Primary hepatic angiosarcoma with multifocal metastases in the gastrointestinal tract

A 64-year-old man presented with a 2-month history of melena. He also had a history of chronic renal disease, hypertension, and diabetes mellitus, and he had worked at a leather factory for decades. He was diagnosed as having primary hepatic angiosarcoma and had undergone right lobectomy 4 months before this admission. Esophagastroduodenoscopy and colonoscopy failed to detect the bleeding source. Capsule endoscopy showed multiple small hemorrhagic spots in the small intestine. Anterograde double-balloon enteroscopy (DBE) demonstrated multiple ulcerative, sessile polyps with oozing in the small intestine and a 2.5-cm tumor with hemorrhage in the distal jejunum (Fig. 1 a, b). The tumor was resected by polypectomy; however, intermittent melena was still present. Retrograde DBE demonstrated a 2.5-cm ulcerative tumor with hemorrhage in the ascending colon (Fig. 1 c), which was also removed by polypectomy. Histopathological examination of the ulcerative polyps and tumors showed spindle tumor cells arranged in solid sheets rich in minute vascular channels (Fig. 2 a). Immunohistochemical study of tumor cells showed positivity for CD31 (Fig. 2 b) and CD34 (Fig. 2 c). A diagnosis of metastatic angiosarcomas was made. Although thalidomide was prescribed for the patient, he died within 3 months of the diagnosis.

Angiosarcoma, a rare high grade vascular malignant tumor, most frequently involves the skin and subcutaneous tissues. It can also occur in breast, liver, spleen, bone, ovaries, and the adrenal glands. The etiology of angiosarcoma is still unclear, but in recent decades, several environmental carcinogens such as Thorotrast, vinyl chloride and arsenic compounds have been thought to be associated with angiosarcoma [1]. The most common site of metastases of primary hepatic angiosarcoma is lung followed by spleen [2]. Intestinal metastases in primary hepatic angiosarcoma are extremely rare. The most common presenting symptoms of intestinal angiosarcoma include abdominal pain, hemorrhage, vomiting, anemia, and weakness. Early detection and diagnosis, followed by operative resection, remains the mainstay of management of this tumor and significant long-term survival. The prognosis of patients with angiosarcoma is very poor [3].
Fig. 2  a Histopathological section of the resected tumor showed spindle tumor cells arranged in solid sheets rich with minute vascular channels (hematoxylin and eosin, original magnification × 400). b Immunohistochemical study of the tumor cells showing diffuse and strong positivity for CD31 (original magnification × 100). c Immunohistochemical study of tumor cells showing diffuse and strong positivity for CD34 (original magnification × 100).

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