.

Two days after admission, the rectal bleeding recurred, requiring blood transfusion; a laparotomy was also performed that day. At operation, a 5 × 5 cm black mass was found arising from the antimesenteric surface of the sigmoid colon. The greater omentum was adherent to the mass and numerous black satellite lesions, measuring 0.2 to 2 cm in diameter, were scattered throughout the greater omentum and on the surface of the pelvic peritoneum.

The pelvic organs, remaining colon, small bowel, and liver were normal. There were no enlarged abdominal lymph nodes. A sigmoid colectomy and omentectomy were performed, removing over 95 percent of the macroscopic disease. The patient made an uncomplicated recovery from surgery. Three years later, on follow-up, she was alive and well with no evidence of disease. Her platelet count and fibrin degradation products were within the normal range at the initial presentation and during follow-up.

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FIG. 1. Angiosarcoma of the sigmoid colon measuring 5.5 X 3.0 X 2.5 cm. A. A small segment of sigmoid colon from which the fascia and attached omentum have been dissected to expose the extramural and intramural portions of the lesion. The external surface is irregular as a result of 2 to 3 mm diameter blood-filled blebs. B. The opposite cut surface shows the blood-filled interior, some attached fat, and the domelike intracolonic projection surmounted by a necrotic, ulcerated, nipplelike projection from the mucosal surface.

FIG. 2. Photomicrograph of the large vascular spaces containing blood, thrombus, and polypoid masses of tumor showing a sinusoidal anastomosing pattern and papillae (hematoxylin and eosin; X 50).

FIG. 3. Photomicrograph of a solid portion of the angiosarcoma showing vascular channels lined by pleomorphic endothelial cells containing a few mitoses indicated by arrows (hematoxylin and eosin; X 250).

Pathology

The resected sigmoid colon showed a dark, blue-black, ovoid mass (3.5 X 3.0 X 2.5 cm) protruding from its antimesenteric surface, which, on the cut surface, extended for a further 2.0 cm through a breach in the wall of the colon to produce a mucosal nodule 2.0 cm high and 2.0 cm wide at its base. The outer surface of the lesion, where it was free from the omentum, had a blistered appearance produced by numerous 0.2- to 0.3-cm diameter blood-filled blebs (Fig. 1). The cut surface was composed of vascular spaces containing free blood and blood clot. The mucosal surface of the colon, apart from the main lesion, was unremarkable. Further blood-filled nodules 0.1 to 1.0 cm in diameter were seen on the external surface of the colon adjacent to the main lesion and throughout the resected greater omentum (13.0 X 5.0 X 1.0 cm).

Microscopically, the lesion consisted of numerous, freely anastomosing, vascular channels of varying size ranging from sinusoidal to large angulated venous and cavernous spaces infiltrating the pericolic fat, attached omentum, and extending through the muscularis propria into the submucosa and mucosa of the colon. Large vascular spaces contained tufts of endothelial cells projecting into their lumina (Fig. 2). The plump or elongated endothelial cells showed anisocytosis, nuclear pleomorphism, scattered mitotic figures, and strongly expressed factor VIII-related antigen as demonstrated by the immunoperoxidase technique (Figs. 3 and 4). Areas of necrosis and thrombosis were surrounded by clusters of hemosiderin-filled macrophages and lymphocytes within the spindle-celled stroma. Similar