Radiological features of a
symptomatic splenic hamartoma

S. E. Thompson
E. A. Walsh
B. C. Cramer
C. C. Pushpanathan
P. Hollett
L. Ingram
D. Price

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S. E. Thompson · E. A. Walsh (✉)
B. C. Cramer
Department of Radiology,
Janeway Child Health Centre and
Memorial University of Newfoundland,
Janeway Place, St. John's, NF A1A 1R8, Canada

C. C. Pushpanathan
Department of Pathology,
Janeway Child Health Centre and
Memorial University of Newfoundland,
St. John's, NF, Canada

P. Hollett
Department of Nuclear Medicine,
Health Sciences Centre and
Memorial University of Newfoundland,
St. John's, NF, Canada

L. Ingram
Department of Pediatrics,
Janeway Child Health Centre and
Memorial University of Newfoundland,
St. John's, NF, Canada

D. Price
Department of Surgery,
Janeway Child Health Centre and
Memorial University of Newfoundland,
St. John's, NF, Canada

Abstract Symptomatic splenic hamartomas are rare in the pediatric age group, with only four previous reports in the literature. Splenic hamartoma has been reported as a solid homogeneous mass without calcification on CT and ultrasound (US), and only one previous report of the findings on MRI has been published. We report a case of a large symptomatic splenic hamartoma in a 14-year-old girl who presented with splenomegaly, pancytopenia and growth retardation. A solid mass with multiple punctate foci resembling calcifications was seen on US. The mass was heterogeneous and better demarcated on enhanced CT. Radiocolloid scintigraphy demonstrated uptake within the lesion, but less than that of normal spleen. The mass was isointense relative to normal splenic tissue on T1-weighted MRI (0.5 T) and of increased intensity with T2 weighting. At splenectomy, a red pulp hamartoma was identified, which contained nodules of hyalinization and necrosis thought to account for the punctate foci seen on US.

Introduction

Splenic hamartomas (SH) are rare benign tumors with an autopsy incidence of only 0.13 % [1]. They are usually < 3 cm [2] in size, asymptomatic and discovered incidentally at surgery or autopsy [1]. Symptomatic splenic hamartomas (SSH) are extremely rare, particularly in the pediatric age group. Reports of the radiological appearances, especially that of MRI, are therefore limited [3]. We present a case of a large SSH in a girl and the associated radiological manifestations.

Case report

A 14-year-old girl presented following a short history of nonspecific malaise, weakness, left upper quadrant pain and early satiety. On physical examination, there was marked splenomegaly and a midabdominal bruit. She was growth retarded, being below the fifth percentile for both height and weight. Laboratory investigations revealed pancytopenia, with a hemoglobin level of 86 g/l, white blood cell count of $1.9 \times 10^9/\mu l$ and platelet count of $54 \times 10^9/\mu l$. The anemia was microcytic in type with a mean corpuscular volume of 73.7 fl and a mean corpuscular hemoglobin concentration of 21.7 pg.

Sonography demonstrated a large splenic mass of mixed echogenicity relative to normal spleen with punctate, hyperechoic foci resembling calcifications (Fig. 1a). An enhanced CT scan (Fig. 1b), revealed a heterogeneous splenic mass of mixed attenuation, measuring $18 \times 15 \times 11$ cm with a rim of normal splenic tissue seen superiorly and anteriorly. The delineation between the mass and normal splenic parenchyma was more clearly defined by CT than US. A technetium-labelled sulfur colloid scan (Fig. 2) demonstrated some accumulation of radiotracer by the mass, but this was less than the uptake by the normal splenic tissue. A technetium-labelled red blood cell study showed some peripheral accumulation of radiotracer by the mass with incomplete uptake centrally on delayed images.
Fig. 1  a Transverse US scan demonstrating a large splenic mass (M), with multiple hyperechoic foci (arrows) resembling calcifications and compression of the left kidney (K). (S spine). b Enhanced CT scan (L left, R right).

On MRI (0.5 T) the mass was isointense on T1 weighting (Fig. 3a) and hyperintense but heterogeneous on T2 weighting (Fig. 3b) relative to normal splenic tissue. With increased T2 weighting, the overall signal of the mass did not increase compared to that of cerebrospinal fluid (CSF; Fig. 3c). At splenectomy, the spleen weighted 1200 g and the mass consisted of a red pulp SH (Fig. 4).

Discussion

Symptomatic splenic hamartoma was first described by Videbaek in 1953 [4] and symptoms including pancytopenia, fatigue, anorexia, left upper quadrant pain, recurrent infections and growth retardation have been described [5, 6]. To our knowledge, only 20 previous reports of SSH have been published [1, 2, 4–9], and, of these, only four were in the pediatric age group [1, 5, 6]. Three of the four cases were in boys, although in adults SSH has a female sex preponderance [5]. In all four cases, patients presented with hepatosplenomegaly and anemia. Growth retardation, recurrent infections, fever and thrombocytopenia were also present in three cases. Lymphadenopathy and leukopenia were present in two of the four cases.

Microscopically, SH consist of an aberrant mixture of normal constituents of splenic parenchyma. They are predominantly made up of red pulp elements but may have associated lymphoid (white pulp) elements. The differential diagnosis of a splenic mass in a child includes cyst, hamartoma, hemangioma, lymphoma and leukemia [10–12]. Splenic lymphangiomas and metastases are rare in this age group [12]. The radiological features on CT and US have been described previously as a homogeneous solid mass without evidence of calcification [3]. SH containing true calcifications have, however, been reported in some cases [8]. In our patient, the lesion was a heterogeneous solid mass containing multiple punctate foci resembling calcifications on US. However, pathologically these foci were shown to represent nodules of hyalinization and necrosis. While uptake of sulfur colloid by a splenic mass, as in this case, would indicate functioning splenic elements, SH may frequently appear as a photopenic area [3]. In addition, one previous case report of a splenic hemangioma demonstrated increased radiotracer uptake on sulfur colloid scan [13].

The only previously published report of the MRI appearance of SH [3] demonstrated features similar to those in our case, namely isointensity on T1-weighted images and hyperintensity on T2-weighted images relative to normal splenic tissue. In addition, in our case, with increasing T2 weighting (TR 2800/TE 90–3500/120) a corresponding increase in signal intensity equivalent to that of CSF was not demonstrated as would be