Colonic metastasis from subcutaneous angiosarcoma: A diagnostic dilemma

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Abstract Primary angiosarcoma is an extremely rare and aggressive soft-tissue malignancy of endothelial cell origin that occurs most frequently in the skin and subcutaneous tissues of the extremities. Presence of this disease in the intestine as a primary or metastatic deposit is an extremely uncommon incident, and might causes diagnostic confusion with primary colonic neoplasm. We encountered epithelioid angiosarcoma of the sigmoid colon in a 59-year-old male patient who presented with occasional bleeding per rectum and had also had an angiosarcoma arising from the subcutaneous tissue of the right thigh. Retrospective review of histopathology and immunohistochemistry of the two specimens were helpful in the final diagnosis.

Keywords Angiosarcoma · Colonic metastasis

Introduction

Angiosarcoma is a rare and aggressive neoplasm that constitutes approximately 1–2% of all soft tissue tumor [1]. Most frequently, it arises from skin and soft tissue of the extremities, retroperitoneum, breast, bone, liver and spleen. However, gastrointestinal angiosarcoma, as a metastatic or primary disease, is extremely uncommon [2, 3]. Due to its clinical presentation being similar to other gastrointestinal malignancy, preoperative diagnosis often becomes challenging. Here we report a case of epithelioid angiosarcoma of sigmoid colon, which also presented with a similar neoplasm arising from the subcutaneous tissue of right thigh.

Case report

A 59-year-old male had presented with history of malena, occasional bleeding per rectum and intermittent colicky abdominal pain for the last 5 months. In addition, there was a gradually increasing painful swelling of the right lower thigh for the last 8 months. Examination revealed a vague ill-defined mildly inflamed mass in the anterior aspect of right lower thigh, while rest of the system was unremarkable. Following an uninformative per rectal examination and gastroduodenoscopy, colonoscopic examination revealed a relatively large sessile polypoidal growth at the mid-sigmoid region. Endoscopic biopsy showed normal colonic epithelium. MRI scan of the thigh showed a well-defined homogeneously enhancing hyper-dense septated lesion measuring $5.6 \times 4.3 \times 8.1$ cm, suggestive of inflammatory or infective etiology. FNAC revealed blood cells, and antibiotics started for a presumed inflammatory swelling. Except for a low hemoglobin level

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(6.5 gm/dl), routine blood investigations including LFTs and coagulation profile were within normal limits. Contrast CT scan of abdomen and a chest X-ray did not demonstrate any metastasis. He underwent an uneventful laparoscopic sigmoid colectomy and histopathology was reported as malignant gastrointestinal stromal tumor (GIST) (Fig. 1). Subsequent immunohistochemistry demonstrated strong reactivity to cytokeratins (CKs) and immunonegativity for CK-20, CK-7, c-KIT protein, CD34, S-100, HMB45 and desmin, thereby raising the possibility of this being a poorly differentiated colonic adenocarcinoma.

In the meanwhile, the swelling over the thigh increased in size necessitating readmission and wide excision by the Orthopaedic surgeons. Histology of resected specimen showed high-grade epithelioid angiosarcoma (Fig. 2). This was corroborated by immunohistochemistry, which revealed

Fig. 1 Photomicrograph of sigmoid colon tumor shows vascular spaces lined by atypical endothelial cells and frequent mitotic figures

Fig. 2 Photomicrograph of soft tissue tumor of thigh shows vascular spaces lined by atypical endothelial cells. Intracytoplasmic RBCs and vascular lumina are noted

cells expressing CKs CD31, CD34, CKs and Fli-1 markers. The light microscopic features of the thigh and colonic neoplasm were exactly identical and the previous diagnosis of colonic adenocarcinoma was therefore amended to angiosarcoma!

The patient has received 3 cycles of doxorubicin and ifosfamide based chemotherapy and is surviving without disease recurrence at 3-month follow up.

Discussion

Angiosarcoma is derived from vascular endothelial cells, and is classically represented by freely anastomosing vascular channels lined by atypical endothelial cells [4]. Such preponderance of grossly atypical endothelial cells were also found within the coalesced vessels in this patient. Microscopic appearance might vary from well-differentiated tumor simulating benign haemangioma to grossly undifferentiated and solid tumor simulating adenocarcinoma [4]. Colon is an infrequent site of this disease, and when present, it might produce signs and symptoms similar to other colonic malignancy. Therefore, it is easy to misdiagnose colonic angiosarcoma as haemangioma, adenocarcinoma or GIST and such had been the situation in this case.

Angiosarcoma usually expresses variety of endothelial cell markers depending on the degree of differentiation. Of these, CD31, CD34 and CKs are most characteristic, while CK-20 is virtually absent in all gastrointestinal angiosarcoma, but is a characteristic feature of colonic adenocarcinoma [2]. The colonic tumor of this patient expressed nonspecific CKs while were immunonegative for CK-20 and c-KIT marker. This negates the diagnosis of an adenocarcinoma or GIST, thereby creating confusion in this patient. Thus, histological revision of sigmoid and subcutaneous specimen became necessary and revealed similar microscopic features in both the lesions leading to the diagnosis of angiosarcoma (Figs. 1 and 2).

Angiosarcoma commonly metastasizes to lungs, lymph nodes, soft tissues, bone, liver, and very infrequently to other sites [5]. Literature search revealed scant report of colonic metastasis of this disease [6] and it is difficult to ascertain whether the colonic neoplasm was primary or metastatic spread from the thigh. The latter option is a strong possibility because the disease is notorious for its metastatic potential [7] but the scenario becomes even more confusing, because angiosarcoma could be multifocal at the very outset [8].

Surgery with sizeable normal tissue margin and adjuvant radiotherapy is the mainstay of therapy for localized disease, but poses considerable difficulty in managing metastatic disease. There is very limited role of adjuvant chemotherapy, but doxorubicin and ifosfamide based chemotherapy have shown some promise in palliation of metastatic disease [9].

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