Answer

Littoral Cell Angioma

Littoral cell angioma, a rare primary vascular tumor of the spleen, was first described by Falk et al[1] in 1991. This neoplasm is believed to arise from the endothelial cells that line the red pulp sinuses (littoral cells) because it has characteristic morphologic and immunophenotypic features that demonstrate dual endothelial/histiocytic differentiation. Littoral cell angiomas occur over a wide age range (median age, 49 years) and there is no gender predilection. Patients may be asymptomatic or present with pain or fever of unknown origin. Splenomegaly is almost always present. Hypersplenism-related anemia or thrombocytopenia is occasionally seen.

Littoral cell angiomas typically consist of single or, more often, multiple cystic lesions well delineated from surrounding splenic tissue but without a definite capsule. Microscopically, these lesions are characterized by dilated anastomosing vascular channels lined with polygonal endothelial cells. Papillary fronds protrude within the vascular channels. Exfoliated histiocytic cells are frequently found in the lumina. The lining cells exhibit no significant nuclear pleomorphism or mitotic activity. Additional features may include intracytoplasmic and/or extracytoplasmic hemosiderin, extramedullary hematopoiesis, and calcification. The diagnosis can be validated by immunostaining that indicates the lining cells are positive for endothelial marker (factor VIII–related antigen) and histiocytic markers (CD68 and lysozyme).

Levy et al[2] analyzed computed tomographic features in 8 cases of littoral cell angioma and found that the presence of numerous hypoattenuating splenic masses is the most consistent finding. On delayed contrast–enhanced images, the lesions may become isoattenuating with the adjacent spleen tissue. The differential diagnosis for multiple hypoattenuating splenic lesions includes many neoplastic and nonneoplastic disorders. Adenopathy or disease in other organs, which is usually seen in patients with splenic metastasis and lymphoma, is not present in patients with littoral cell angioma. The nonneoplastic disorders that may have a similar computed tomographic appearance include infection and systemic diseases such as sarcoidosis. The patient's clinical condition will help to distinguish these disorders from littoral cell angioma.

Although littoral cell angiomas were originally thought to be benign, littoral cell angioma with malignant features has been reported.[3] Furthermore, a relationship between littoral cell angioma and visceral malignancies has been observed. The associated malignancies include colorectal, renal, and pancreatic adenocarcinoma and lymphoma.[4] It has been postulated that littoral cell angioma reflects an altered immune state or conditions requiring increased phagocytic function.[1] Close follow-up and careful investigation in search of a second neoplasm have been recommended. In the patient described, concomitant hepatitis B virus infection and chronic urticaria may represent a putative association or can be entirely incidental. The patient remains asymptomatic after 1 year of follow-up.

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REFERENCES

1. Falk S, Stutte HJ, Frizzera G. Littoral cell angioma: a novel splenic vascular lesion demonstrating histiocytic differentiation. *Am J Surg Pathol*. 1991;15:1023-1033.
2. Levy AD, Abbott RM, Abbondanzo SL. Littoral cell angioma of the spleen: CT features with clinicopathologic comparison. *Radiology*. 2004;230:485-490.
3. Ben-Izhak O, Bejar J, Ben-Eliezer S, Vlodavsky E. Splenic littoral cell haemangioendothelioma: a new low-grade variant of malignant littoral cell tumour. *Histopathology*. 2001;39:469-475.
4. Chatelain D, Bonte H, Guillevin L, Balladur P, Flejou JF. Small solitary littoral cell angioma associated with splenic marginal zone lymphoma and villous lymphocyte leukaemia in a patient with hepatitis C infection. *Histopathology*. 2002;41:473-475.