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Myoid angioendothelioma of the spleen mimicking metastatic disease in a patient with rectal cancer: a radiologic-pathologic correlation ☆

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ABSTRACT

Myoid angioendothelioma of the spleen is an uncommon, benign vascular tumor that is morphologically characterized by a composite of vascular spaces and stromal cells with myoid feature. Herein, we report a case of the myoid angioendothelioma of the spleen, concurrent with rectal adenocarcinoma. A 41-year-old woman presented with hematochezia for several weeks. Grossly, the rectal mass was a 2.5 × 2-cm ulcerative fungating lesion. The splenic mass was a 2.2 × 2-cm well-circumscribed lesion. Microscopically, the rectal mass was a well-differentiated adenocarcinoma that invaded into the pericolic adipose tissue. The splenic mass was composed of slit-like vascular spaces and fascicles of elongated stromal cells. Vascular endothelial cells were immunopositive for CD31, factor VIII–related antigen, and CD34 but negative for CD8. Stromal cells were immunopositive for smooth muscle actin but negative for desmin.

1. Introduction

Vascular neoplasms of the spleen are rare and include hemangioma, littoral cell angioma, lymphangioma, hemangioendothelioma, hemangiopericytoma, and angiosarcoma. The myoid hemangioendiothelioma of the spleen is an extremely rare vascular tumor, characterized histologically by a mixture of vascular endothelial cells and stromal cells displaying myoid/myofibroblastic differentiation. In the literature, only 7 cases of myoid angioendothelioma of the spleen have been reported so far [1-3]. Herein, we present the radiologic and pathologic findings of another rare case of incidental myoid angioendothelioma of the spleen in a patient with concurrent rectal cancer and liver metastasis.

2. Case Report

A 41-year-old woman with a history of hypertension and asthma was admitted to our hospital because of a recent hematochezia. On the physical examination of the rectum, a mass lesion was palpable at 4 cm from the anal verge. Laboratory findings revealed elevated carcinoembryonic antigen (CEA) (69.7 ng/mL, normal range, ~5.0 ng/mL) but normal range of carbohydrate antigen 19-9 (CA 19-9). Contrast-enhanced computed tomographic scan revealed an eccentric thickening in the left wall of the lower rectum. There were several enlarged pararectal lymph nodes as well as a 1.5-cm splenic mass, with irregular low density. 18F-fluorodeoxyglucose positron emission tomography (FDG-PET) revealed hypermetabolic wall thickening of the lower rectum (maximal standard uptake value [SUV], 12.53), focal hypermetabolic nodule of the hepatic segment 5 (maximal SUV, 5.71), and hypermetabolic mass of the spleen (maximal SUV, 4.41) (Fig. 1). For further evaluation of the mass in liver and spleen, a magnetic resonance (MR) imaging was performed with a 1.5-T MR scanner, and we acquired T2-weighted image, precontrast T1-weighted image, and dynamic contrast-enhanced images, which were obtained at the arterial phase, portal phase, and 10-minute–delayed phase after gadolinium administration. The hepatic mass in segment 5 was depicted slightly hyperintense on T2-weighted image (Fig. 2A) and hypointense on precontrast T1-weighted image (Fig. 2B). On dynamic contrast-enhanced MR images, the arterial phase image (Fig. 2C) showed peripheral enhancement with irregular rim appearance and no significant enhanced portion in the central part of the lesion. The portal phase (Fig. 2D) and delayed phase (Fig. 2E) images showed gradually decreased enhancement of the lesion and hypointensity relative to surrounding liver parenchyma. The splenic mass was

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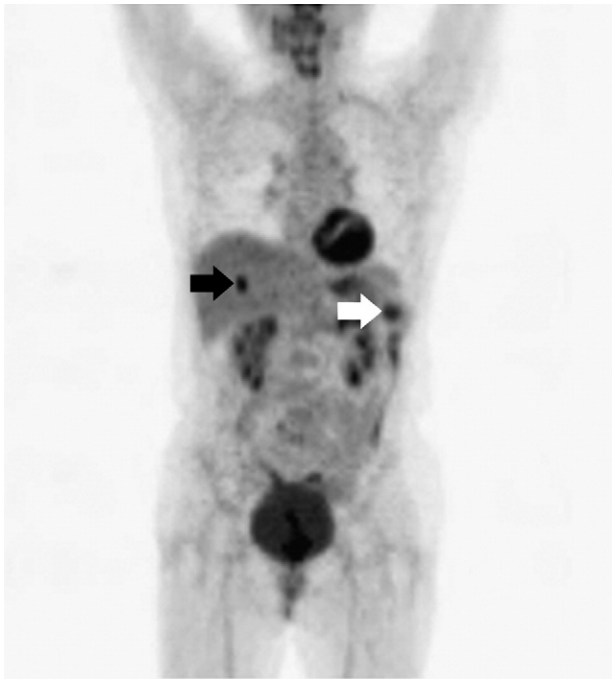

Fig. 1. The FDG-PET reveals hypermetabolic wall thickening of the lower rectum, focal hypermetabolic nodule of the hepatic segment 5 (black arrow), and hypermetabolic mass of the spleen (white arrow).

depicted as a relatively well-defined, round mass with central hyperintensity and peripheral hypointensity on T2-weighted image (Fig. 2F). Precontrast T1-weighted image (Fig. 2G) showed isointense to hypointense area relative to normal spleen parenchyma. Dynamic contrast-enhanced MR images showed initial peripheral rim enhancement on arterial phase (Fig. 2H) and progressive centripetal pattern of enhancement on portal (Fig. 2I) and delayed phase (Fig. 2J). Under the clinical impression of rectal cancer with metastasis to lymph nodes, liver, and spleen, surgery was determined to be the best treatment option. Thus, the patient underwent ultralower anterior resection with lymph node dissection, enucleation of the hepatic mass on segment 5, and splenectomy. Macroscopically, the rectal mass was located at the lower rectum, below the peritoneal reflection, and was a 2.5 × 2-cm ulcerated lesion. Its cut surface revealed a gray-whitish, firm and ulceroinfiltrative lesion that invaded the subserosal adipose tissue. Microscopically, the rectal mass was a well-differentiated adenocarcinoma mainly composed of tubules from the mucosa to the subserosal adipose tissue (Fig. 3A). Of 18 regional lymph nodes, 3 revealed metastatic adenocarcinoma. The hepatic mass was a 0.5 × 0.5-cm, ill-defined, and gray-whitish lesion. The hepatic mass was metastatic adenocarcinoma (Fig. 3B). The splenic mass was a 2.2 × 2-cm, round, firm, well-circumscribed, and yellow-whitish lesion (Fig. 3C). The splenic mass was a solid, well-circumscribed, and capsulated lesion composed of numerous capillary caliber vessels embedded in an eosinophilic matrix of plump stromal cells (Fig. 3D). The vascular channels were slit-like to mildly dilated and lined by a layer of cytologically bland endothelial cells (Fig. 3E). There was no tufting or layering of the endothelium. In between the vascular channels, there was moderate amount of stromal cells arranged either in short fascicles or haphazardly. The stromal cells displayed spindle or polygonal shape, indistinct cell borders, fibrillary cytoplasm, and spindle or somewhat irregular nuclei, with fine chromatin pattern and occasional small nucleoli. Rare stromal cells showed nuclear pseudoinclusions (Fig. 3E, inset). Neither mitoses nor necrosis was found. Scattered inflammatory cells, predominantly lymphocytes, were

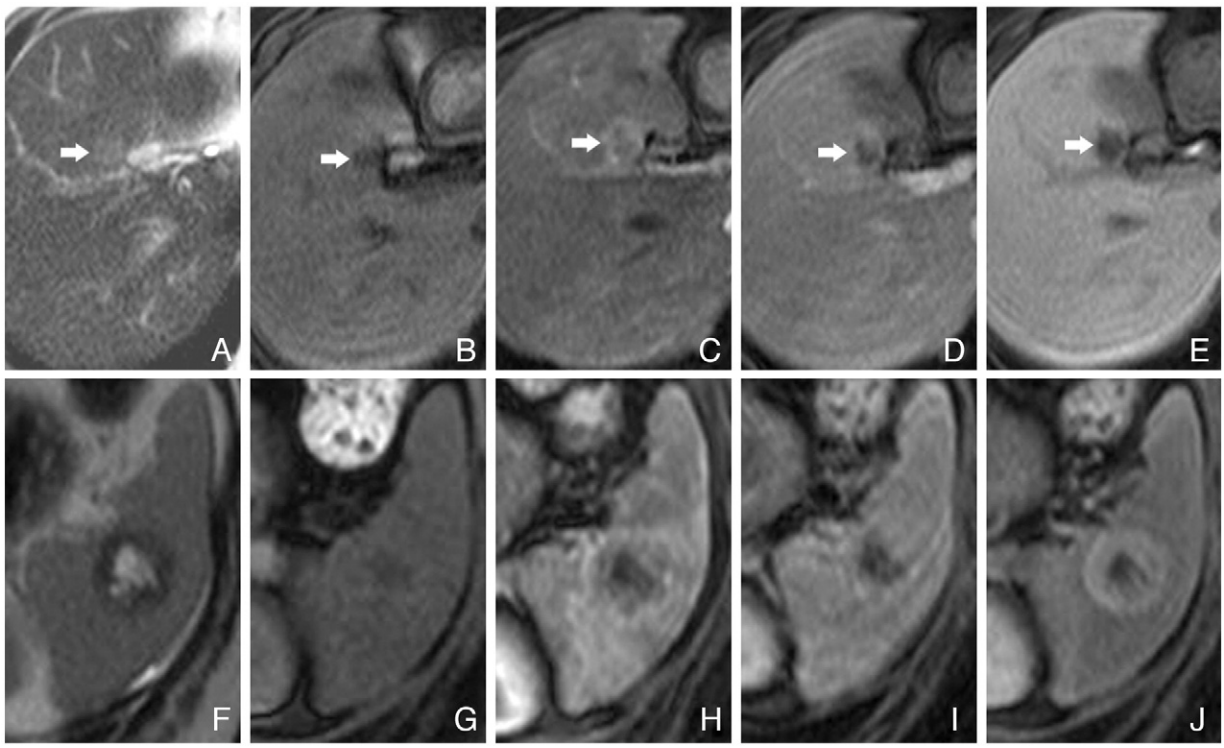

Fig. 2. The hepatic mass in segment 5 (arrow) shows slightly hyperintense relative to liver parenchyma on T2-weighted MR image (A) and hypointense on precontrast T1-weighted image (B). Dynamic contrast-enhanced MR images obtained at the arterial phase (C), portal phase (D), and 10-minute–delayed phase (E) after gadolinium administration. The lesion is depicted as peripheral rim enhancement with low signal in center on arterial phase (C) and slightly decreased enhancement on portal phase (D). On delayed phase (E), the lesion shows marked hypointensity compared with liver parenchyma. The splenic mass shows relatively well-demarcated, round mass with central hyperintensity and peripheral hypointensity on T2-weighted MR image (F). Precontrast T1-weighted image (G) shows isointense to hypointense area relative to spleen parenchyma. Dynamic contrast-enhanced MR images (H-J), which were obtained by the same scan as C to E, show initial peripheral enhancement on arterial phase (H) and progressive centripetal pattern of enhancement on portal (I) and delayed phase (J).