Case Report

Retroperitoneal fibrosis: A rare mimicker of a perirenal hematoma✩,★★

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ABSTRACT

Retroperitoneal fibrosis (RPF) is a relatively rare entity, characterized by chronic inflammation and fibrosis in the retroperitoneal periaortic tissues. Due to this rarity, the diagnosis might be delayed or challenging, especially with atypical presentations of RPF. We report the imaging findings of an incidental finding of active RPF in an asymptomatic 84-year-old male patient. This patient was initially diagnosed with a perirenal hematoma due to an atypical imaging presentation on a routine staging computed tomography scan of the chest and the abdomen to stage a recently discovered prostate cancer. However, due to the persistence of the perirenal lesion, subsequent magnetic resonance imaging of this lesion was conducted, which resulted in the final diagnosis of active RPF. With this case report, we want to address attention to the potential atypical presentation of RPF, which could mimic a perirenal mass and demonstrates the importance of a broad differential diagnosis. Secondly, we want to point out the importance of magnetic resonance imaging in the evaluation of RPF, since it can differentiate between inactive or chronic idiopathic RPF and active idiopathic RPF or malignant RPF. Finally, this case shows the importance of comparing current imaging findings with previous imaging results, since a perirenal hematoma would reduce in size on follow-up imaging, which was not the case in our patient.

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Case report

At the end of 2018, an 84-year-old male patient underwent a computed tomography (CT) of the thorax and abdomen (at our institution) to stage a recently diagnosed prostate adenocarcinoma (Gleason 7 [3 + 4]). This showed some suspicious retroperitoneal lymph nodes along the aorta, but no evidence of metastasis. Interestingly, the staging CT demonstrated the incidental finding of a left-sided perirenal mass and a mild dilatation of the left renal pelvis (Fig. 1). Despite the absence of documented trauma, the tentative diagnosis of a perirenal hematoma was made.

Surgical history revealed an enucleation prostatectomy 10 years prior and transurethral resection of a bladder polyp 9 years prior. Histology of the resected bladder polyp showed

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a noninvasive low grade papillary transitional cell carcinoma, stadium TaG1, which was entirely resected. Urinary laboratory values were normal.

On subsequent 68Ga-prostate-specific membrane antigen (PSMA) positron emission tomography scan at the beginning of 2019 however, the suspected lymphadenopathy was not PSMA avid. Hence, the prostate adenocarcinoma was defined as stadium T2-3N0M0.

The prostate adenocarcinoma was subsequently treated with combined hormonal (decaptepyl) and local radiotherapy (56 Gray in total).

One year later, in January 2020, a follow-up CT urography was performed for the evaluation of the presumed left-sided perirenal hematoma and the mild hydronephrosis. This showed persistence of the perirenal “mass” with a density of 40 Hounsfield units on unenhanced series (Fig. 2). There was an extension to the left psoas muscle and also caudally towards the pelvic cavity. The left renal pelvis and calyces still appeared mildly dilated (Fig. 3). No ureteral abnormalities were noted. There were no signs of active bleeding on the contrast-enhanced series (Fig. 4). Regarding the right upper urinary tract, normal findings were reported.

Given the persistence of the perirenal “mass” on the CT urography, with similar imaging findings compared to the previous CT and positron emission tomography imaging, the suggested diagnosis of perirenal hematoma was questioned. Subsequent magnetic resonance imaging (MRI) of the kidneys was performed 3 weeks later for further differentiation.

This showed mild anterior displacement of the left kidney with a surrounding discrete T2 hyperintense infiltrating cuff in the left perirenal space with encasement of the renal vessels and the proximal ureter extending towards the aorta (Fig. 5). There was also caudal extension alongside the iliopectineous muscle. On T1, the mass appeared hypointense (Fig. 6). Upon intravenous contrast administration, the mass showed vivid enhancement in the portal venous phase and faint, homogeneous contrast enhancement in the renal parenchymal phase (Fig. 7). The mass also showed diffusion restriction with a low apparent diffusion coefficient (Fig. 8). The morphological appearance of this left perirenal mass was similar to the previous CT findings.

The tentative diagnosis of active retroperitoneal fibrosis (RPF) was made with the differential diagnosis of a perirenal lymphoma or perirenal extending leukemia. Nonetheless, due to the persistent character of the perirenal lesion (unchanged morphological appearance over 1 year), perirenal lymphoma and perirenal extending leukemia were considered very unlikely.

Two months after the first MRI investigation, a new MRI of the kidneys was performed for re-evaluation, showing unchanged imaging findings, further supporting the tentative diagnosis of active RPF. Given the patient's history of prostate adenocarcinoma, the patient was clinically diagnosed with secondary RPF after prostate adenocarcinoma. No further follow-up imaging was planned.
Discussion

Idiopathic RPF and most secondary forms of RPF are characterized by the proliferation of aberrant fibrous and inflammatory tissue, typically around the infrarenal abdominal aorta and often progressing in the retroperitoneum with the envelopment of ureters and inferior vena cava [1,2].

On rare occasions, there may be involvement of retroperitoneal structures such as the kidney and the renal pelvis [1]. One can imagine that the occurrence of RPF in atypical locations could delay the diagnosis, as happened in our case with a predominant perirenal localization [1].

The most common clinical presentation of idiopathic RPF is flank-, back-, or abdominal pain. Besides these rather aspecific symptoms, other complaints can occur, which can be linked to the growth and secondary compressive or infiltrative features of the retroperitoneal plaque formation such as ureteral compression and deep vein thrombosis. In 56%-100% of patients with idiopathic RPF, the fibro-inflammatory tissue causes obstructive uropathy and subsequent renal failure due to ureteric entrapment [1]. Our patient however was asymptomatic and did not suffer from any significant renal complication.

The estimated incidence of the idiopathic form of RPF is 0.1-1.3 cases/100,000 persons per year [3]. While the idiopathic type of RPF is the most common, the differential diagnosis is broad, such as secondary forms of RPF, malignancy, and hematoma [2].

Given the presence of a prostate adenocarcinoma, a definitive differentiation between an idiopathic or a secondary form of RPF is difficult based on imaging alone, since prostate cancer can be a possible cause of secondary RPF. The chronic therapy with β-receptor blocking agents in our patient, which is also a possible cause of secondary RPF, makes an idiopathic form of RPF also less likely [2].

Idiopathic RPF has 2 disease stages with a distinct microscopic appearance, which also results in different imaging characteristics. The early stage is characterized by edematous and highly vascular and hypercellular tissue, associated with active inflammation. The late or chronic stage shows marked fibrosis and can contain scattered calcifications. These different tissue characteristics are hypothesized as possible parameters to monitor treatment response [1,2].
Unfortunately, there are no standardized diagnostic criteria to diagnose idiopathic RPF [1].

Idiopathic RPF is most commonly diagnosed with CT or MRI. On CT, it presents as a homogeneous plaque surrounding the abdominal aorta and encircling the common iliac arteries. Extension of the plaque around the ureter and inferior vena cava is frequently seen. The density of this plaque on a non-contrast-enhanced CT approaches muscle density. Upon contrast admission, this plaque can show a variable degree of contrast enhancement. Quantification of contrast enhancement on CT is difficult and cannot be used confidently to assess the metabolic activity in patients with idiopathic RPF [1–3].

On MRI, the intensity of idiopathic RPF is typically low on T1-weighted imaging [2,3]. A hematoma however would present with high signal intensity on nonenhanced T1-weighted imaging, which also excludes our initial diagnosis of a perirenal hematoma [1]. On T2-weighted imaging, the intensity is variable, depending on the activity of the plaque. In active stages, there is high signal intensity on T2-weighted imaging, while in chronic idiopathic RPF T2 signal intensity is typically low. Chronic or inactive stages of idiopathic RPF can also be differentiated from active idiopathic RPF by the absence of restricted diffusion and the absence of contrast-enhancement in chronic or inactive idiopathic RPF. Active idiopathic RPF, malignant RPF, and retroperitoneal malignant neoplasms exhibit similar signal intensity on T2-weighted imaging, restricted diffusion, and show similar contrast-enhancement characteristics. The inability to confidently differentiate active RPF from malignant RPF and retroperitoneal malignant neoplasms poses a major challenge [1–4]. This was in fact also a difficulty in our case. Nevertheless, given the indolent behavior over time the perirenal lesion is very unlikely to be a true retroperitoneal malignant mass.

The aim of treatment is to stop the progression of the inflammatory reaction and ideally to reduce the size of this fibro-inflammatory tissue. Corticosteroids are the first-line therapy to treat idiopathic RPF. For secondary forms of RPF, the treatment approach should be based on the cause. As already mentioned, ureteric obstruction is a possible complication of RPF and in most cases of ureteric obstruction, surgery is done. Surgery also has the advantage to biopsy some tissue for further investigation. The treatment prognosis of secondary RPF

Fig. 7 – MRI of the kidneys. Axial contrast-enhanced T1 fat-saturated gradient-echo images. Early arterial phase (A), portal venous phase (B), renal parenchymal phase (C). The lesion displays vivid enhancement in (B) and faint enhancement in (C).

MRI, magnetic resonance imaging.

Fig. 8 – MRI of the kidneys. Diffusion-weighted imaging (DWI) and ADC map. Axial series. The perirenal lesion has a hyperintense signal on DWI sequences (A) and a hypointense signal on the ADC map (B), indicating restricted diffusion. ADC, apparent diffusion coefficient; MRI, magnetic resonance imaging.
varies greatly between etiological causes. Idiopathic RPF on the other hand, has a remission rate after corticosteroid therapy between 75% and 95%. Imaging with CT and MRI can be used to assess changes in the size of the retroperitoneal tissue. However idiopathic RPF has relapse rates of up to 72% and can be considered as a chronic-relapsing entity [1,3].

Patient consent statement

Consent to participate: informed consent was obtained from the patient.

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