Online Submissions: wjg.wjgnet.com
www.wjgnet.com
wjg@wjgnet.com

World J Gastroenterol 2007 December 28; 13(48): 6603-6604
World Journal of Gastroenterology ISSN 1007-9327

CASE REPORT

Littoral-cell angioma of the spleen: A case report

Ángel Cosme, Ángel Tejada, Luis Bujanda, Manuel Vaquero, José Luis Elorza, Evelia Ojeda, Unai Goikoetxea

Ángel Cosme, Ángel Tejada, Luis Bujanda, Unai Goikoetxea, Department of Gastroenterology, Hospital Donostia, Spain
Manuel Vaquero, Department of Pathology, Hospital Donostia, Spain
José Luis Elorza, Departamet of Surgery, Hospital Donostia, Spain
Evelia Ojeda, Department of Internal Medicine, Hospital Donostia, Spain
Correspondence to: Ángel Cosme, Department of Gastroenterology, Hospital Donostia, Paseo Beguiristain, s/n, 20014, San Sebastián (Gipuzkoa), Spain. acosme@chdo.osakidetza.net
Telephone: +34-985-943003463 Fax: +34-985-943007065
Received: July 6, 2007 Revised: October 1, 2007

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. LCA is a benign vascular tumor of the spleen that needs to be included in the differential diagnosis of multiple splenic nodules.

Key words: Littoral-cell angioma; Spleen

Cosme Á, Tejada Á, Bujanda L, Vaquero M, Elorza JL, Ojeda E, Goikoetxea U. Littoral-cell angioma of the spleen: A case report. *World J Gastroenterol* 2007; 13(48): 6603-6604

http://www.wjgnet.com/1007-9327/13/6603.asp

INTRODUCTION

Littoral-cell angioma (LCA) is a rare primary tumor of the spleen that was first described by Falk *et al* in 1991[1]. Considered a benign condition, this neoplasm arises from the red cell pulp sinuses and has intermediate features between those of endothelial and histiocytic cells. To the best of our knowledge, there have been 11 cases of LCA reported in the English-language literature since the description of this disease in 1991[2]. We report a case of LCA in a patient who had symptoms of weakness, pain and change in bowel habits. 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 gastrectomy due to peptic ulcer at the age of 63 years and polypectomy of 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 $\geqslant$ 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 hypointensive 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 (Figure 2), but CD8 and CD3 were negative. Ten months after surgery, the patient was asymptomatic.

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

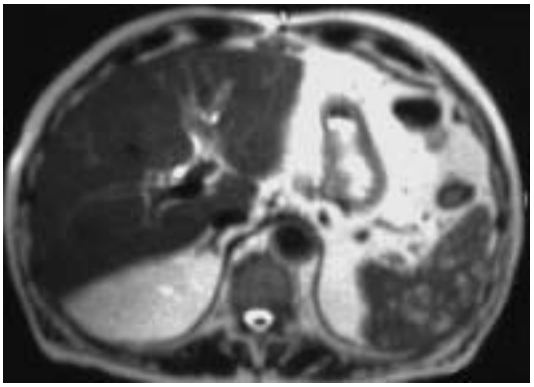

Figure 1 MRI: multiple hypointense lesions in the spleen (T_2).

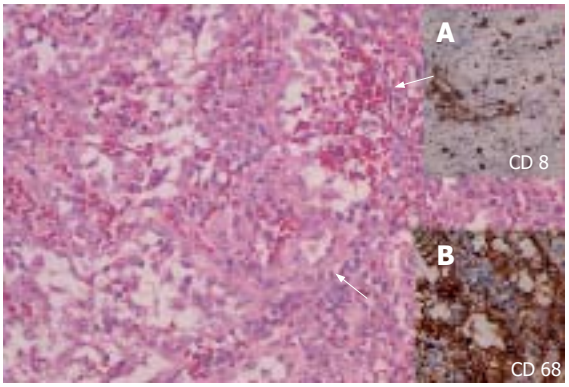

Figure 2 Vascular proliferation, well established (arrows), and very similar to normal spleen sinusoids. **A**: Negative for CD8; **B**: Positive for histiomonocytic markers.

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-histiocitic 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[12], 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

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 **Najera L**, Dotor AM, Santoja C. Littoral cell angioma of the spleen. A case report and review of the literature. *Rev Esp Patol* 2006; **39**: 49-53
4 **Tan YM**, Chuah KL, Wong WK. Littoral cell angioma of the spleen. *Ann Acad Med Singapore* 2004; **33**: 524-526
5 **Johnson C**, Goyal M, Kim B, Wasdahl D, Nazinitsky K. Littoral cell angioma. *Clin Imaging* 2007; **31**: 27-31
6 **Goldfeld M**, Cohen I, Loberant N, Mugrabi A, Katz I, Papura S, Noi I. Littoral cell angioma of the spleen: appearance on sonography and CT. *J Clin Ultrasound* 2002; **30**: 510-513
7 **Dascalescu CM**, Wendum D, Gorin NC. Littoral-cell angioma as a cause of splenomegaly. *N Engl J Med* 2001; **345**: 772-773
8 **Rosso R**, Paulli M, Gianelli U, Boveri E, Stella G, Magrini U. Littoral cell angiosarcoma of the spleen. Case report with immunohistochemical and ultrastructural analysis. *Am J Surg Pathol* 1995; **19**: 1203-1208
9 **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
10 **Lin CH**, Yu JC, Shih ML, Peng YJ, Hsieh CB. Littoral cell angioma of the spleen in a patient with hepatocellular carcinoma. *J Formos Med Assoc* 2005; **104**: 282-285
11 **Collins GL**, Morgan MB, Taylor FM 3rd. Littoral cell angiomatosis with poorly differentiated adenocarcinoma of the lung. *Ann Diagn Pathol* 2003; **7**: 54-59
12 **Bisceglia M**, Sickel JZ, Giangaspero F, Gomes V, Amini M, Michal M. Littoral cell angioma of the spleen: an additional report of four cases with emphasis on the association with visceral organ cancers. *Tumori* 1998; **84**: 595-599
13 **Erçin C**, Gürbüz Y, Hacihanefioğlu A, Turgut Karakaya A. Multiple littoral cell angioma of the spleen in a case of myelodysplastic syndrome. *Hematology* 2005; **10**: 141-144
14 **Tholouli E**, Roulson JA, Byers R, Burton I, Liu Yin JA. Littoral cell angioma of the spleen in a patient with severe aplastic anaemia. *Haematologica* 2003; **88**: ECR33

S- Editor Liu Y **L- Editor** Kerr C **E- Editor** Yin DH