Case Report

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대한영상의학회지

Primary Colonic Epithelioid Angiosarcoma with Hepatic Metastasis: A Case Report

간전이를 동반한 대장 상피모양혈관육종: 증례 보고

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Colonic angiosarcoma is an extremely rare and aggressive malignant tumor with poor prognosis. We report a case of colonic epithelioid angiosarcoma with colonic obstruction and rapidly progressive hepatic metastasis in a 44-year-old female. Abdominal CT revealed a heterogeneously enhancing irregular mass in the ascending colon, causing proximal bowel distension. The patient underwent surgery, and histopathological examination revealed a poorly differentiated carcinoma. A follow-up liver dynamic MRI after 4 months revealed newly developed diffusely scattered numerous small nodules in both hepatic lobes with peripheral and nodular marked arterial hyperenhancement, raising the suspicion of hepatic angiosarcoma. A pathologic second opinion was obtained, and additional immunohistochemistry revealed colonic epithelioid angiosarcoma. The patient showed progressive hepatic metastasis on follow-up abdominal CT after 6 months and died 8 months after initial diagnosis. We describe an educational case of colonic angiosarcoma, a rare malignant tumor, with rapidly progressive hepatic metastasis that showed radiologic findings suggestive of angiosarcoma and enabled a re-diagnosis for proper treatment and prognosis prediction.

Index terms Angiosarcoma; Colonic Neoplasms; Neoplasm Metastasis; Intestinal Obstruction; Multidetector Computed Tomography; Magnetic Resonance Imaging

INTRODUCTION

Angiosarcoma is rare and aggressive malignant vascular or lymphatic origin tumor, representing less than 1% of soft-tissue sarcomas (1). Angiosarcomas arise most commonly in skin, head and neck region, particularly in scalp (2). Angiosarcoma in gastrointestinal tract is extremely rare, more likely in stomach and small bowel (3). Primary colorectal angiosarcoma is even more rare, accounts for 0.0012% of colorectal cancer (4)

and only 33 cases reported worldwide to our knowledge (3). Case reports in the literature discuss pathologic characteristics but not radiologic features (1). Angiosarcomas show poor prognosis with systemic metastasis to lung, liver, bone and soft tissue (1). There was no reported case of primary colonic angiosarcoma and hepatic metastasis with supporting radiologic features in Korea, to our knowledge. Herein, we report of an extremely rare case of primary colon epithelioid angiosarcoma which showed rapidly progressive hepatic metastasis with radiological findings of CT and MR including serial follow up.

CASE REPORT

A 44-year-old female represented diffuse abdominal pain from 4 days ago and a week history of constipation. She had no significant past medical history. Physical examination revealed diffuse abdominal tenderness and rebound tenderness and the vital sign was stable. Laboratory findings revealed normal range of complete blood count with mild elevation of inflammatory markers with erythrocyte sedimentation rate (83 mm/hr) and C-reactive protein (1.32 mg/dL). Carcinoembryonic antigen (CEA) and CA 19–9 were normal.

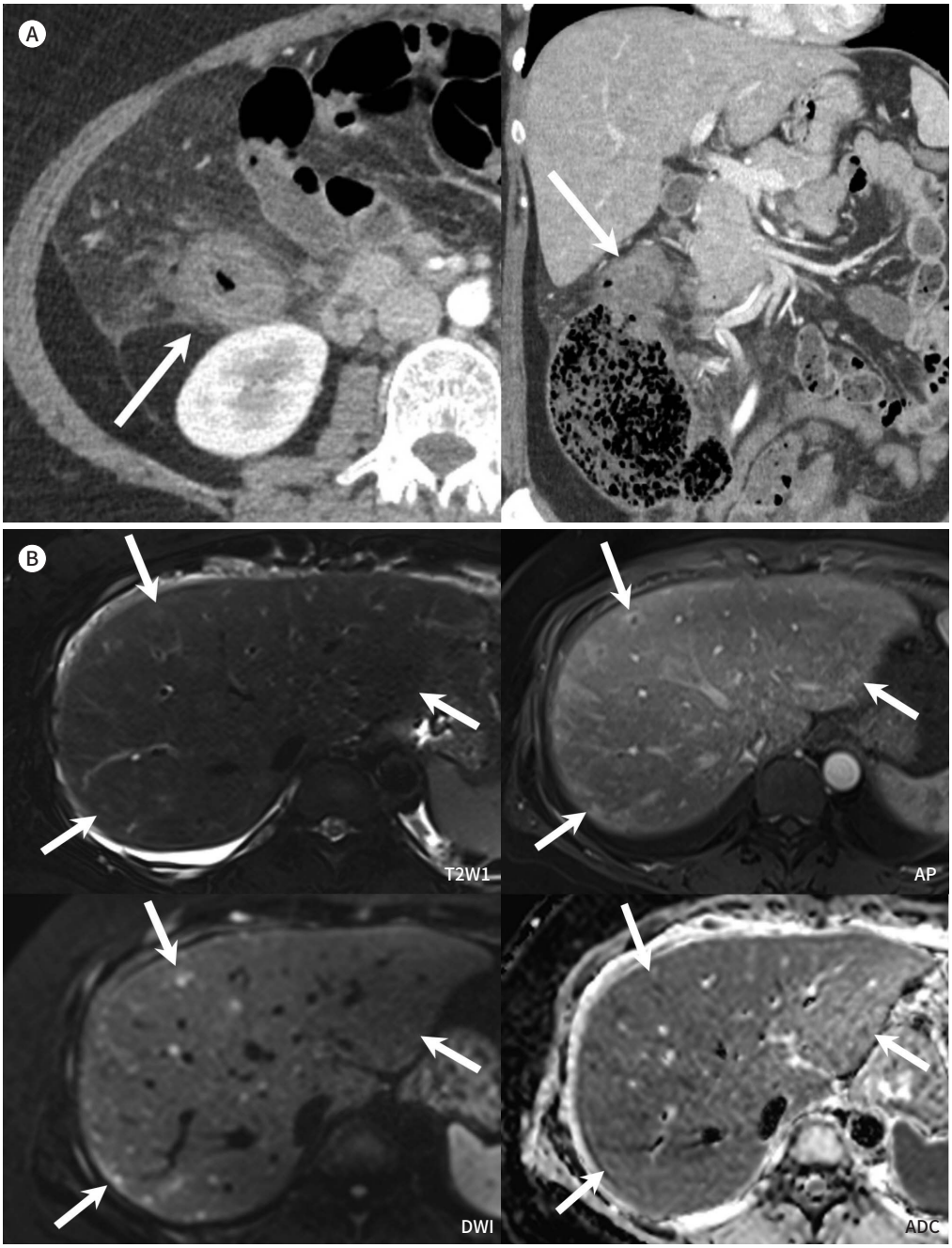
Abdomen simple radiography showed gaseous distension of small bowel loops and feces filled distension of ascending colon and cecum. Axial and coronal contrast-enhanced abdominal CT showed 4 cm sized heterogeneously enhancing irregular shaped mass with pericolic fat infiltration in distal ascending colon, associated with luminal narrowing of distal ascending colon and feces-filled proximal bowel distension in ascending colon, cecum, and distal ileum (Fig. 1A). There were multiple conglomerated enlarged pericolic lymph nodes more than seven in number. There was no evidence of distant metastasis including liver and other solid organ. The impression was distal ascending colon cancer with metastatic lymphadenopathy and associated colonic obstruction, with CT staging of T3/N2b/M0.

The patient underwent extended right hemicolectomy for distal ascending colon mass with gross R0 resection. Histopathologic report suggested primary or metastatic poorly differentiated carcinoma, with 4 cm size of ulceroinfiltrative lesion involving entire wall with pericolic fat spreading. There was proliferation of uniform epithelioid cells in solid nests mostly and stromal hyalinized desmoplasia accompanied by vascular proliferation with frequent areas of intravascular tumor emboli. But there were no consistent immunohistochemical findings. The patient underwent adjuvant chemotherapy for presumed poorly differentiated carcinoma of colon.

On follow up PET-CT after four months showed multiple hypermetabolic lesions in liver and the patient underwent liver dynamic MRI. Diffusely scattered numerous ill-defined small T2 subtle hyperintense nodules were newly appeared in both hepatic lobes with peripheral and nodular marked arterial hyperenhancement and diffusion restriction (Fig. 1B), strongly suspected as hepatic angiosarcoma in that of disseminated hypervascular lesions.

Second opinion for histopathologic diagnosis was asked to outside hospital revealed positive on CD31 and E26 transformation specific (ETS)-related gene (ERG) on additional immunohistochemistry, which are specific endothelial cell markers for epithelioid angiosarcoma and were not performed on initial immunohistochemical stains. Immunohistochemical staining was performed again in our hospital and finally revealed positive on CD31 as outside hospi-

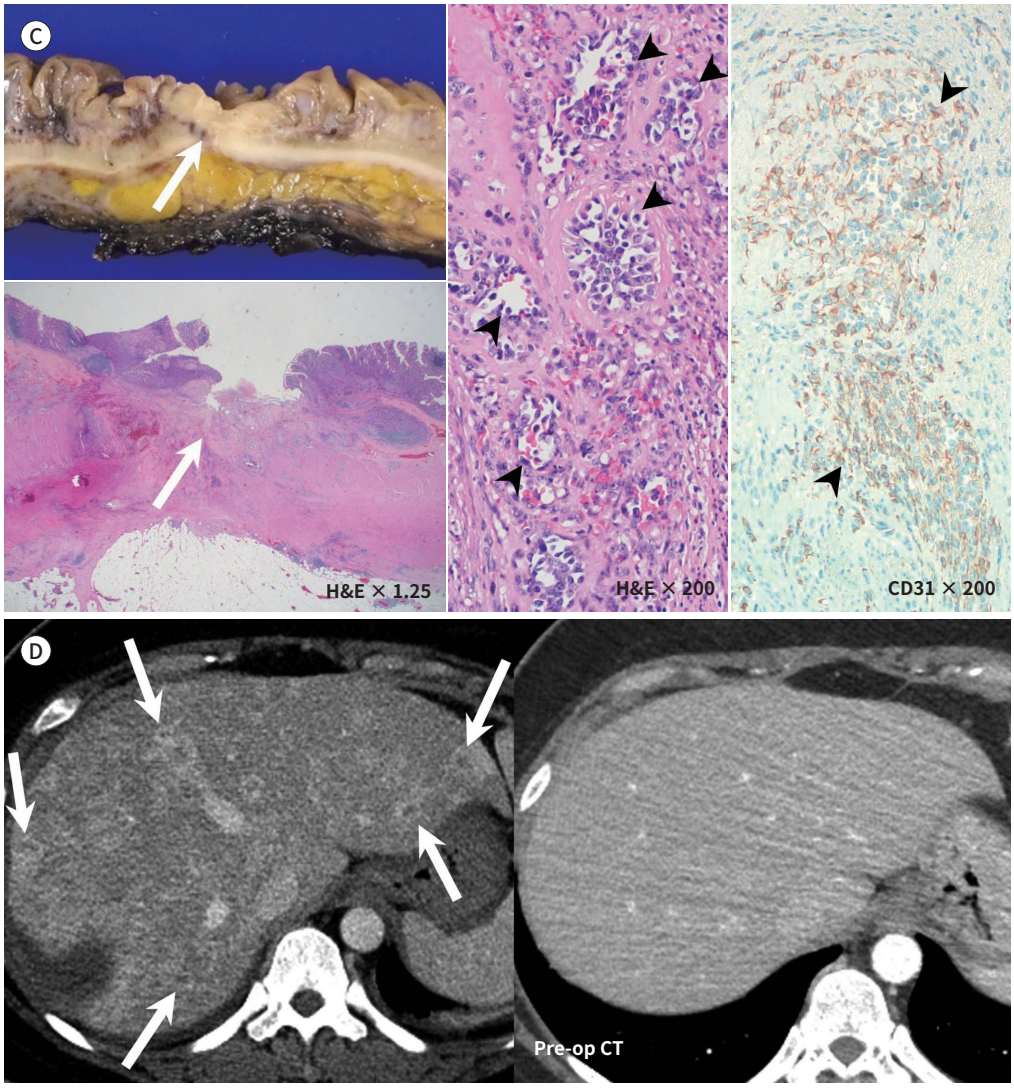
Fig. 1. A 44-year-old female with the primary colonic epithelioid angiosarcoma with hepatic metastasis.
A. Initial axial and coronal contrast-enhanced abdominal CT images show a heterogeneously enhancing irregular mass (arrows) in the distal ascending colon with pericolic fat infiltration. Focal luminal narrowing of the ascending colon and feces filled distension of the proximal colon are also seen.
B. Follow-up axial liver dynamic MRI after 4 months shows newly developed diffusely scattered numerous ill-defined small T2 subtle hyperintense nodules in both hepatic lobes with peripheral and nodular marked arterial hyperenhancement and diffusion restriction (arrows, more than indicated).
ADC = apparent diffusion coefficient, AP = arterial phase, DWI = diffusion-weighted imaging, T2WI = T2-weighted image



tal, suggesting epithelioid angiosarcoma (Fig. 1C).

The patient changed chemotherapy regimen on epithelioid angiosarcoma. On axial contrast-enhanced abdominal CT after six months showed diffusely scattered numerous small nodules in both hepatic lobes with peripheral and nodular marked hyperenhancement and central hypoattenuation, newly appeared compared to pre-operative abdominal CT (Fig. 1D), suggesting progression of liver metastasis of angiosarcoma. The patient showed progression of disease with pneumonia despite of chemotherapy and antibiotics. The patient finally expired

Fig. 1. A 44-year-old female with the primary colonic epithelioid angiosarcoma with hepatic metastasis.
C. Cut section of the gross specimen reveals an ill-defined transmural gray-white lesion (arrow). Low-power view shows an ill-defined transmural infiltrative tumor with surface ulceration (arrow, H&E stain, × 1.25). High-power view reveals irregular vascular channels lined by atypical cells (arrowheads, H&E stain, × 200) that are positive on immunohistochemical staining for CD31 (stained in brown) (arrowheads, × 200).
D. Follow-up axial contrast-enhanced abdominal CT image after 6 months shows diffusely scattered numerous small nodules in both hepatic lobes with peripheral and nodular marked hyperenhancement and central hypoattenuation (arrows, more than indicated) that are newly appeared compared to pre-operative abdominal CT. Furthermore, loculated fluid collection in the subcapsular area of the liver is newly observed.
H&E = hematoxylin and eosin, op = operation



after two months from CT and eight months from initial diagnosis.

This study was approved by the Institutional Review Board of our institution and the requirement for informed consent was waived (IRB No. SCHUH 2021-03-028).

DISCUSSION

Primary colonic angiosarcoma is extremely rare with only 33 cases reported in the literature to our knowledge (3). The median age at presentation was 56 years with range of 16–85 years (3). The risk factors of angiosarcoma include chronic lymphedema, radiotherapy, chemical toxins (including vinyl chloride, thorium dioxide and arsenic), foreign bodies and familial syndromes such as neurofibromatosis (2). Symptoms are nonspecific, including gastrointestinal bleeding, perianal pain and obstructive bowel symptoms (4). Most common location is sigmoid colon and anorectum, followed by cecum and ascending colon (3). Our patient presented abdominal pain and constipation with colon obstruction but did not reveal certain risk factor described above.

Most common histologic type is epithelioid, followed by spindle and mixed type (5). Gross pathology shows hemorrhagic, spongy masses due to intrinsic vascular nature and has indistinct borders (6). Macroscopically most tumors are solid, some are cystic with hemorrhage and necrosis (6). Histology shows epithelioid cells arranged in solid sheets or nests, similar with poorly differentiated carcinoma (5, 6). Immunohistochemistrical study is extremely helpful for diagnosis with endothelial markers of CD31 and ERG (5).

Radiologic finding of colonic angiosarcoma does not much differ from other malignant epithelial tumors except bizarre enhancement and prominent peritumoral vascular structures, representing increased vascularity in vascular origin tumor (7). There is rare case report of colonic angiosarcoma with radiographic findings. A case report showed a large mass in sigmoid colon over 10 cm size with colonic wall thickening, flocculent gas, and central hemorrhage (8). Our case showed about 4 cm size of irregular shaped mass with heterogeneous enhancement in ascending colon with pericolic infiltration but no definite central hemorrhage.

Angiosarcoma shows frequent hematogenous spread, affects lung, liver, and bone (2), with hepatic metastasis in about 18% of cases (3). Most common CT finding of metastatic angiosarcoma in liver is multiple hypoattenuating lesions, with variable findings including peripheral enhancement, cystic lesions with fluid-fluid levels or marked nodular enhancement, similar to that of primary hepatic angiosarcoma (9). There is radiologic–pathologic correlation of peripheral enhancement corresponded to proliferation of tumor cells with central hemorrhagic necrosis. And cystic lesions may show fluid-fluid level on CT and MRI, represents severe hemorrhagic necrosis (9). Liver MR image shows T1 heterogeneous hypointense, T2 hyperintense lesions with peripheral enhancement and fluid-fluid levels (9). In our case, CT and MRI scans revealed diffusely scattered numerous nodules with peripheral and nodular marked hyperenhancement with central hypoattenuation and T2 hyperintensity in liver relatively, consistent with hepatic angiosarcoma. Those lesions did not show internal fluid-fluid level may be attributed to small size not enough to cause severe hemorrhagic necrosis.

Angiosarcoma has a poor prognosis with 5-year survival rate in range of 12% to 35% with median survival of 7 months (1, 2). Prognostic factors with poor outcome include tumor size

larger than 5 cm, old age, metastatic disease at presentation, poor performance status and primary site from viscera and retroperitoneum (2). Mainstay of treatment is complete surgical resection of the tumor. Adjuvant radiation therapy may improve survival and chemotherapy including doxorubicin, paclitaxel and imatinib can be used with unclear actual benefit (3). Recently vascular targeting agents are under study (1). Lesions in liver may benefit from embolization (1). Our patient showed 8 months survival in young age (44-year-old) with tumor size less than 5 cm in ascending colon, underwent extended right hemicolectomy with gross R0 resection followed by paclitaxel after diagnosis of colon epithelioid angiosarcoma.

In conclusion, we described the educational case of extremely rare primary epithelioid colon angiosarcoma with rapidly progressed hepatic metastasis which showed radiologic findings to give a clue to suspect angiosarcoma and make re-diagnosis for proper treatment and prognosis prediction despite of nonspecific radiologic finding of primary lesion.

Author Contributions

Conceptualization, H.S.S.; data curation, L.J., H.S.S., H.J.; formal analysis, L.J., H.S.S., H.J.; funding acquisition, H.S.S.; investigation, all authors; methodology, K.H., J.S.; project administration, H.S.S.; resources, H.S.S., H.J.; software, L.J., H.S.S.; supervision, H.S.S., H.J.; validation, K.H., J.S.; visualization, L.J., H.S.S., J.S.; writing—original draft, L.J., H.S.S., H.J., K.H.; and writing—review & editing, all authors.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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간전이를 동반한 대장 상피모양혈관육종: 증례 보고

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대장 혈관육종은 매우 드문 악성 종양이며 빠르게 진행하고 나쁜 예후를 보인다. 우리는 장폐쇄를 일으킨 대장 상피모양혈관육종의 빠르게 진행한 간전이가 있었던 44세 여자 환자의 사례를 경험하여 이를 보고하고자 한다. 복부 CT에서 불균질하게 조영증강되는 상행결장의 종괴로 인한 근위부 장 확장 소견이 있었고 수술 후 병리적으로 저분화암종으로 보고되었다. 4개월 후 추적 간 MRI에서 테두리 및 결절성의 현저한 동맥기 조영증강을 보이는 수많은 작은 간 결절들이 새로 생겼으며 간 혈관육종이 의심되는 형태로 보였다. 추가 면역조직화학검사 병리적 이차 의견에서 대장 상피모양혈관육종이 진단되었다. 환자는 6개월 후 추적 복부 CT에서 빠른 진행성 간 전이를 보였으며 8개월 후 사망하였다. 빠른 진행을 보이는 혈관육종의 간 전이의 영상 소견을 통해 혈관육종을 의심하고 재진단할 수 있었던 드문 대장 혈관육종의 교훈적인 사례로 생각된다.

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