Case report

Infarction of an accessory spleen secondary to splenic vein thrombosis in essential thrombocytosis

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Abstract

Accessory spleen is an incidental finding of no clinical significance in most patients. Essential thrombocytosis is a chronic, myeloproliferative disorder with a clinical course complicated by thrombotic episodes. We report on a case of essential thrombocytosis presented with infarction of the accessory spleen secondary to splenic vein thrombosis.

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Accessory spleen is a frequent congenital anomaly of the spleen occurring 3–20% of the population [1–8]. The clinical significance of an accessory spleen is limited to its hyperplasia after splenectomy and to the relatively infrequent condition of its torsion. Infarction of an accessory spleen secondary to torsion is a well-defined entity [3–6]. Essential thrombocytosis is a hematological disease that may cause venous thrombosis in different parts of the body and splenic infarction is not an unexpected complication of the disease [9,10]. However, infarction of an accessory spleen in essential thrombocytosis has not been previously reported.

We present the radiological findings of a case of essential thrombocytosis presented with infarction of the accessory spleen secondary to splenic vein thrombosis.

1. Case report

A 44-year-old woman with the diagnosis of essential thrombocytosis was admitted with pain and tenderness in her left hypochondrium. The patient was evaluated by sonography that revealed splenomegaly and a wedge shaped hypoechoic infarct area in the posterior-inferior part of the spleen. A spheric mass in 5 cm in diameter was noticed in the splenic hilum (Fig. 1a). The echogenicity of the mass was lower than the spleen and was similar to the infarcted area of the spleen. Splenic vein in the splenic hilum was found to have thrombosis and multiple collateral veins were observed in that area. Partial thrombosis of the splenic vein was demonstrated in the midline of the abdomen, in its course beneath the pancreas (Fig. 1b). The patient was then examined by computed tomography (CT) after the use of contrast material, which showed that the mass was in similar density with the infarcted area of the spleen and the splenic vein thrombosis was clearly demonstrated (Fig. 2). A further evaluation with pre- and post-contrast magnetic resonance (MR) imaging was performed (Fig. 3). The internal structure of the mass was shown to be inhomogeneous due to the hemorrhagic components seen as hyperintense areas both in the mass and in the infarcted areas of the spleen on gradient echo T1-weighted images. T2-weighted images demonstrated hypointense areas of retracted clot and hyperintense areas of free fluid inside of the mass. The lesion did not show enhancement during breath-hold dynamic contrast enhanced gradient echo images. The mass with the properties of its localization, similar appearance with the infarcted area of the spleen in all
examination modalities, lack of contrast enhancement and also with the finding of splenic vein thrombosis, was considered to be an enlarged infarcted accessory spleen. Clinical approach was conservative for the infarction of the original spleen so surgical treatment was not considered for the original or for the accessory spleen. The patient was symptomatically treated and was followed-up for 3 years. The lesion regressed in diameter during the follow-up and became undetectable on final MR imaging. The infarction area of the original spleen healed with fibrosis. No other complications occurred.

2. Discussion

Embryological development of spleen occurs from the mesenchymal cells migrating to the dorsal mesogastrium. Incomplete fusion of splenic tissue gives rise to formation of an accessory spleen. Accessory spleen is very frequent and considered as an anatomical variant seen in up to 20% of the population, commonly located in the splenic hilum, along the splenogastric ligament or in the course of splenic vessels [1–3]. The vascular pedicle of an accessory spleen is commonly related to the splenic hilum but may also be related to the tail of the pancreas, gastroplenic ligament, small bowel mesentery or to the vessels of the gastric fundus [3,5].

Accessory spleen has a silent clinical course and may remain undetected for life. Clinical importance appears when splenectomy is considered or when spontaneous torsion of an accessory spleen occurs. It rarely may cause intra-abdominal bleeding secondary to spontaneous rupture. Hyperplasia of an accessory spleen may be observed after splenectomy, which may reflect the relapse of the hematological diseases. The torsion, on the other hand, is a well-known complication that may occur in an accessory spleen causing acute abdominal findings [3–7]. In torsion of the accessory spleen, the venous return is disturbed first causing hemorrhagic infarction of the parenchyma. The same hemodynamics holds true for the presented case in which the splenic vein thrombosis caused venous infarction of the accessory spleen as well as the original spleen. Thrombosis is a common complication in essential thrombocytosis and splenic venous infarction may be observed as a consequence [9,10]. However, infarction of an accessory spleen in an essential thrombosis patient has not been previously reported.

The imaging findings of infarction of accessory spleen, reported in torsion cases, include a mass in or near the splenic hilum, commonly larger than the expected size of an accessory spleen [3–7]. It is seen as a hypoechoic mass in US and non-enhancing, hypodense lesion on CT. MR imaging has been reported to demonstrate the hemorrhagic components occurring secondary to venous type of infarction. Magnetic susceptibility effects of blood degrading products as deoxyhemoglobin or methemoglobin, as well as
the retraction of the clot determine the signal intensity on
MR. The MR sequences that are sensitive to susceptibility
effects are helpful in demonstration of hemorrhage. In
the presented case, infarcted accessory spleen includes
hyperintense areas on gradient echo T1W images corre-
sponding to methemoglobin content. The retraction of
clot and the presence of free fluid mainly determine the
signal on spin echo T2W images. The lack of contrast
enhancement and appearance of a thick pseudcapsule on
post-contrast images are expected findings in accessory
splenic infarction.

The radiological differential diagnosis of a mass in this
localization with these radiological findings includes
pancreatic masses and pseudocysts, enteric or omental
cysts, intestinal duplication cysts and abscesses [7]. The diagnosis of the presented case is based on the finding of
similarity of the internal structure of the lesion to the
infarcted area of the spleen in all imaging modalities.
Conservative clinical approach has limited us to provide a
pathological specimen. The infarction area in the native
spleen healed with fibrosis and the infarcted accessory
spleen probably has gone to autosplenectomy.

The vascular compromise of accessory spleen has been
reported in several cases with torsion of the accessory
spleen; however, infarct of an accessory spleen secondary to
splenic vein thrombosis has not been previously reported.
Also, the demonstration of this entity in an essential
thrombocytosis patient does not exist in the literature. The
patient presented here is unique in these aspects.

3. Summary

Essential thrombocytosis is a hematological disease that
may cause venous thrombosis in different parts of the body.
However, infarction of an accessory spleen in essential
thrombocytosis has not been previously reported.

A 44-year-old woman with the diagnosis of essential
thrombocytosis was admitted with pain and tenderness in
her left hypochondrium. Sonographic examination revealed
splenomegaly and a wedge shaped hypoechoic infarct area
in the posterior-inferior part of the spleen. A spheric mass
in 5 cm in diameter was noticed in the splenic hilum.
Partial thrombosis of the splenic vein was demonstrated in

Fig. 3. Spin echo T2-weighted (TR/TE = 3500/160) (upper left); dynamic contrast enhanced gradient echo T1-weighted (TR/TE/FA = 108/4/65), pre-contrast
(upper right), early-arterial (lower left) and late-venous (lower right) MR images of the patient at the same level with CT. The internal structure of the lesion is
inhomogeneous on T2-weighted image with hypointense areas of retracted clot and hyperintense areas of free fluid. Pre-contrast gradient echo T1-weighted
image shows the hemorrhagic components as hyperintense areas both in the mass (arrow) and in the infarcted areas of the spleen. The central part of the lesion
does not show enhancement but a peripheral rim-like enhancement occurs together with the splenic parenchyma.
the midline of the abdomen. CT of the patient showed that the mass was in similar density with the infarcted area of the spleen and the splenic vein thrombosis was better observed. Further evaluation with MR imaging demonstrated hemorrhagic components in the mass as well as in the infarcted areas of the spleen. The lesion did not show enhancement during breath-hold dynamic contrast enhanced gradient echo images. The mass with the properties of its localization, similar appearance with the infarcted area of the spleen in all examination modalities, lack of contrast enhancement and also with the finding of splenic vein thrombosis, was considered to be an enlarged infarcted accessory spleen. The patient was symptomatically treated and was followed-up for 3 years. The lesion regressed in diameter during the follow-up and became undetectable on final MR imaging. The infarction area of the original spleen healed with fibrosis. No other complications occurred.

Infarction of an accessory spleen secondary to splenic vein thrombosis has not been previously reported. Also, the demonstration of this entity in an essential thrombocytosis patient does not exist in the literature. The reported case is unique in these aspects.

References


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