Imaging for abdominal involvement in amyloidosis

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ABSTRACT

Involvement of the abdominal organs has variable presentations mostly without specific findings. The objective of this pictorial essay was to illustrate the computed tomography and magnetic resonance imaging (MRI) findings of abdominal involvement in systemic amyloidosis. Heterogeneous appearance of the liver, periportal involvement, diffuse low signal intensity of spleen on T2-weighted MRI, and thickened bowel wall may be helpful imaging findings when accompanied by presence or history of chronic inflammatory disease and clinical suspicion for amyloidosis.

Radiologic features

Radiologic findings of abdominal involvement in systemic amyloidosis are nonspecific and can present in a variety of ways. Heterogeneous appearance of liver, periportal involvement, diffuse low signal intensity of spleen on T2-weighted images and thickened bowel wall can be helpful if accompanied by history of chronic inflammatory disease and clinical suspicion for amyloidosis.

Hepatic involvement typically results in organ enlargement with abnormal liver function tests. Amyloid can accumulate in the hepatic parenchyma throughout the sinusoids or in vessel walls (Fig. 1) (8). On US, hepatic involvement of amyloidosis has nonspecific findings such as, heterogeneous echogenicity, diffuse or focal areas of decreased parenchymal attenuation (Fig. 1). The most important CT and MRI findings of hepatic involvement in systemic amyloidosis are hepatomegaly, heterogeneous appearance of liver (Figs. 1, 2) and periportal involvement (Fig. 1). Periportal involvement is seen as low signal intensity on T1-weighted and T2-weighted images. Diffuse low signal intensity on T2-weighted images can be an indication of diffuse involvement of hepatic amyloidosis (Fig. 3). It has been...
Imaging findings of abdominal amyloidosis

Main points

- Radiologic findings of abdominal involvement in systemic amyloidosis are nonspecific and can present in a variety of ways.
- Together with the clinical history of inflammatory disease in a patient, heterogeneous appearance of liver, periportal involvement, diffuse low signal intensity of spleen on T2-weighted images with bowel wall thickening findings can suggest systemic amyloidosis.
- Interpretation of multiple organ involvement by CT and/or MRI and getting acquainted with these imaging findings is essential for early diagnosis and prompt management.

reported that asymmetric contour, triangular hepatomegaly (its apex at the falciform ligament) and heterogeneous attenuation might aid in distinguishing amyloidosis from other infiltrative diseases (5). Hepatic rupture and prominent calcification can rarely be seen (2, 9). The differential diagnosis includes cirrhosis, fatty liver, and iron overload. Differentiation from cirrhosis can be made by absence of segment four atrophy, fatty liver by absence of signal drop in out-of-phase images, and iron overload by absence of signal drop in in-phase images.

As in the liver, the main radiologic finding of splenic involvement in amyloidosis is splenomegaly, which occurs in 4%–13% of patients (5). Calcification and absence of contrast enhancement can rarely be seen (particularly on CT examinations) usually due to vascular involvement and diffuse parenchymal amyloid infiltration (Fig. 4) (5). Some studies in the literature indicated that MRI of amyloidosis demonstrated splenic involvement with high signal intensity on T1-weighted images and low signal intensity on T2-weighted images (Fig. 3) (2, 10, 11). Low signal intensity on T2-weighted imaging findings might be due to reduced blood content caused by accumulation of amyloid in the spleen (5). The differential diagnosis includes iron overload. In-phase and out-of-phase images or T2* measurements may help to differentiate iron overload from amyloidosis.

Kidney involvement is a prominent result in systemic amyloidosis and almost 50% of the patients with secondary amyloidosis

Figure 1. a–g. A 59-year-old woman with primary amyloidosis. Ultrasonography (a) shows diffuse heterogeneous liver parenchyma. Axial contrast-enhanced T1-weighted images (b, c) demonstrate diffuse heterogeneous liver parenchyma with periportal involvement (c, black arrow). Note also diffuse irregular wall thickening of stomach (white arrow). Axial contrast-enhanced CT images (d, e) show periportal involvement (black arrows) and diffuse wall thickening of stomach (white arrow). In panel (f), microscopic finding (hematoxylin and eosin stain; original magnification, ×400) reveals acute amorphous amyloid deposition in the portal area (arrows). Panel (g) shows immunohistochemical staining (×400) of amorphous depositions in liver.

Figure 2. a–d. A 27-year-old man with familial Mediterranean fever. Axial T2-weighted image (a) demonstrates heterogeneous liver parenchyma. After one year, axial contrast-enhanced CT images (b, c) demonstrate infarct areas in the liver. Note also enlargement in the right adrenal gland (c, arrow). In panel (d), microscopic finding (H-E stain; original magnification, ×400) reveals acute amorphous amyloid deposition around blood vessels in the portal area (arrows).
die of renal failure (8). Radiologically, kidneys are generally imaged as smaller with cortical thinning—referred as amyloid contracted kidneys—which is found in nearly 50% of systemic amyloidosis cases (Fig. 5) (12). Enlarged kidneys with smooth contours may be seen, probably in the early phase (Fig. 3). US characteristics of renal involvement are nonspecific; the renal parenchyma shows diffusely increased echogenicity and corticomedullary differentiation is present. Doppler US findings of renal involvement are also nonspecific and consist of a normal or increased interlobar artery resistive index, which can be seen in other renal parenchymal diseases, corresponding to increased peripheral vascular resistance of the affected kidney. Other renal imaging findings are focal renal parenchymal mass, renal calcifications, and renal pelvic masses due to amyloid deposition and perirenal soft tissue mass with calcifications (13).

The main radiologic finding of adrenal gland involvement is enlargement of the gland (Fig. 2). This appearance is a nonspecific finding, which can occur in adrenal gland hyperplasia and granulomatous diseases (14).

According to the previous reports, small bowel is the most commonly involved organ in the gastrointestinal system (15, 16). The most frequent imaging finding of small intestinal amyloidosis is regular thickening of the folds (focal or diffuse), which displays edema due to ischemia as a result of vascular accumulation. Barium enema findings include regular thickening of the folds, jejunulation of the ileum, granular mucosal pattern, tiny polypoid protrusions, and amyloid tumor (5). Descending and rectosigmoid colon are the most commonly involved sites of the large intestine. Barium enema findings of colonic involvement are nonspecific: i.e., luminal narrowing, loss of colonic haustrations, thickened mucosal folds, and a nodular mucosal pattern. CT and MRI of small and large intestinal involvement include bowel wall thickening, dilatation, and mesenteric infiltration (Figs. 3, 4). MRI plays an important role in demonstrating bowel wall thickening and thickening of the mesenteric and perirectal fat planes. Mesenteric involvement can be seen as high signal intensity on T2-weighted images.

Patients with gastric involvement have symptoms resulting from pyloric obstruction (17). Mucosal granules, mucosal or intramural filling defects and thickened folds can be seen in barium enema studies. The pertinent imaging findings include diffuse or focal gastric wall thickening, focal deposits, amyloid tumors, and gastric wall calcifications (Fig. 1). Gastric involvement with diffuse wall thickening can be demonstrated by MRI (Fig. 1). Diffuse or focal wall thickening mimick gastric tumors, thus histologic analysis is essential.

As in the other organs, involvement of the ureters and bladder may be focal or diffuse. Amyloid deposition frequently involves the
lower ureter and is usually unifocal. Linear submucosal or intramural calcification of the renal pelvis and ureter is a characteristic of amyloidosis. CT or MRI can demonstrate filling defects, irregular ureteral narrowing, stricture, and diffuse or focal wall thickening (Fig. 6) (16). Diffuse bladder wall thickening or a filling defect in the lumen can be seen. Linear submucosal or intramural calcification of the bladder wall is specific for amyloidosis. It is difficult to distinguish focal masses from primary bladder cancer.

Conclusion
In conclusion, abdominal involvement can occur in primary and secondary forms of systemic amyloidosis. Imaging findings are usually nonspecific and display variable presentations. Together with a clinical history of inflammatory disease, heterogeneous appearance of liver, periportal involvement, diffuse low signal intensity of spleen on T2-weighted images, and bowel wall thickening findings can suggest systemic amyloidosis. Getting acquainted with these imaging findings is essential for the early diagnosis and prompt management.

Conflict of interest disclosure
The authors declared no conflicts of interest.

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