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CASE REPORT

Pediatric littoral cell angioma of the spleen: multimodality imaging including diffusion-weighted imaging

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Abstract Littoral cell angioma (LCA) is a rare primary splenic vascular tumor originating from littoral cells lining the splenic red pulp sinuses. LCAs are rarely seen in children. We present the US, CT, and MRI findings including diffusion-weighted imaging (DWI) in a 2-year-old boy with histologically proven LCA. Previous studies on liver lesions have shown that DWI allows differentiation of vascular tumors from primary neoplasms and metastatic disease. The current case indicates that increased ADC values within the splenic lesions suggest a vascular etiology, which might help narrow the differential diagnosis.

Keywords Littoral cell angioma · Spleen · Child · Diffusion-weighted imaging · ADC

Introduction

Littoral cell angioma (LCA) is a rare primary splenic vascular tumor that originates from littoral cells lining the splenic red pulp sinuses. LCA was first described by Falk et al. [1, 2] in 1991. LCA is typically diagnosed in adults (mean age 48 years), rarely in children [1]. LCA might present with signs related to hypersplenism such as thrombocytopenia, anemia and splenomegaly or with constitutional symptoms such as fever and abdominal pain, or as an incidental finding [3]. LCAs are primarily benign tumors; malignant transformation has, however, been reported [4]. Imaging findings are nonspecific; differential diagnoses might include vascular malformations, infectious or metastatic disease [5]. We report a 2-year-old boy who presented with thrombocytopenia and coagulopathy caused by a splenic LCA. US, CT, and MRI findings are presented and correlated with diffusion-weighted imaging (DWI).

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Case report

A 2-year-old boy presented to an outside hospital because of a mild right frontal swelling that occurred after minor head trauma (the boy had hit his head on a chair in day-care). Subsequent head CT showed a small subgaleal hemorrhage without focal intracranial lesion. Neurological examination was unremarkable and after the routine survey period the child was discharged home. The parents returned, however, to the emergency department of our hospital because the swelling increased significantly overnight. Clinical examination revealed a large subgaleal hematoma. Neurological examination was again unremarkable. Physical examination revealed profound splenomegaly. Hematological tests showed significant anemia (hemoglobin 6.4 g/dl), MCV 75 fl, reticulocyte count 6%, thrombocytopenia, low fibrinogen (99 mg/dl), elevated D-dimers (16.73 mg/l) and low platelets. Abdominal US revealed a significantly enlarged, heterogeneous spleen with several round, ill-defined, heterogeneously hypoechoic

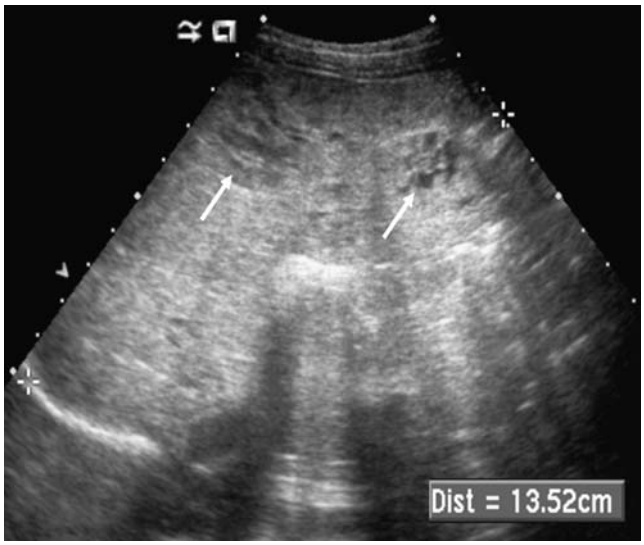

Fig. 1 US image shows marked splenomegaly, a few round, ill-defined, heterogeneously hypoechoic foci (*arrows*) within diffuse heterogeneous splenic parenchyma

focal lesions (Fig. 1). No lymphadenopathy or other focal lesions were visible.

For further characterization, a multiphase contrast-enhanced CT scan was performed. In addition to the splenomegaly, multiple round hypodense (on arterial phase) lesions of various sizes were seen throughout the spleen (Fig. 2). These lesions showed mild progressive contrast enhancement on early and late venous-phase imaging. No additional lesions were seen in other abdominal solid organs. The most likely diagnosis was believed to be multiple splenic hemangiomas. Infectious disease, lymphoma or metastatic disease seemed unlikely. On MRI, the lesions appeared well-circumscribed, predominantly T1-hypointense and T2-hyperintense (Fig. 3), with mild progressive contrast enhancement on dynamic imaging (Fig. 3). Gradual contrast enhancement resulted in near complete filling of the lesions on delayed venous-phase imaging (Fig. 3). The lesions were hypointense on isotropic DW images (Fig. 3). Apparent diffusion coefficient (ADC) maps showed an increased diffusion of the lesions compared to the normal-appearing splenic tissue (Fig. 3). ADC measurements were performed in five representative lesions. The mean ADC value was $2{,}765.6\times10^{-6}$ mm^2/s (standard deviation 460×10^{-6} mm^2/s). The mean ADC value of the surrounding normal-appearing splenic parenchyma was 874.6×10^{-6} (standard deviation 11.4×10^{-6} mm^2/s). Our presumptive diagnosis was hemangiomatosis. Lymphangiomatosis was less likely given the gradual contrast enhancement and filling in of some of the lesions. The anemia and thrombocytopenia were thought to be secondary to consumptive coagulopathy and active bleeding.

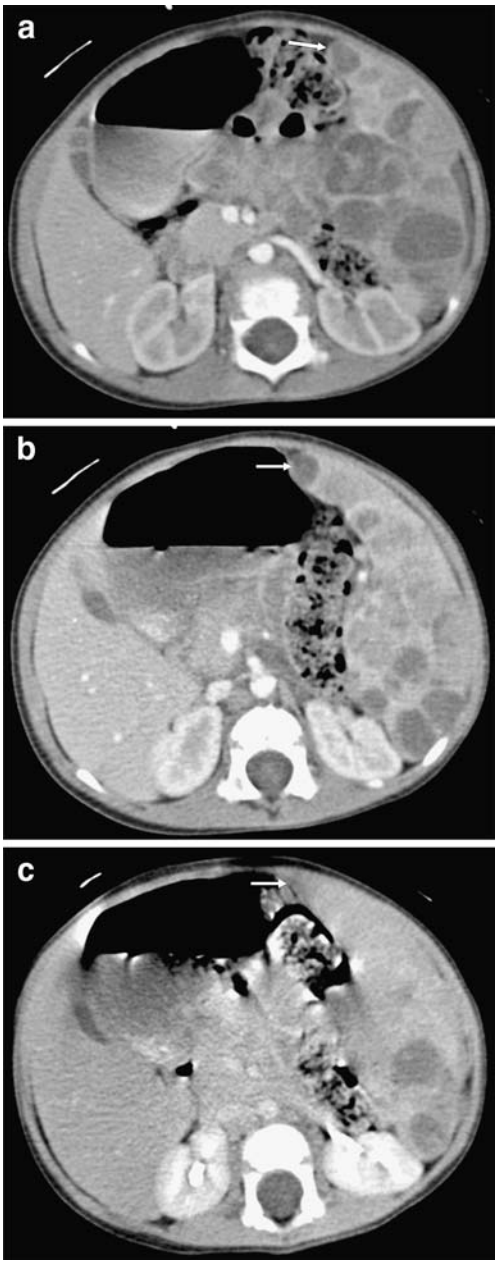

Fig. 2 Axial CT images of the spleen show multiple round lesions of various sizes throughout the spleen. Gradual contrast enhancement of the lesions are nicely depicted (*arrows*) on the same cut of the spleen on the early arterial phase image (**a**), the early venous phase image (**b**), and the late venous phase image (**c**)

With the exception of supportive treatment for the coagulopathy, no medical therapy was attempted for the splenic lesions. Endovascular embolization of the spleen was decided upon to increase the platelet count. Selective celiac angiography demonstrated splenomegaly with multiple splenic parenchymal defects consistent with multiple splenic lesions (Fig. 4). Approximately 50% of the spleen was targeted for ablation. With the catheter engaged in the celiac artery, a Progreat microcatheter (Terumo Interven-

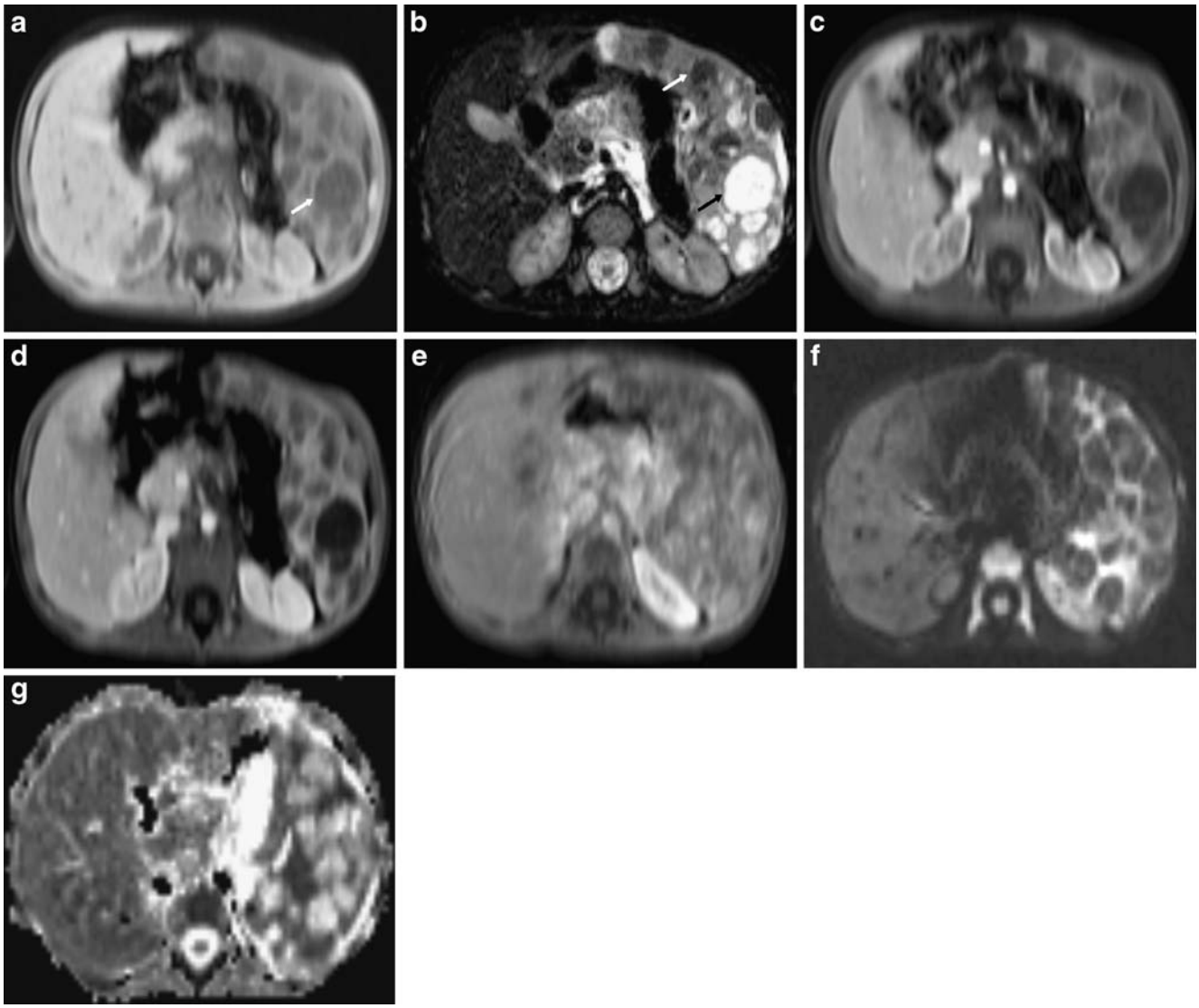

Fig. 3 MRI. **a** Axial T1-W image demonstrates well-demarcated hypointense lesions (*arrow*). **b** The lesions are predominantly hyperintense (*black arrow)* on axial T2-W images. Few of these lesions are hypointense (*white arrow)* because of hemosiderin deposition. **c–e** These lesions show mild progressive contrast enhancement on dynamic gadolinium-enhanced MR images in the early phase (**c**), the late venous phase (**d**), and the delayed phase (**e**). **f** The lesions are hypointense on the isotropic DW image. **g** ADC map shows an increased diffusion of the lesions compared to the normal-appearing splenic tissue

tional Systems, Somerset, NJ) and wire were used to select the inferior segmental branches of the splenic artery accounting for supply to the inferior 50% of the spleen. In each of these positions, particle embolization with 300–500 μm embospheres was performed until stasis of the target vessels was achieved. This was done under fluoroscopy and without evidence of reflux. Postembolization angiography demonstrated absent parenchymal blush of the inferior pole of the spleen (Fig. 4). The platelet count gradually normalized after embolization, with fibrinogen remaining low and D-dimers elevated. Subsequent review of the angiography study, the cross-sectional imaging results and the clinical history with our vascular anomalies team (S.M.) and an outside vascular anomalies expert (Dr. Patricia Burrows) suggested the diagnosis of LCA. Because of the continued coagulopathy, splenectomy was performed. Histopathological examination confirmed splenic LCA. Areas of necrosis were seen within the spleen related to prior embolization. The child's coagulation normalized and he was discharged from hospital 4 days after splenectomy.

Discussion

LCA is a rare primary splenic vascular tumor that originates from the splenic littoral cells. The term "littoral" is derived from the Latin noun *littoris*, meaning shore. Littoral cells

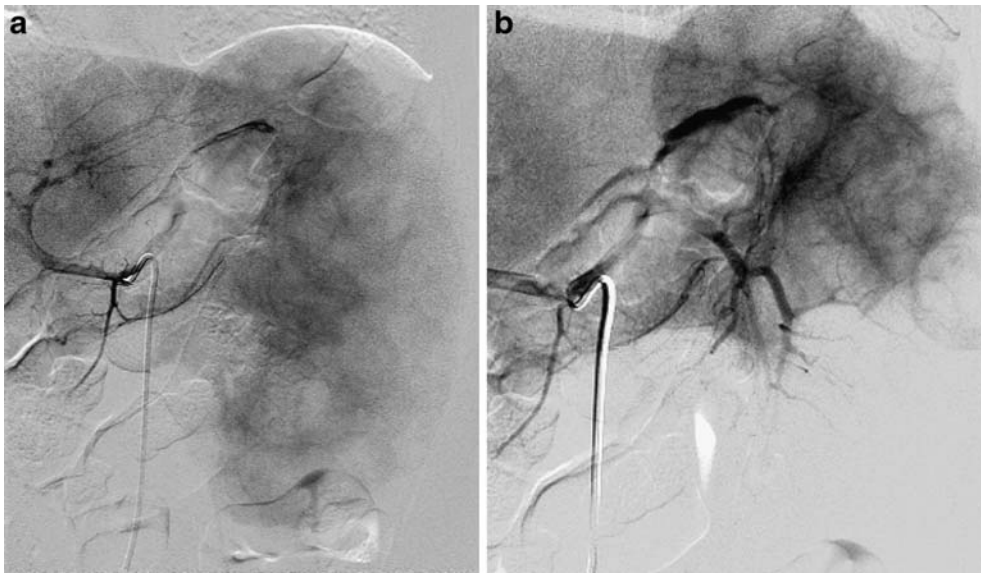

Fig. 4 Angiography. **a** Selective splenic angiogram reveals a significantly enlarged spleen with multiple filling defects. **b** After embolization the inferior half of the spleen is devascularized

typically line the splenic red pulp sinuses. Gross pathology often reveals a large spleen with multiple nodular lesions. The histopathological features of LCA are based on the presence of anastomosing vascular channels of variable size lined with flat and tall endothelial cells, focal papillary fronds extended into the vascular channels and normal splenic sinuses at the periphery of the lesions in the splenic red pulp. Immunohistochemical staining for factor 8, CD31 and CD68 are characteristic of LCA that show both endothelial and histiocytic cells. Cellular atypia is typically absent [1, 2]. Although there is no age predilection, LCA usually occurs in adults and appears to be extremely rare in children. Only two pediatric cases have been reported. Falk et al. [1] included one patient in their initial description of 17 patients with LCA. Anton-Pacheco et al. [6] described one additional case of LCA, in a 1-year-old girl. There appears to be no gender predilection**.** Clinically, patients might present with symptoms of hypersplenism including anemia, thrombocytopenia and splenomegaly. However, most frequently patients are asymptomatic and LCA is discovered incidentally [3]. In our patient the enlarging subgaleal hematoma initiated work-up of the child that revealed a coagulopathy and hypersplenism leading us to the diagnosis of LCA.

Imaging findings including US, CT and MRI have been described in several articles [2, 5–7]. Our patient showed a significantly enlarged spleen with multiple rounded, focal lesions that showed a delayed enhancement/filling in of the lesions. Contrast-enhanced imaging with either CT or MRI showed the multiplicity of the lesions in better detail than US. The imaging findings on US, CT and MRI are unfortunately nonspecific. Differential diagnosis is broad and includes vascular malformations such as hemangiomatosis and lymphangiomatosis, vascular tumors such as hemangioendothelioma and hemangiopericytoma, infectious processes such as tuberculosis and fungal disease, as well as malignant processes such as lymphoma and metastatic disease. Currently, final diagnosis is only possible by histopathological examination. Bhatt et al. [2] reported that MRI can be especially helpful in the diagnosis of LCA by showing T1- and T2-hypointense hemosiderin deposition within the angiomas. In our patient, the majority of lesions were T1-hypointense and T2-hyperintense; however, a smaller percentage were T1- and T2-hypointense, indicating hemosiderin deposition. Our patient showed increased ADC values measured within the lesions compared to the normal-appearing surrounding splenic parenchyma. These data allow the differential diagnosis to be narrowed.

A previous study on liver lesions has shown that DWI allows differentiation of vascular tumors from primary hepatic neoplasms and metastatic disease [8]. Hepatic hemangiomas have been reported to reveal increased ADC values compared to normal liver parenchyma. Despite the fact that the diffusion characteristics of the spleen cannot directly be compared with those of the liver, our case indicates that increased ADC values within the splenic lesions suggest a vascular etiology/lesion [8]. Splenic abscesses, fungal infections and solid splenic tumors including metastatic disease become less likely, as these lesions typically show reduced ADC values. The measured ADC values within the splenic LCA lesions were slightly higher, as in previously reported liver hemangiomas. The exact reason for this difference in ADC values is unclear. We can only suggest that this can possibly be explained by the fact that the liver is a very different organ from the spleen and, secondly, the ADC measurements of the liver hemangiomas in this study were performed in adult patients.

In conclusion, MRI in combination with DWI and ADC analysis might narrow the differential diagnosis of multifocal splenic lesions. Further studies incorporating larger patient groups are necessary; our case, however, indicates that the inclusion of a DWI/ADC sequence/analysis should be considered in the diagnostic work-up of patients presenting with multifocal splenic lesions.

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