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Images of Interest / Imagens de Interesse

Primary Retroperitoneal Amyloidosis: A Rare Cause of Obstructive Uropathy

Amiloidose Retroperitoneal Primária: Uma Causa Rara de Uropatia Obstrutiva

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Abstract

Retroperitoneal amyloidosis, one of the rarest forms of amyloidosis, is characterized by the deposition of insoluble fibrillar proteins in the retroperitoneum. It typically presents on CT and MRI as a soft-tissue thickening or mass with progressive calcification, carrying a risk of obstructive uropathy. The imaging characteristics of retroperitoneal amyloidosis may be similar to other conditions, such as retroperitoneal fibrosis, making image-guided biopsy and histologic evaluation crucial for a definitive diagnosis. This article presents the case of an 81-year-old man with hydronephrosis caused by primary retroperitoneal amyloidosis, an uncommon complication of this rare disease.

Keywords

Retroperitoneal amyloidosis; Obstructive uropathy; Computed tomography; Magnetic resonance imaging.

Resumo

A amiloidose retroperitoneal, uma das formas mais raras de amiloidose, é caraterizada pela deposição de proteínas fibrilares insolúveis no retroperitoneu. Apresenta-se habitualmente na TC e RM como um espessamento ou massa de partes moles com calcificações progressivas, acarretando o risco de uropatia obstrutiva. As características imagiológicas da amiloidose retroperitoneal podem ser semelhantes a outras patologias, tal como a fibrose retroperitoneal, tornando a biópsia guiada por imagem e avaliação histológica cruciais para um diagnóstico definitivo. Este artigo descreve o caso de um homem de 81 anos com hidronefrose causada por amiloidose retroperitoneal primária, uma complicação incomum desta doença rara.

Palavras-chave

Amiloidose retroperitoneal; Uropatia obstrutiva; Tomografia computorizada; Ressonância magnética.

Case Report

An 81-year-old man presented to the hospital with a three-month history of fatigue, abdominal discomfort, intermittent right flank pain, and weight loss of 15 kg. No additional symptoms or clinical signs were observed. The patient had a history of colon cancer in remission for ten years. Laboratory tests revealed mild anemia (hemoglobin 11.8 mg/dL) and acute renal dysfunction (serum creatinine 1.4 mg/dL). Tumor markers were within normal ranges.

A contrast-enhanced computed tomography (CT) of the abdomen (Figure 1) showed an infiltrative soft-tissue thickening of the right retroperitoneal fat. This process encased and compressed the right renal pelvis and the proximal ureter, causing hydronephrosis. No intralesional calcifications were observed. No additional foci of disease were identified in the remaining abdominal and pelvic organs. On magnetic resonance imaging (MRI) scan (Figure 2), the lesion exhibited intermediate signal intensity on fat-suppressed T1-weighted images (WI) and slightly decreased signal on T2-WI. Gradient-echo images showed a diffuse signal decrease on opposed-phase compared to in-phase images.

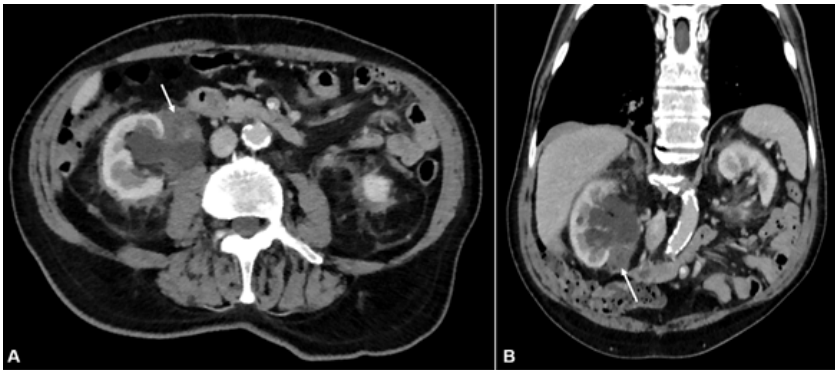

Figure 1 – Computed Tomography. Contrast-enhanced axial (A) and coronal (B) planes show an infiltrative soft-tissue thickening of the right retroperitoneal fat (arrow), causing hydronephrosis.

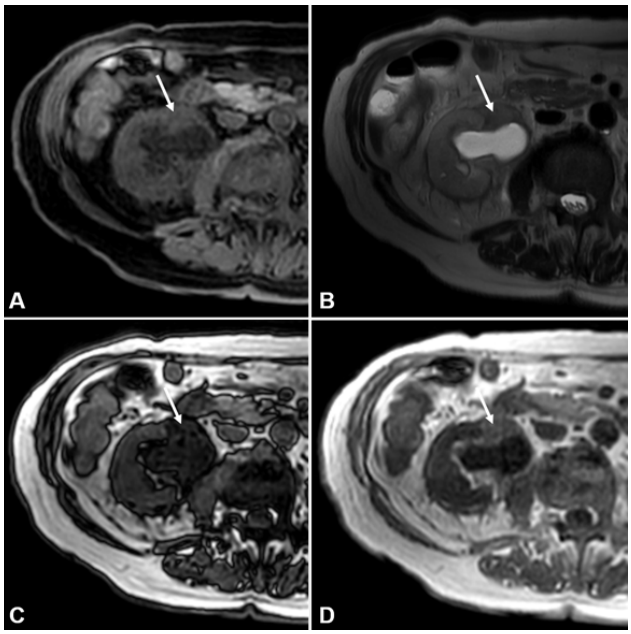

Figure 2 – Magnetic Resonance Imaging. The lesion shows intermediate signal intensity on fat-suppressed T1-WI (arrow, A) and slightly decreased signal on T2-WI (arrow, B). There is a diffuse signal drop-off on opposed-phase (arrow, C) compared with in-phase images (arrow, D).

The lesion was submitted to a CT-guided biopsy (Figure 3), and the pathology report of the specimen revealed the diagnosis of lambda light-chain retroperitoneal amyloidosis (Figure 4).

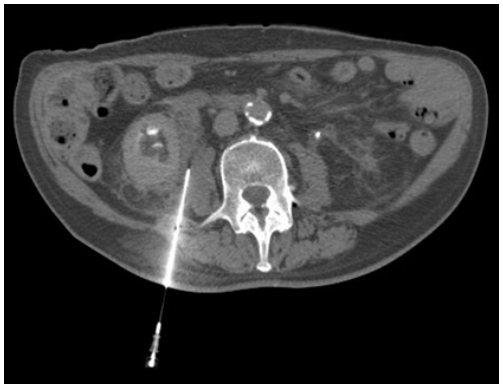

Figure 3 – Computed Tomography. Image-guided biopsy of the retroperitoneal lesion.

Discussion

Amyloidosis is a group of rare disorders characterized by the pathological extracellular deposition of insoluble fibrillar proteins. This process compromises tissue function, potentially leading to significant morbidity and mortality. The protein component determines the type of amyloidosis.[1,2] Primary amyloidosis is classified as amyloid light-chain (AL) disease. Secondary amyloidosis, often called amyloid A (AA) disease, is typically associated with chronic inflammatory conditions. Amyloidosis can be further categorized as localized or systemic, with the systemic type characterized by the deposition of protein fibrils in multiple organs.[2]

Retroperitoneal involvement is one of the rarest forms of amyloidosis presentation, posing clinical and imaging challenges. Amyloid deposits can infiltrate the retroperitoneum, causing soft-tissue thickening or mass formation, which may undergo calcification over time. This infiltrative process can cause compression of the renal pelvis and ureters, potentially leading to hydronephrosis, a rare complication of amyloidosis.[1,2]

CT and MRI typically reveal a diffuse replacement of normal retroperitoneal fat with a non-fatty soft-tissue component. On MRI, this tissue usually exhibits intermediate signal intensity on T1-WI and low signal intensity on T2-WI. In our case, the lesion exhibited just a slightly decreased signal on T2-WI, probably due to superimposed inflammatory changes. Additionally, amyloid-infiltrated tissues often show a diffuse signal drop on opposed-phase gradient-echo images due to the mixture of fat and water in the voxels of fatty soft tissues infiltrated by amyloid.[1]

The differential diagnosis for infiltrative retroperitoneal soft-tissue processes includes retroperitoneal fibrosis, desmoplastic tumors, and lymphoma. Histologic assessment is essential for a definitive diagnosis, with imaging playing a crucial role in guiding biopsy procedures.[1,2]

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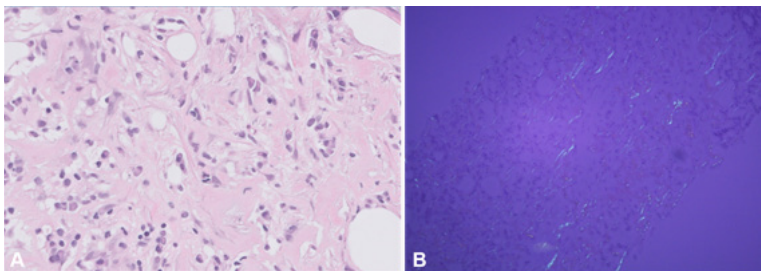

Figure 4 – Histopathological examination. (A) Hematoxylin and eosin-stained (HE) histologic section showing amorphous material consistent with amyloid. (B) Congo red-stained section under a polarizing microscope reveals apple-green birefringence, characteristic of amyloid.

Ethical Disclosures / Divulgações Éticas

Conflicts of interest: The authors have no conflicts of interest to declare.
Conflitos de interesse: Os autores declaram não possuir conflitos de interesse.
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Suporte financeiro: O presente trabalho não foi suportado por nenhum subsídio ou bolsa.

Confidentiality of data: The authors declare that they have followed the protocols of their work center on the publication of data from patients.
Confidencialidade dos dados: Os autores declaram ter seguido os protocolos do seu centro de trabalho acerca da publicação dos dados de doentes.
Protection of human and animal subjects: The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki).

Protecção de pessoas e animais: Os autores declaram que os procedimentos seguidos estavam de acordo com os regulamentos estabelecidos pelos responsáveis da Comissão de Investigação Clínica e Ética e de acordo com a Declaração de Helsínquia da Associação Médica Mundial.

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