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AMYLOIDOSIS: AN UNUSUAL CAUSE OF MESENTERIC, OMENTAL AND LYMPH NODE CALCIFICATIONS

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We present a rare case of amyloidosis initially presenting with giant calcified inguinal adenopathy. Further imaging revealed diffuse calcifications within the mesentery and greater omentum.
Amyloid deposition may mimic chronic granulomatous disorders and primary or secondary neoplastic conditions. Although definite diagnosis is made on histology, the radiologist should include amyloidosis in the differential diagnosis in the absence of a clinical history of neoplastic disorders or chronic infection, especially if extensive intralesional calcifications are seen. Ultrasound may be useful to target solid noncalcified areas in easily accessible extra-abdominal locations.

Key-word: Amyloidosis.

Case report

An 81-year-old female presented with bilateral inguinal soft tissue swellings. The lesions were painless and of hard consistency on palpation. The clinical notes of the patient mentioned biopsy of a similar soft tissue lesion at the left groin 20 years earlier, revealing a nonspecific benign fibrous lesion at histology. Further clinical history was unremarkable. According to the patient, the masses were slowly increasing in volume during the last 6 months. Computed Tomography (CT) showed bilateral giant adenopathy with intralesional coarse calcifications (Fig. 1). In addition, multiple small nodular calcifications were seen in the omentum. Subsequent CT of the upper abdomen confirmed diffuse calcifications along the greater omentum, mesentery, the hepatic fissure, peritoneal surface of the spleen and splenic hilum (Fig. 2).

Magnetic Resonance Imaging (MRI) of the groin showed heterogeneous signal of the enlarged inguinal lymph nodes on both pulse sequences. There was no enhancement of the intralesional calcifications, whereas the noncalcified areas showed vivid enhancement (Fig. 3).

Lesion heterogeneity was also seen on ultrasound. The calcified areas showed retroacoustic shadowing, whereas the solid components were hypoechoic with increased power Doppler signal (Fig. 4). An ultrasound guided biopsy was performed at the level of the noncalcified solid portions of the left groin

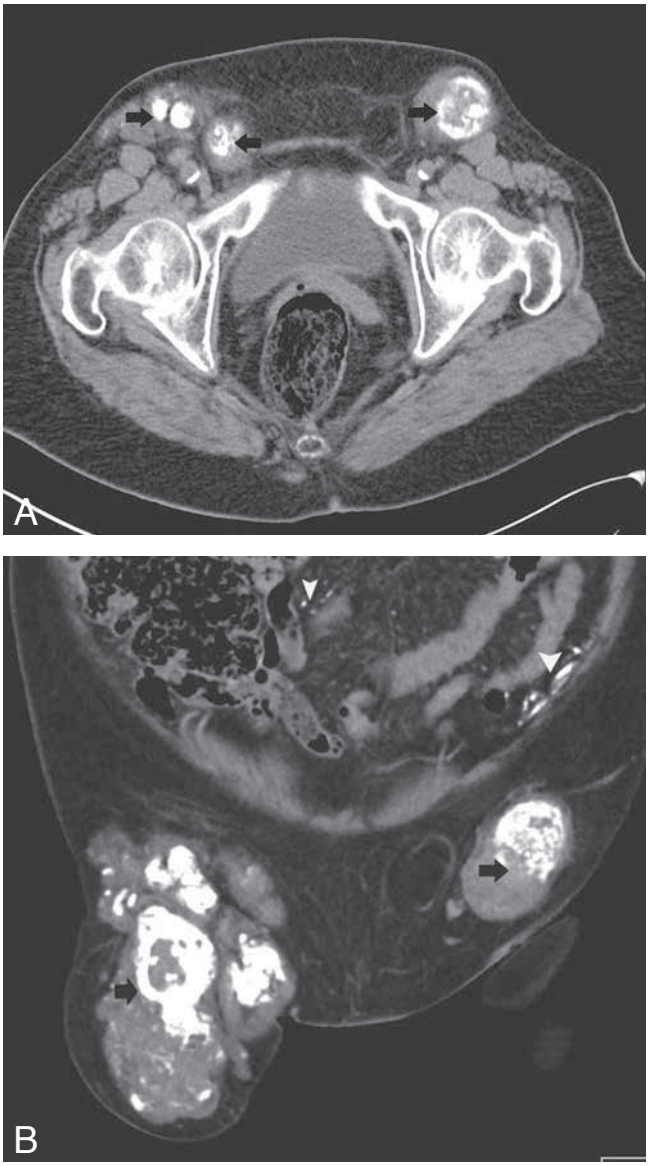

Fig. 1. — CT scan of the pelvis. Axial (A) and coronal reformatted image (B) shows bilateral massive inguinal adenopathy with intralesional coarse calcifications (black arrows). Note also small nodular calcifications within the omentum (white arrowheads).

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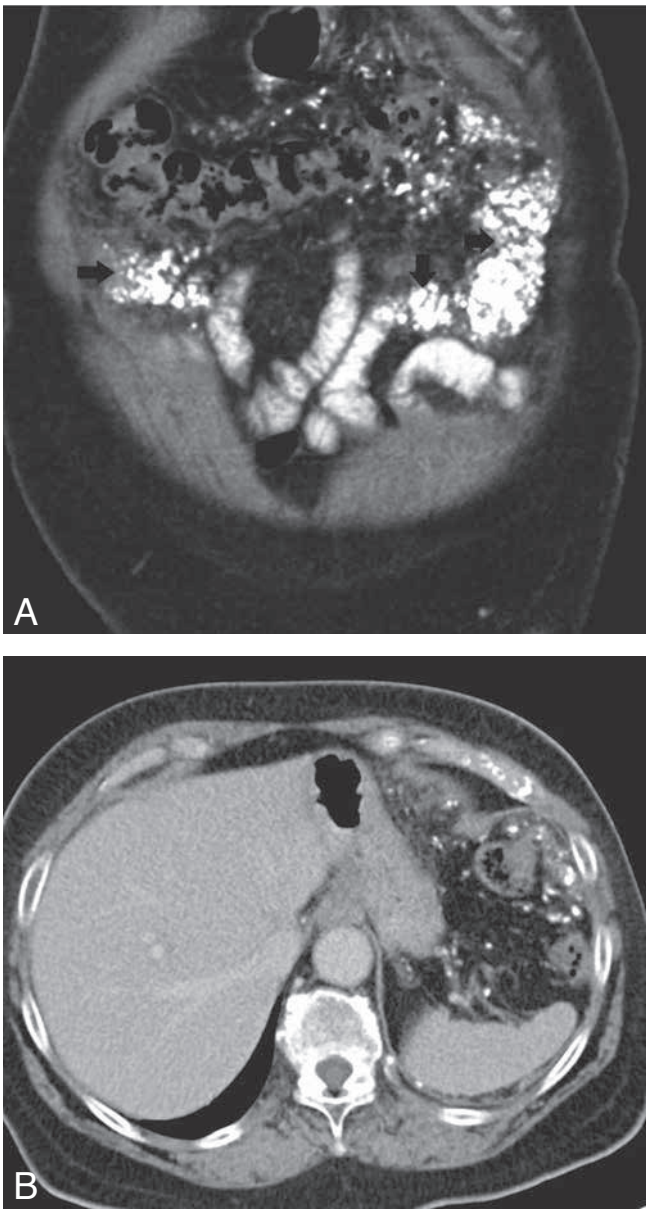

Fig. 2. — CT scan of the upper abdomen. Coronal reformatted image (A) showing thickening of the omentum which contains a mantle of speckled calcifications (black arrows). Axial image (B) revealing multiple nodular calcifications within the mesentery, the splenic hilum and the posterior surface of the spleen. Axial image at a slightly lower level (C). Note subtle calcification at the hepatic fissure (white arrowheads).

lesion. Histology of the biopsy specimen showed amyloid deposits on Congo red stain, exhibiting apple-green birefringence on polarized light (Fig. 5).

Discussion

Amyloidosis consists of a heterogeneous group of disorders characterized by widespread extracellular deposition of amyloid, an insoluble fibrillar protein, in multiple organs and tissues (1-4). The current classi-

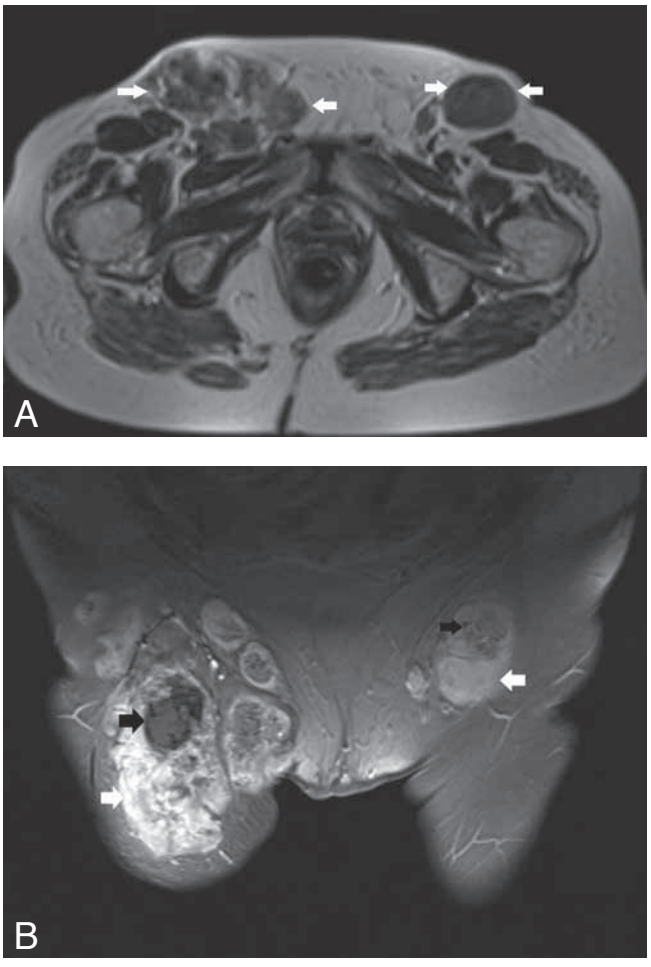

Fig. 3. — MR imaging of the groins. Axial T2-WI (A) and coronal fat-suppressed T1-WI after intravenous administration of gadolinium contrast (B). The enlarged lymph nodes and are of heterogeneous signal on T2-WI, with the calcified areas being hypointense (white arrows). The noncalcified components showed vivid enhancement (white arrows), whereas the calcified solid components are nonenhancing (black arrows) (B).

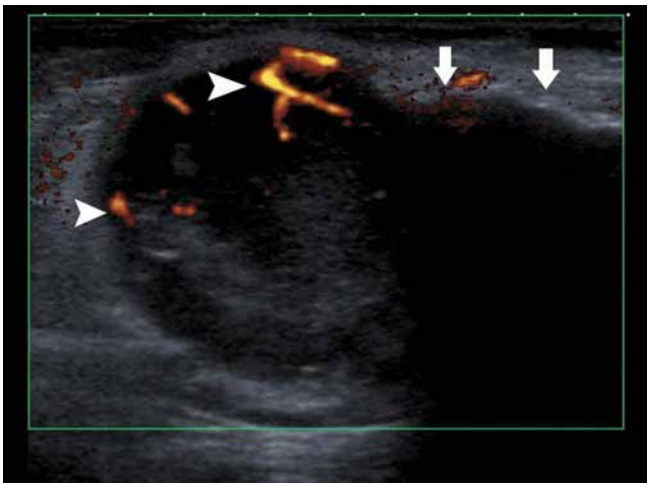

Fig. 4. — Ultrasound of the groins showing enlarged inguinal lymph nodes with calcified areas demonstrating retroacoustic shadowing (white arrows). The noncalcified solid parts are hypoechoic with foci of increased Doppler flow (white arrowheads).

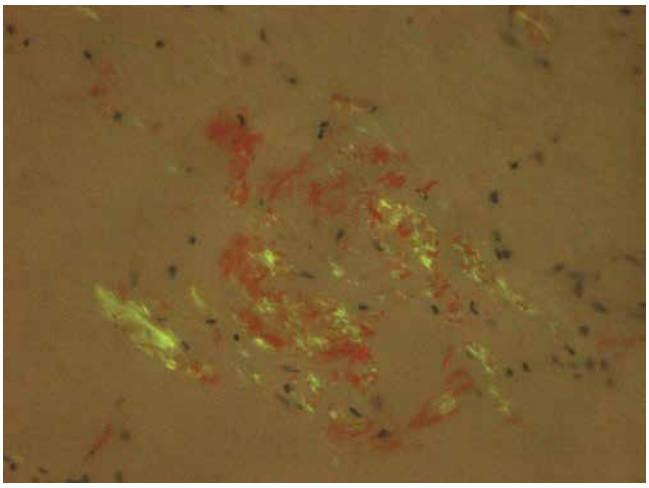

Fig. 5. — Photomicrograph of the histopathologic specimen (Congo red stain) showing apple-green birefringence on polarized light.

fication of amyloid disease is based on the specific type of protein fibril involved in the amyloid deposits, and uses an abbreviation of the protein, prefixed with the letter A. For example, amyloidosis caused by transthyretin (TTR) is termed "ATTR". The nomenclature of amyloid protein fibril is complex and has been revised recently by the nomenclature Committee of the International Society of Amyloidosis (5, 6). The literature contains many clinical and histochemical classifications that were used when the chemical diversity of amyloid disease was poorly understood. One such historical classification distinguishes systemic and localised forms of amyloidosis. Systemic amyloidosis refers to involvement of more than one body organ or system, whereas localised amyloidosis affects only one body organ or tissue type. Another older classification divides primary or secondary forms. Primary amyloidosis arises from a disease with disordered immune cell function such as multiple myeloma and other immunocyte dyscrasias. Secondary amyloidosis occurs as a complication of some other chronic inflammatory or tissue destructive disease. The Committee of the International Society of Amyloidosis recommends to suppress these confusing alternative classifications.

Amyloidosis has been described in various anatomic locations including the central nervous system, head and neck, chest, heart, lymph nodes, breast, gastrointestinal and genitourinary tract, spleen, subcutis, musculoskeletal system (1-4, 7-10).

Involvement of the omentum, mesentery and peritoneum are extremely rare manifestations of this disease (1-4). The disease may be either nodular or diffuse (4). In the diffuse form, amyloid deposition in the greater omentum, mesentery and retroperitoneum usually results in hazy infiltration of the fat within these structures on CT (4). Extensive calcifications within these deposits are rare (2), but are highly suggestive of amyloidosis (2, 4). Enlargement of retroperitoneal lymph nodes is seen in the nodular form.

Lymph node involvement occurs in up to 37% of patients and the hilar, mediastinal and para-aortic lymph nodes are most commonly involved, but other locations have been reported as well (8, 9). In our patient, the dominant finding at presentation was the presence of huge calcified inguinal lymphadenopathy.

On imaging, the involved lymph nodes are usually enlarged and may contain intralesional calcifications. CT is the preferred technique for demonstration of calcifications, although ultrasound may be a useful technique for guiding biopsy, particularly in lymph nodes which are easily accessible. The biopsy should be targeted to noncalcified solid parts, because of the soft consistency of these components. Non calcified areas may demonstrate increased color Doppler signal, in keeping with vascularized components.

Apart from evaluation of cardiac amyloidosis (7), MRI of amyloid deposition in soft tissues and lymph nodes has been rarely described (10). MR appearance may be variable and is usually heterogeneous on all pulse sequences. Densely packed amyloid fibrils, dense collagen and calcifications appear hypointense on T2-weighted images. High signal intensity areas on T2-WI may be due to necrosis, whereas hemorrhagic, proteinaceous or eosinophilic components may be of high signal on T1-WI (10). Contrast enhancement can also be observed in lymph nodes (8).

The differential diagnosis of calcified peritoneal, mesenteric and omental amyloidosis includes primary or metastatic neoplasms that may provoke desmoplastic reaction, such as peritoneal mesothelioma and ovarian carcinoma (2). Ascites is usually present in these cases, whereas ascites is only seen in 20% of cases of peritoneal amyloidosis, secondary to associated cardiac, hepatic or renal failure (4). In our case, there was no ascites. Another differential diagnosis consists of infectious disorders of the peritoneum and greater omentum, such as tuberculosis. The differential diagnosis of massive lymphadenopathy includes lymphoma, infection, metastatic disease, and granulomatous diseases. Rosai-Dorfman disease, Castleman's disease, Bartonellosis, and rare inflammatory disorders such as Kimura's disease and Kichuchi-Fujimoto disease should be considered in the differential diagnosis in case of enhancing lymph nodes (8). Chronic tuberculosis or granulomatous disease should be excluded when lymph nodes are calcified.

Histologic confirmation is required for definite diagnosis of amyloid deposition. Although routine hematoxylin-eosin stains can be suggestive, the diagnosis should be confirmed by a Congo Red stain, where examination under polarized light reveals a characteristic apple-green colour, due to the birefringence of the amyloid deposition.

Conclusion

Although rare, amyloidosis should be considered in the differential diagnosis of diffuse omental, mesenteric and peritoneal calcifications and in the presence of massive calcified lymphadenopathy.

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