CASE REPORT

Littoral cell angioma in a splenunculus: a case report

N VENKATANARASIMHA, MBBS, MRCP, FRCR, S HALL, MBBS, P SURESH, MRCP, FRCR and M P WILLIAMS, FRCR

Department of Radiology, Derriford Hospital, Plymouth PL6 8DH, UK

ABSTRACT. Littoral cell angioma (LCA) is a rare primary splenic vascular tumour that arises from the littoral cells that line the red pulp sinuses. It is usually asymptomatic and is discovered incidentally on imaging for other pathologies. Radiologists should be aware of these lesions as they may be mistaken for malignant lesions and lead to unnecessary surgery. We present a case of LCA recurrence within a splenunculus that was discovered incidentally in a 60-year-old patient being investigated for right upper quadrant pain.

Received 6 September 2009
Revised 22 November 2009
Accepted 25 November 2010

DOI: 10.1259/bjr/60430925

© 2011 The British Institute of Radiology

Littoral cell angioma (LCA) is a rare tumour of the spleen that was described first in 1991. Its imaging features in the spleen are well described, but this is first case report of a recurrence of this condition in a previously normal splenunculus and its appearances on ultrasound, CT and MRI.

Case report

A 60-year-old male was investigated for haematuria. Ultrasound demonstrated normal renal tracts, but incidentally showed multiple hypoechoic nodules within a normally sized spleen. CT was performed for further evaluation and demonstrated multiple low hypoattenuating lesions within the spleen (Figure 1). After an initial non-diagnostic CT-guided splenic biopsy, the patient underwent splenectomy. This was complicated by pancreatitis and portal vein thrombosis seen on the post-operative CT, which also demonstrated a homogeneously enhancing 2 cm splenunculus (Figure 2). Histopathology of the spleen showed LCA.

Seven years later the patient represented with cholestatic jaundice caused by gallstones. Ultrasound (Figure 3) and CT (Figure 4) performed at the second admission again showed chronic portal vein thrombosis with associated collateral vessels. These imaging modalities also demonstrated a 6 cm splenunculus with multiple low density lesions that were identical in pattern to the lesions seen in the native spleen.

MRI demonstrated the lesions within the splenunculus as well-defined nodules that were hypointense on T_1 weighted (Figure 5) and hyperintense on T_2 weighted images (Figure 6). Dynamic post-gadolinium images showed no significant contrast enhancement of the lesions.

The identical imaging appearances of the lesions within the splenic tissue, and the fact that these were incidental findings at each presentation, helped us to conclude that the lesions in the splenunculus were likely to represent a recurrence of LCA. The MRI appearances further supported this diagnosis as the signal characteristics of the lesions were consistent with LCA as described in the literature. A 6 month follow-up ultrasound examination showed no change in the appearances of the lesions within the splenunculus.

Discussion

Primary vascular tumours of the spleen are uncommon and mostly haemangiomas. Littoral cell angioma of the spleen is a rare vascular tumour originally described in 1991 by Falk et al [1] and is unique to the spleen. This benign tumour arises from the specialised lining cells of the venous sinuses of the red pulp known as “littoral cells”, which expresses both histiocytic and vascular endothelial antigens.

The exact incidence of LCA is unknown. There is no gender or age predilection although the median patient age reported is 49 years [1]. Patients are usually asymptomatic and the tumour is discovered incidentally at imaging for other pathologies. Occasionally, patients present with pain, pyrexia of unknown origin, splenomegaly and symptoms or laboratory evidence of

Address correspondence to: N Venkatanarasimha, Level 6, Derriford Hospital, Derriford Road, Plymouth PL6 8DH, UK. E-mail: nandashettykv@yahoo.com

Figure 1. Contrast-enhanced portal venous phase CT at initial presentation shows multiple hypodense lesions of various sizes (arrows) within the spleen.

hypersplenism [2]. The vast majority of cases of LCA described in the literature are benign, but a few cases of malignancy have been reported [3].

Radiologically, LCA usually presents as multiple nodules within the spleen [4] and rarely as a solitary mass [5]. The most common CT manifestation of LCA is splenomegaly with innumerable masses [4]. The lesions appear as hypodense nodules of varying size, ranging from 0.2 to 6 cm, and demonstrate contrast enhancement on the portal venous phase becoming isodense to the surrounding splenic parenchyma on delayed-phase images [6].

On MRI, the lesions typically appear isointense to slightly hypointense on T_1 weighted images and hyperintense on T_2 weighted images [2]. The ultrasound

Figure 2. Contrast-enhanced portal venous phase CT performed post-splenectomy shows a homogeneously enhancing splenunculus (arrow) without any focal lesion.

Figure 3. At second presentation, transabdominal sonography demonstrates an enlarged splenunculus with several well-defined hypoechoic nodules (arrow) of varying sizes. These demonstrated no internal vascularity.

appearance of LCA is again non-specific and includes both heterogeneous echotexture without any definite nodules and well-defined nodules that are hypoechoic (as in our case) or hyperechoic [3]. Colour Doppler assessment can demonstrate both central and peripheral vascularity that helps to distinguish LCA from cavernous haemangiomas [7].

At pathology, the gross specimen of spleen shows a nodule or nodules with blood or blood products varying in colour from dark red or brown to black, depending on the chronicity of the blood. Microscopically, LCA is characterised by the presence of vascular channels that

Figure 4. Contrast-enhanced portal venous phase CT performed at the second presentation shows an enlarged splenunculus with multiple hypoattenuating lesions (arrows) of varying sizes. In addition there are extensive varicose vessels around the gallbladder (arrowheads) and in the greater omentum, which are in keeping with portosystemic shunts occurring as a consequence of portal vein thrombosis.

Figure 5. T_1 weighted VIBE (volumetric interpolated breath-hold sequence) sequence (repetition time (TR)/echo time (TE); 8 ms/5 ms; slice thickness 3 mm; matrix size 192×256). The nodules show hypointensity (arrows).

are lined by tall endothelial cells and papillary fronds extending into the vascular channels [1, 4].

Although several reports have described the imaging appearances of LCA, they all are non-specific, requiring pathology for a definitive diagnosis. The differential for multiple hypodense splenic lesions on CT includes both neoplastic and inflammatory conditions. These include splenic haemangiomas, lymphangiomas, hamartomas, angiosarcomas, metastases, lymphoma and infectious processes like pneumocystis, fungal infections and atypical bacterial infections, which can all have similar appearances on CT [6].

Conclusion

Littoral cell angioma should be considered in the differential diagnosis of multiple lesions within the spleen. In the above case report, lesions with similar imaging appearances within the splenunculus to those of LCA of the native spleen alerted the radiologist to a recurrence. A repeat biopsy was avoided in view of the multimodality imaging appearances of LCA.

Figure 6. Axial T_2 weighted HASTE (half fourier acquisition single shot turbo spin echo) sequence (repetition time (TR)/echo time (TE); 1000 ms/86 ms; slice thickness 4 mm; matrix size 224×256). The nodules show hyperintensity (arrows).

References

1. Falk S, Stutte HJ, Frizzera G. Littoral cell angioma — a novel splenic vascular lesion demonstrating histiocytic differentiation. *Am J Surg Pathol* 1991;15:1023–33.
2. Español I, Lerma E, Fumanal V, Palmer J, Roca M, Domingo-Albós A, et al. Littoral cell angioma with severe thrombocytopenia. *Ann Haematol* 2000;79:46–9.
3. Abbott RM, Levy AD, Aguilera NS, Gorospe L, Thompson WM. From the archives of the AFIP: primary vascular neoplasms of the spleen radiologic–pathologic correlation. *Radiographics* 2004;24:1137–63.
4. Levy AD, Abbott RM, Abbondanzo SL. Littoral cell angiona of the spleen: CT features with clinicopathologic comparison. *Radiology* 2004;230:485–90.
5. Arber DA, Strickler JG, Chen YY, Weiss LM. Splenic vascular tumors: a histologic, immunophenotypic, and virologic study. *Am J Surg Pathol* 1997;21:827–35.
6. Kinoshita LL, Yee J, Nash SR. Littoral cell angioma of the spleen: imaging features. *AJR* 2000;174:467–9.
7. Bhatt S, Simon R, Dogra VS. Littoral cell angioma. Sonographic and color Doppler features. *J Ultrasound Med* 2007;26:539–42.