Littoral-cell angioma of the spleen: A case report

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Abstract

Littoral-cell angioma (LCA) is a primary splenic vascular tumor that arises from the normal littoral cells lining the sinus channels of the splenic red pulp. We report a case of LCA of the spleen, which has been infrequently communicated in the literature. A 76-year-old man with a 2-wk history of weight loss, abdominal pain and changes in bowel habits was admitted to our hospital. Imaging studies (CT and MRI) showed multiple lesions in the spleen. Splenectomy was performed. Lining cells were positive for CD31/CD68 markers. Our case was associated with a serrated colonic adenoma. A CT scan showed multiple lesions in the spleen. Pathology was conclusive for serrated adenoma. An enhanced CT scan of the abdomen showed multiple round, hypodense lesions in the spleen. The combination of CT and MRI showed multiple lesions in the spleen. Postoperative pathology examination confirmed the final diagnosis of LCA.

CASE REPORT

A 76-year-old man was admitted to our hospital with a 2-wk history of weakness, weight loss, anorexia, hypogastric abdominal pain and change in bowel habits. His past medical history showed gastritis due to Helicobacter pylori at the age of 63 years and polypectomy due to villous colonic adenoma 2 years before admission. Physical examination was normal. No splenomegaly was found. Results of routine laboratory tests were normal. Colonoscopy identified a polylobulated polyp 3 cm in size in the sigmoid colon, and polypectomy was performed. Pathology was conclusive for serrated adenoma. An enhanced CT scan of the abdomen showed multiple round, hypodense lesions in the spleen. Abdominal MRI revealed multiple splenic hypointense lesions (Figure 1). Our presumptive preoperative diagnosis was lymphoma or hemangioma. Splenectomy was performed. Microscopically, lesions consisted of anastomosing vascular channels with papillary projections and cyst-like spaces. They were lined with endothelial cells that showed hemophagocytosis. Lining cells were positive for both vascular (CD31) and histiomonocytic (CD68) markers. The final histological diagnosis was Littoral-cell angioma.

DISCUSSION

LCA of the spleen may occur at any age (1-77 years; median age, 50 years), with no sex-based predilection (female: male ratio, 5:3)[2,3]. Clinically, patients have splenomegaly, abdominal pain, pyrexia of unknown origin[4] or hypersplenism. LCA may present as an incidental finding. LCA may appear as single or multiple lesions in the spleen. Splenic lesions ranged from 0.2 to 6.0 cm. An extensive list of possibilities such as multiple hemangiomas, lymphoma, metastatic disease, and disseminated infections caused by fungi, mycobacteria, Pneumocystis carinii and sarcoidosis, should be considered in the differential diagnosis of multinodular splenomegaly. CT, MRI, US, and Tc99m RBC scan characteristics have been correlated with histological and immunohistochemical pathological features[5]. Consistent CT features of splenic LCA reported in the medical literature.
literature include low-attenuating lesions on contrast-enhanced images\(^2,6\). Morphological diagnosis is based on the presence of anastomosing vascular channels lined with tall endothelial cells, focal papillary fronds, and normal splenic sinuses at the periphery of the lesion. Combination of morphological and immunohistochemical analyses that show a hybrid endothelial-histiocytic phenotype establish the diagnosis of LCA\(^1,7\). Two case reports have described variants of LCA with histological features of malignancy\(^8,9\).

One third of the previously reported cases were associated with tumors of visceral organs, including colorectal, renal, hepatocellular\(^10\), lung\(^11\), and pancreatic adenocarcinomas, malignant lymphoma\(^13\), myelodysplastic syndrome\(^13\), or aplastic anaemia\(^14\). Since the malignant potential of LCA has not been firmly established in the literature, we recommend close clinical follow-up of patients with LCA of the spleen.

REFERENCES