Hemangioendotheliomas are poorly differentiated neoplasms built from strands of endothelial cells and have a very narrow lumen filled with blood cells. Hemangioendotheliomas are commonly detected during the initial six months of life, concerning mainly soft tissues and skin. It is the most common benign liver cancer in children. In adults, hemangioendotheliomas are very rare findings, being mostly described in the liver, lungs, brain and bones. Hemangioendotheliomas of the large intestine are extremely rare in adults. Cancer developing there is of high degree of malignancy. The authors present the case of 68 year old patient in whom preoperative benign tumor with ulcers were diagnosed. Intraoperative macroscopic picture suggested malignant process. The patient underwent anterior rectal resection with regional lymphadenectomy. The postoperative histopathologic evaluation with immunohistochemical studies angiosarcoma was diagnosed. A four year survival free of cancer was obtained, and the patient died due to myocardial infarct. Literature describes the short periods of survival of patients with vascular sarcoma of the colon. The most common reason for decease are metastases with massive haemorrhages. Bleeding tendency results from growing Kasabach-Merritt syndrome. It is characterized by profound thrombocytopenia and coagulopathy. The presented patient is a unique example of angiosarcoma with long period of survival.

**Keywords:** angiosarcoma, large intestine, anterior resection of rectum, treatment, surgery.

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**Angiosarcoma of the large intestine – a case report**

Zbigniew Kamocki, Robert Wilamowski, Joanna Reszeć, Konrad Zaręba, Boguslaw Kędra

1Department of Medical Pathomorphology, Medical University of Białystok, Poland
22nd Department of General and Gastroenterological Surgery, Medical University of Białystok, Poland

**Introduction**

Angiosarcomas are poorly differentiated neoplasms composed of stands of endothelial cells and have a very narrow lumen filled with blood cells. They usually develop during childhood within superficial or deep soft tissues. Angiosarcomas frequently originate from the vascular wall, usually from medium and large sized veins. They are malignant but rarely result in recurrences or metastases to lungs and lymph nodes. Severe atypia is associated with a more aggressive course [1]. Angiosarcomas are commonly detected during the initial six months of life, since they usually involve the soft tissues of head and skin. They are the most frequently found benign neoplasms of the liver in children [2]. In adults angiosarcomas are very rare findings, being mostly described in the liver, lungs, brain and bones. Angiosarcomas of the large intestine are extremely rare in adults.

**Case report**

A 68-year-old patient was admitted due to loose bowel movements with traces of blood observed for a period of two weeks. The symptoms were initially recorded by the patient 6 months prior to admission and took the form of blood in the stool. The patient’s body weight did not change over this period of time. Sigmoi doscopy revealed an elevated ulcerative lesion 20 mm in diameter, located 12 cm from the anal verge along with two small polyps (4 mm and 8 mm) situated 25 cm from the anal verge. Histopathology of the ulceration showed a tubular adenoma with chronic mucosa inflammation. Above the splenic flexure of the transverse colon, the colonic mucosa was pink and glossy with a mesh of dilated vessels suggestive of chronic inflammation. Remaining segments of the large intestine were normal on sigmoidoscopic examination.

In spite of the benign pathologic result the patient was qualified for en bloc surgical resection on these aforementioned findings. A hard, exophytic tumor (5 cm × 5 cm) was found intra-operatively on the mesorectal side of the rectum. It was gray with cherry foci of blood extravasations. The patient underwent resection of the upper rectum and sigmoid colon (with formation of end-to-end anastomosis with mechanical stitches) with regional lymphadenectomy. No complications were noted in the postoperative period and the patient was discharged eight days after surgery.

Histopathology of the tumor revealed a G3 malignant angiosarcoma with chronic inflammation of the colonic mucosa (Fig. 1–6). The surgical resection margin was negative and no metastases were found in regional lymph nodes.

**Discussion**

Colorectal angiosarcoma is a very rare finding. Only a few well-documented cases of angiosarcoma in this location have been reported in the literature.
The rare occurrence and high aggressiveness of this tumor preclude designing an optimal therapeutic approach. Surgical treatment seems to be the only effective treatment modality in cases of this malignancy with a colorectal localization. The reported cases of angiosarcoma have documented early spread of this malignancy after surgical treatment and a short overall survival. Among these cases was a 77-year-old patient reporting a history of constipation and bleeding from the lower alimentary tract, who was operated on for colorectal angiosarcoma. Numerous liver metastases were detected following the resection of the primary tumor and the patient died six months after surgery [3]. Another reported case involved a 72-year-old patient with pulmonary metastases and colorectal angiosarcoma as the underlying primary malignancy. Initially, this patient was subjected to partial sigmoid resection. He was re-operated on several months later due to massive rectal bleeding, and intraperitoneal spread was detected. Left-side hemicolectomy was performed along with partial stomach resection and total

**Fig. 1.** Fragment of intestinal wall with neoplastic infiltration of muscular layer. On the left visible intestinal mucosa with lymphoid tissue. HE, magnification 50×

**Fig. 2.** Area of neoplastic necrosis infiltrated by neutrophils. HE, magnification 50×

**Fig. 3.** Neoplastic tissue consisting of epithelioid cells. Numerous erythrocytes between neoplastic cells. HE, magnification 100×

**Fig. 4.** Atypical, epithelioid cells without tendency to form blood vessel structures. Some cells form single cell intracytoplasmic lumens containing single erythrocytes. Mitotic figures present. HE, magnification 400×

**Fig. 5.** Immunohistochemical staining of endothelial cells with CD31 antibody. HE, magnification 100×
Omentectomy. The patient died a few days after the procedure due to intraperitoneal hemorrhage [4]. Additionally, the case of a 60-year-old female patient was presented, who was also operated on due to colorectal angiosarcoma. This patient died four months after surgery due to distant metastases [5]. Little is known on the long-term outcomes of adjuvant treatment (radiotherapy and chemotherapy) following the surgical resection of angiosarcomas. Komorowski presented a case involving a 19-year-old patient, who was diagnosed with this malignancy. The patient was operated on and received four subsequent courses of chemotherapy and radiotherapy. At follow-up after 18 months, the patient was still alive with no signs of recurrence [6]. Up to now, the longest postoperative survival in a colorectal angiosarcoma patient was reported by Smith et al., where a 16-year-old female patient survived 36 months after surgery [7]. However, in most of the above-described cases, patients diagnosed with colorectal malignancies died within one year after resection due to neoplastic spread or postsurgical complications. In contrast, our patient survived four years after angiosarcoma resection and died due to myocardial infarction. Therefore, this is the first reported case of such long-term survival (48 months) after resection of colorectal angiosarcoma. Neither positive lymph nodes nor distant metastases were found intraoperatively in this patient. Also no post-operative complications were noted. In conclusion, the surgical approach implemented in our patient proved efficient, and seems the only effective modality in cases of angiosarcoma of such unusual locations as the large intestine. Further research is needed in order to verify the role of adjuvant treatment after surgical resection.

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Address for correspondence
Zbigniew Kamocki
Department of Medical Pathomorphology
Medical University of Bialystok
ul. Waszyngtona 13
15-269 Bialystok
e-mail: zkamocki@yahoo.com

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