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Colonic Hemangiopericytoma

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Background	Hemangiopericytoma is an uncommon perivascular tumor. Descending colon hemangiopericytoma is extremely rare. Here, we report the clinical, ultrasonography, contrast-enhanced computed tomography (CT), and pathologic findings of an HPC arising from the posterior wall of the descending colon in a 42-year-old man.
Summary	Abdominopelvic contrast-enhanced CT showed a 7 cm heterogeneous tumor characterized by internal necrotic material detected in the left lumbar region extending to the left iliac fossa, infiltrating the descending colon. Intraoperative findings revealed a well-capsulated solid tumor surrounding the posterior wall of the descending colon. Positive CD34 and STAT6 markers, along with negative S100, CD117, DOG1, and smooth muscle actin, confirmed the diagnosis of hemangiopericytoma. Follow-up imaging showed no signs of recurrence or metastasis.
Conclusion	HPCs should be included in the differential list of a sizeable mesenchymal tumor with heterogeneous enhancement in the regions of the tumor.
Key Words	hemangiopericytoma; mesenchymal tumor; gastrointestinal stromal tumor; pericytes; STAT6; spindle cell tumor
Abbreviations	HPC: hemangiopericytoma GIST: gastrointestinal stromal tumor US: ultrasonography MFH: malignant fibrous histiocytoma

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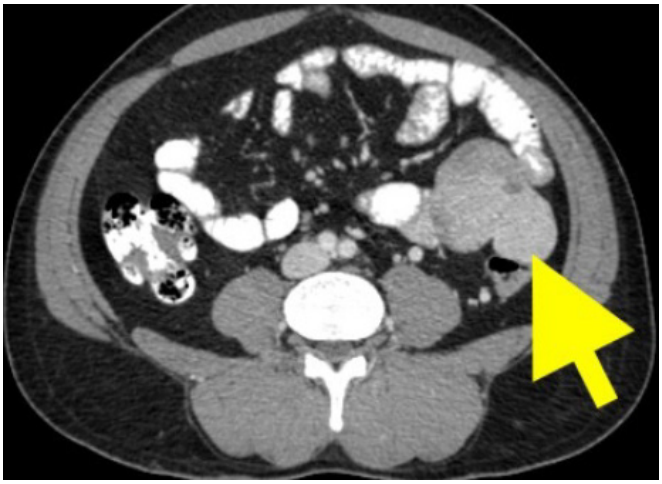
Case Description

A 42-year-old man presented with complaints of left-sided abdominal pain associated with bleeding per rectum of 30 days duration. He underwent a laparotomy for resection of a retroperitoneal malignant tumor in 2009; on histopathological examination, the tumor was consistent with malignant fibrous histiocytoma. The physical and rectal examination did not reveal any abnormalities. Complete blood count, serum electrolytes, creatinine, and urea were within the normal range, and screening for viral markers was negative. He underwent abdominal ultrasonography, which revealed a large rounded lesion, predominantly hypoechoic lesion of size more than 7.2 × 7.7 cm in the left hemipelvis, anterior to left iliac vessels and close to the midline. The lesion showed increased vascularity, and the site of the origin of the lesion could not be defined. Colonoscopy showed a nodular area with evidence of luminal compression and a visible vessel with an intermittent spur. Therapeutic intervention was not performed as there was no active bleeding.

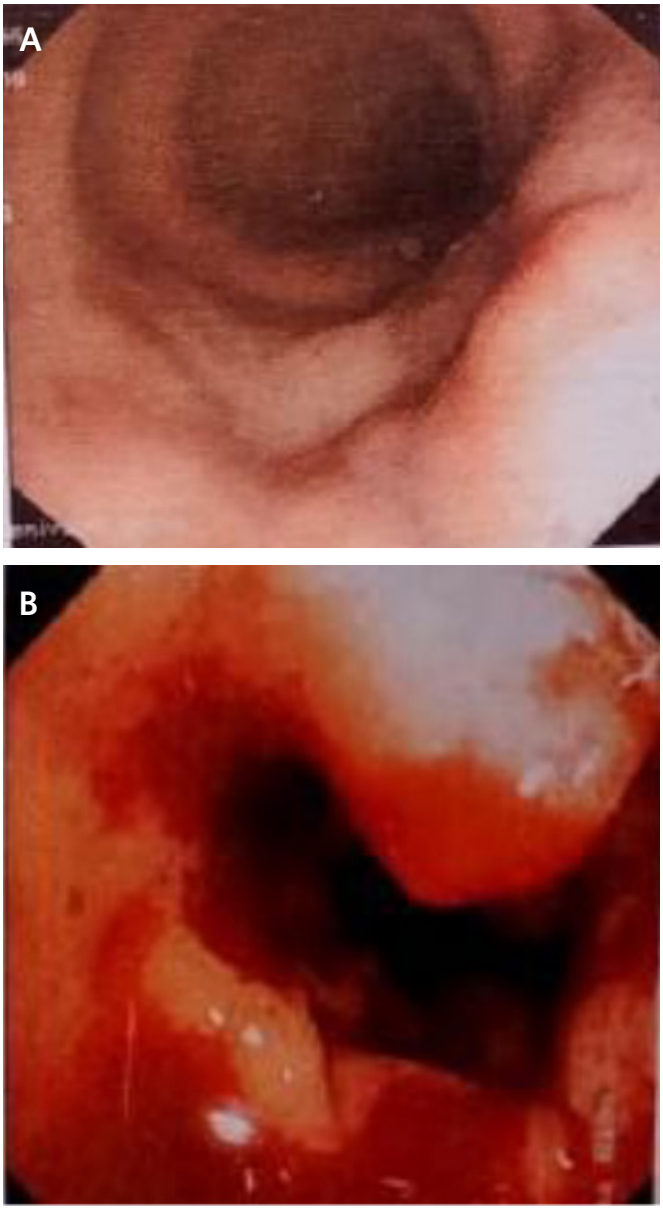
An abdominopelvic contrast-enhanced computed tomography scan showed a soft tissue attenuating mass lesion measuring about 72 × 70 × 60 mm in the left lumbar region extending into the left iliac fossa (Figure 1). There was a heterogeneous gradual enhancement in the tumor with a few foci of necrotic areas. Multiple enhancing venous channels were seen in the vicinity of the lesion, particularly in the posterior and left lateral aspects; it was encasing the adjacent descending colon, causing compression of its lumen. It appeared consistent from the adjacent psoas muscle. The kidney was separate. There was no evidence of metastasis.

Figure 1. CECT of Abdomen. Published with Permission



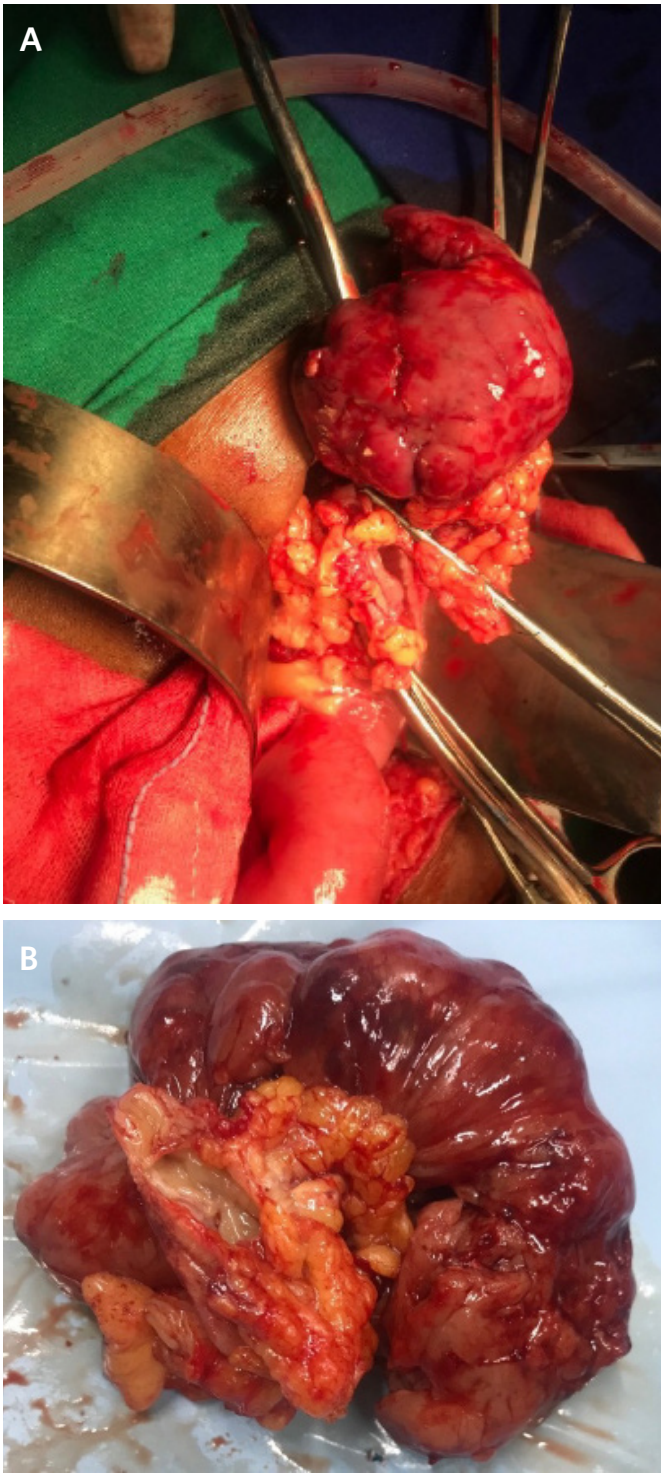
Scan demonstrates a soft tissue mass lesion located in the left lumbar region, with infiltration into the descending colon.

Figure 2. Colonoscopy Showing Nodular Area in Descending Colon. Published with Permission



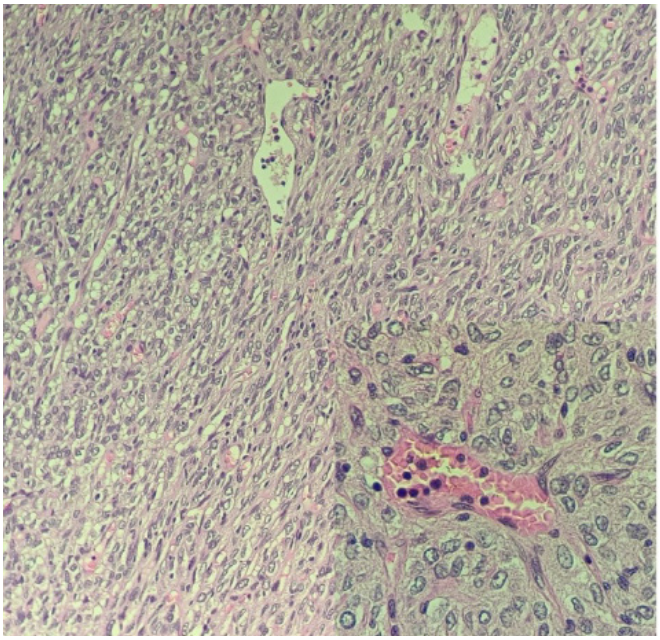
The patient underwent surgery after a thorough evaluation of clinical and imaging findings. Intraoperatively, there was a well-capsulated and circumscribed solid tumor on the posterior wall of the descending colon. Tumor resection, along with involved colonic segment, was done with end-to-end anastomosis. His postoperative period was uneventful.

Figure 3. Intraoperative Image of Lesion and Resected Specimen Along with Part of Descending Colon. Published with Permission



The gross specimen showed a well-encapsulated mass 4 × 3 cm in size. The cut section of the tumor mass showed a greyish-white, fleshy appearance and was multinodular in some places. Microscopically, the specimen showed highly cellular pleomorphic spindle-shaped cells with a patternless, fascicular, whorled, storiform, and palisading architecture. The vascular network was prominent. Tumor cells were invading through the serosal layer of the attached colonic segment. With immunochemical histology stains, the tumor cells reacted positively for CD34 and STAT6 and negatively for CD117, S100 protein, smooth muscle actin, and DOG1. The tumor had low malignant potential activity.

Figure 4. Microscopic Histological Section Showing Infiltrating Cellular Spindle Cells Tumor. Published with Permission



Tumor cells are arranged in patternless, fascicular, whorled, storiform, and palisading patterns. The cytoplasm is moderate, spindly, and amphophilic to clear, with defined cellular borders. Mitotic activity 5-7/50 HPF seen. Stroma is scant and collagenous with mild lymphoplasmacytic infiltrate seen. Foci of hemorrhages and coagulative necros noted. Tumor has rich vascularity.

Six months post-surgery, a subsequent abdominal ultrasound examination showed no signs of tumor recurrence or metastasis. Additionally, a follow-up colonoscopy was performed and found to be normal.

Discussion

Wagner first described the solitary fibrous tumor in 1870.[1] Hemangiopericytoma (HPC) is an uncommon perivascular tumor and accounts for 1% of primary vascular tumors.[2] HPC develops from the specialized cells lining the capillaries, pericytes, which are small, spindle, or oval-shaped cells lining capillaries.[3] It occurs most frequently in the extremities, pelvis, head, neck, and meninges.[2] The neoplasm occurs most commonly in the mesenchymal soft tissue, but hemangiopericytomas are prevalent in other viscera, including the brain, oral cavity, thyroid, urinary bladder, and prostate.[4]

The first description of HPC was published in 1942 by Stout and Murray.[5] HPC is a rare tumor that originates from Zimmermann pericytes, which are primitive mesenchymal cells closely linked with endothelial cells of capillaries.[5] Pericytes, which are contractile cells encircling capillaries and postcapillary venules,[6] primarily manifest in the pelvis, lower extremities, and retroperitoneum. It appears to have no predilection for age[7] and exhibits a male-to-female ratio of 1.8.[8]

HPCs typically originate from the stomach[4] and small bowel,[9] with rare occurrences in the esophagus, colon, and rectum. Symptoms in the large intestine may include obstruction, bleeding, and intussusception.[10] Recurrence rates are notably high (up to 52%),[11] underscoring the importance of long-term follow-up post-primary tumor resection. Other characteristic features are various paraneoplastic symptoms, commonly hypoglycaemia[12] and hypertension,[13] as the tumor cells can secrete insulin-like substances. The imaging features of colonic HPCs are nonspecific. A large HPC can have necrosis and cystic changes, but calcifications are rare.

Intense heterogeneous enhancement is observed in contrast-enhanced CT along with multiple enhancing venous channels near the lesion, indicating the tumor's vascular origin. Still, there is uncertainty about the origin of the tumor. Magnetic resonance imaging is the generally chosen modality of imaging for detecting the organ of origin of a mesenchymal tumor. MRI was not performed on our patient. MRI shows the characteristic sign "flow void phenomena," often emerging from hypervascular tumors.[8] They are hyperintense on gadolinium-enhanced images. Lipomatous HPCs are benign variants of HPCs.[14] Colonic HPCs must be differentiated from gastrointestinal stromal tumors(GIST) and retroperitoneal tumors. GISTs rarely cause lymph nodal metastasis. Other differential diagnoses should be considered if extensive lymph nodal metastasis is observed.[15] It is challenging to differentiate GISTs and common retroperitoneal tumors like leiomyosarcoma, liposarcoma, and malignant fibrous histiocytoma (MFH) on the background of imaging findings.

Conclusion

Colonic HPC is a rare tumor with nonspecific imaging findings. HPCs should be included in the differential list of a sizeable mesenchymal tumor with heterogeneous enhancement in the regions of the tumor.

Lessons Learned

Patients who undergo complete surgical resection have an 80% survival rate at five years.[16] Adjuvant radiotherapy is recommended. Research indicates the benefit of radiotherapy for controlling local recurrence of tumors that were not completely excised. Given the high recurrence rate of these tumors, long-term follow-up, including annual abdominal and pelvic ultrasound examinations, is essential.

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