Littoral cell angiomas of the spleen associated with solid pseudopapillary tumor of the pancreas

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Littoral cell angiomas (LCA) of the spleen are vascular tumors of unknown etiology arising from the littoral cells of the splenic red pulp sinuses. Usually a benign and incidental finding, LCA have been repeatedly reported in association with a variety of visceral malignancies and hold the potential for dissemination per se. We encountered a case of a 30 year old female who was diagnosed with solid pseudopapillary tumor of the head and distal pancreas by fine needle aspiration cytology. A distal pancreatectomy with splenectomy was performed in addition to a pylorus-preserving Whipple's procedure and cholecystectomy. Histopathological examination confirmed solid pseudopapillary tumor of the pancreas and showed multiple well-circumscribed anastomosing vascular channels in the spleen. The diagnosis of LCA of the spleen was confirmed by immunohistochemistry that revealed co-expression of endothelial cell marker, CD31 and CD34, along with histiocytic marker, CD68 by the vascular lining cells. LCA has been previously reported in association with colorectal and pancreatic adenocarcinoma, malignant lymphoma, myelodysplasia and autoimmune disorders. We report the first case of LCA associated with solid pseudopapillary tumor of the pancreas.

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Key words: Littoral cells; Spleen; Vascular tumors; Red pulp

INTRODUCTION

Littoral cell angiomas (LCA) are rare vascular tumors of the spleen of uncertain biological behavior[1]. First described by Falk et al in 1991[2], the majority of LCA are asymptomatic incidental findings with no age or sex predilection[3,4]. Splenomegaly is a common feature of all the LCA and a few of them show symptoms of hypersple-
The unique feature of almost all LCA is its immunohistochemical reactivity to CD31 (endothelial marker) and CD68 (histiocytic marker); the latter suggesting the origin of the tumor as the splenic sinus lining littoral cells. LCA have been associated with a variety of visceral malignancies, including colorectal and pancreatic adenocarcinoma, malignant lymphoma and myelodysplasia. To the best of our knowledge, about 35 cases of LCA have been reported to date in the English literature.

CASE REPORT

A 30 year old female with a history of sickle cell disease, SC trait presented with a 2 d history of gradual onset of back and bilateral lower extremity pain with fever and chills. The patient was diagnosed with solid pseudopapillary neoplasm of the pancreas by endoscopic ultrasonography-guided fine needle aspiration cytology. The diagnosis was confirmed by immunohistochemistry that showed a positive reactivity to CD56, synaptophysin, CD10 and alpha-1 antitrypsin. A CT-scan imaging of the abdomen identified an 11 cm tumor in the distal pancreas and a 2 cm tumor of the head of the pancreas with a bridge of preserved pancreatic tissue between the two tumors (Figure 1). A preoperative angiogram showed the dorsal pancreatic artery supplying the distal tumor and the patient underwent a distal pancreatectomy and splenectomy along with a Whipple’s procedure to prevent the overt diabetes. Gross examination of the pancreas showed a yellow-tan, lobulated, well-circumscribed mass located on the anterior aspect of the pancreatic tail (Figure 2) measuring 13 cm × 10 cm × 7.5 cm and a hemorrhagic, focally cystic red-brown tumor measuring 2 cm × 1.5 cm × 1 cm in the supero-anterior aspect of the pancreatic head. Histopathological examination of the tumor in the head and the distal pancreas revealed morphological changes of solid pseudopapillary tumor. Gross examination of the spleen showed a 113 g yellowish-brown nodular organ measuring 10 cm × 6.5 cm × 3 cm. Two dark-brown, well-circumscribed nodules were identified; one measuring 1.1 × 0.7 × 0.4 cm near to the hilum and other measuring 2.5 × 0.7 × 0.3 cm just underneath the capsule. Histopathological examination showed multiple, anastomosing vascular lesions that vaguely resembled splenic sinusoids lined by tall endothelial cells (Figure 3). The vascular lesions were well delimited from the surrounding splenic parenchyma. Immunohistochemistry revealed the co-expression of CD31 (Figure 4A), CD68 (Figure 4B) and CD34 (Figure 5) by the vascular lining cells, confirming the lesion as LCA of the spleen.

DISCUSSION

Since the identification of LCA by Falk et al in 1991, these vascular tumors have been periodically reported in the literature. Two forms of LCA have been described; the more commonly encountered diffuse multiple nodular form as in our case and the rare solitary form.

The differential diagnosis of splenic neoplasm with a radiological imaging similar to LCA is extensive and includes hemangioma, lymphangioma, hamartoma, hemangiopericytoma, hemangioendothelioma, angiosarcoma, lymphoma, metastasis and sarcoidosis.

Clinically, LCA can present as an abdominal mass, mostly due to splenomegaly, with symptoms of hypersplenism with ensuing anemia and/or thrombocytopenia, pulmonary hypertension and pyrexia of unknown origin.
or can be an incidental finding. However, in our case the patient did not have a splenomegaly associated with solid pseudopapillary tumor of the pancreas. More dramatically, LCA has been reported to present as splenic rupture and hemoperitoneum[12,13].

Radiological studies by CT scan, MRI, sonography or nuclear medicine studies, although not conclusive[14], can contribute to diagnosing LCA. A CT-scan imaging shows LCA as hypoattenuating nodules of varying size. Delayed phase imaging on CT-scan reveals the nodules to be isodense to surrounding splenic parenchyma due to delayed filling of the nodules. MRI of the spleen shows hypodense lesions on T1 and T2 weighted scan due to the hemosiderin content of the tumor[15]. However, no hypodense nodules in the spleen were evident on CT-scan imaging in our case. Sonography is rarely helpful as findings vary greatly from isoechoic to hypo- and hyper-echoic lesions[15]. Tc-99m labeled RBC scintigraphy can differentiate splenic lesions from splenic hemangiomas[16].

The pathogenesis of LCA remains unclear but, given its association with autoimmune disorders such as Crohn’s disease and inborn metabolic diseases such as Gaucher’s disease, immune system dysfunction has been postulated as a possible pathogenic mechanism[17,18]. Supporting this hypothesis, other reports have suggested that chronic infection and systemic immunosuppression may contribute to the development of LCA[12,19]. Interestingly, once thought of as a benign and incidental lesion, one third of the reported cases are associated with malignancies of visceral organs including adenocarcinoma of colorectum (most

Figure 3  Histopathology of splenic nodule showing proliferation of spindle cells with anastomosing vascular channels and congestion of large vessels suggestive of LCA (Hematoxylin and Eosin stain). A: ×40; B: ×100.

Figure 4  Immunohistochemistry of splenic nodule showing vascular lining cells reactive to CD31 (A) and CD68 (B), ×100.

Figure 5  Immunohistochemistry of splenic nodule showing vascular lining cells reactive to CD34. A: ×40; B: ×100.
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