Primary Gastric Angiosarcoma Presenting as an Asymptomatic Gastric Submucosal Tumor

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Primary gastric angiosarcoma is a rare gastric tumor. Previously reported cases are limited and usually symptomatic. Preoperative diagnosis is difficult and should be differentiated from adenocarcinoma or gastrointestinal stromal tumor. We report a 55-year-old man with primary gastric angiosarcoma presenting as an asymptomatic gastric submucosal tumor. Abdominal ultrasonography showed a heterogeneous tumor with several notably anechoic portions between the stomach and the gallbladder. Computed tomography showed that the tumor originated from the stomach, and upper gastrointestinal endoscopy showed a large gastric submucosal tumor at the lower body. Laparotomy was performed and a purple circumscribed tumor measuring 8.5 × 5.0 × 4.0 cm was found on the serosal aspect of the lower body. The tumor was soft and spongy with areas of hemorrhagic pools on section. Microscopically, the tumor was composed of proliferative neoplastic spindle to epithelioid cells. The neoplastic cells infiltrated and dissected the smooth muscle as vessel-forming architectures. On immunohistochemical study, the tumor cells were positive for CD34. These findings confirmed a diagnosis of primary gastric angiosarcoma. The patient has been well during the 11-month postoperative follow-up. [J Formos Med Assoc 2007;106(11):961–964]

Key Words: angiosarcoma, stomach, submucosal tumor

Angiosarcomas represent 2% of all sarcomas and most frequently occur in the skin and subcutis. However, it is also known to affect internal organs such as the heart, liver and spleen. Angiosarcoma of the gastrointestinal (GI) tract is rarely reported in the literature. Among reported cases, only two were of primary gastric angiosarcoma, but both were symptomatic and none presented as a gastric submucosal tumor.2,3

Here, we report an extremely rare case of primary gastric angiosarcoma which presented as an asymptomatic gastric submucosal tumor.

Case Report

A previously healthy 55-year-old man was referred to our clinic for an abdominal tumor found incidentally by abdominal ultrasonography (US) during a routine health examination. Abdominal US showed a heterogeneous tumor, 6.5 × 3.8 cm in size, between the stomach and the gallbladder. There were several notably anechoic portions within the tumor (Figure 1A). On admission, physical examinations were unremarkable and the abdominal tumor could not be palpated.
Laboratory data were essentially within normal limits. Stool occult blood was negative. Carcinoembryonic antigen level was 5 ng/dL (normal, < 5 ng/dL).

Contrast-enhanced abdominal computed tomography (CT) showed a tumor with heterogeneous hypodensity originating from the stomach (Figure 1B). Upper GI endoscopy showed a large gastric submucosal tumor covered by normal-appearing mucosa on the posterior wall of the lower body (Figure 1C). Due to clinical suspicion of large gastric stromal tumor, the patient underwent subtotal gastrectomy. At laparotomy, there was a purple circumscribed tumor measuring 8.5 × 5.0 × 4.0 cm on the serosal aspect of the lower body. Macroscopically, the tumor was soft and spongy with areas of hemorrhagic pools on section (Figure 1D). The mucosal surface was grossly uninvolved. Microscopically, abundant neoplastic cells dissected the smooth muscle bundles, sparing the mucosa and submucosa. The tumor was composed of proliferative neoplastic spindle to epithelioid cells. Of note, there were vessel-forming architectures containing blood lakes or solid sheets in the tumor (Figure 2A). The mitotic activity was up to 2–3 per 10 high-powered fields. On immunohistochemical (IHC) study, the tumor cells were positive for vimentin and CD34, focally positive for cytokeratin (AE1/AE3) and negative for CD117 and smooth muscle actin (Figure 2B). These findings confirmed a diagnosis of mixed epithelioid and moderately differentiated primary gastric angiosarcoma.

The postoperative course was smooth and the patient has been well during the 11-month postoperative follow-up.
Discussion

Vascular tumors of the stomach represent only 0.9–3.3% of all gastric neoplasms. Gastric angiosarcoma is extremely rare. To the best of our knowledge, there have been only two cases of primary gastric angiosarcoma reported. They were prominent in view of mucosal involvement. Upper GI endoscopy with biopsy was performed in both cases. One was diagnosed as angiosarcoma, but the other had an erroneous preoperative diagnosis of poorly differentiated adenocarcinoma. On the contrary, our case is unique in its presentation as an asymptomatic gastric submucosal tumor.

The etiologies of angiosarcoma remain obscure, but it has been linked to exposure to radiation, vinyl chloride, Thorotrast and previous chemotherapy. However, no predisposing factor could be identified in this patient. The diagnosis of angiosarcoma in this case was confirmed after combining the gross appearance, cytomorphology and IHC staining, but the differential diagnosis between gastric angiosarcoma and gastric stromal tumor is difficult according to the findings of preoperative imaging studies. However, the preoperative abdominal US and CT morphology of cystic components corresponded to the gross specimen and may provide a diagnostic hint. Histologic examination with IHC staining is important to confirm the diagnosis. Microscopically, angiosarcomas characteristically show two kinds of growth patterns, vasoformative and solid. The vasoformative structures are lined by spindled or plumped anaplastic endothelial cells. The solid growth pattern consists of two cell types: sheets of spindle-shaped cells or large, polygonal epithelioid-shaped cells with abundant amphophilic or eosinophilic cytoplasm. However, in many cases, they might show only epithelioid histology and be difficult to differentiate from other epithelioid tumors. In these cases, IHC staining including CD31, CD34 and factor VIII plays an important role to confirm the vascular origin.

Treatment of gastric angiosarcoma consists of complete surgical resection, sometimes followed by neoadjuvant chemotherapy. Prognosis is poor, and the previously reported patients died within months after diagnosis. Once completely resected, angiosarcoma may also be curable. In this case, the tumor was encapsulated and the resection margin was free of tumor cells. No further radiotherapy or chemotherapy was performed. Close follow-up of the patient is undergoing.

In conclusion, though gastric angiosarcoma is extremely rare, it should be taken into consideration as a rare differential diagnosis of gastric submucosal tumor, especially when cystic components are detected within the tumor. Preoperative diagnosis is difficult, and histopathologic examination is important for diagnosis. CT or endoscopic US-guided fine needle aspiration for obtaining tissue may be helpful.

Figure 2. (A) There are vessel-forming architectures containing blood lakes or solid sheets in the tumor (hematoxylin & eosin; original magnification, 200×). (B) On immunohistochemical study, the tumor cells are positive for CD34 (original magnification, 40×).
References


