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Letter to the Editor

Renal replacement lipomatosis with interstitial nephritis and secondary degenerative changes: A rare entity

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Dear editor,

Renal replacement lipomatosis (RRL) is a rare benign condition characterized by the replacement of the renal sinus, hilum, and perirenal space with extensive fatty tissue. It is also known as replacement lipofibromatosis and is generally observed in individuals aged 60-70 years [1]. RRL is typically associated with renal calculi, chronic inflammation, and hydronephrosis, leading to severe atrophy of the renal parenchyma and, ultimately, a non-functional kidney [2]. A study indicated that in 3,500 intravenous pyelogram cases, the incidence of RRL was 0.66 %, with no significant gender preponderance [3].

We report a case of a 65-year-old male patient who presented with a left renal mass and hematuria. Non-contrast CT of the kidneys, ureters, and bladder (NCCT KUB) revealed a 23 mm calculus at the pelvi-ureteric junction, with extensive peripelvic and periureteric fat stranding and a prominent renal fascia. Left nephroureterectomy was performed, and the excised

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specimen was submitted for histopathological examination. On gross examination, the specimen measured 11.5×7.5×4.5 cm; the ureter measured 7 cm in length and 0.6 cm in diameter. The outer surface was unremarkable. On sectioning, the pelvicalyceal system was markedly dilated and replaced by fat (Figure 1A).

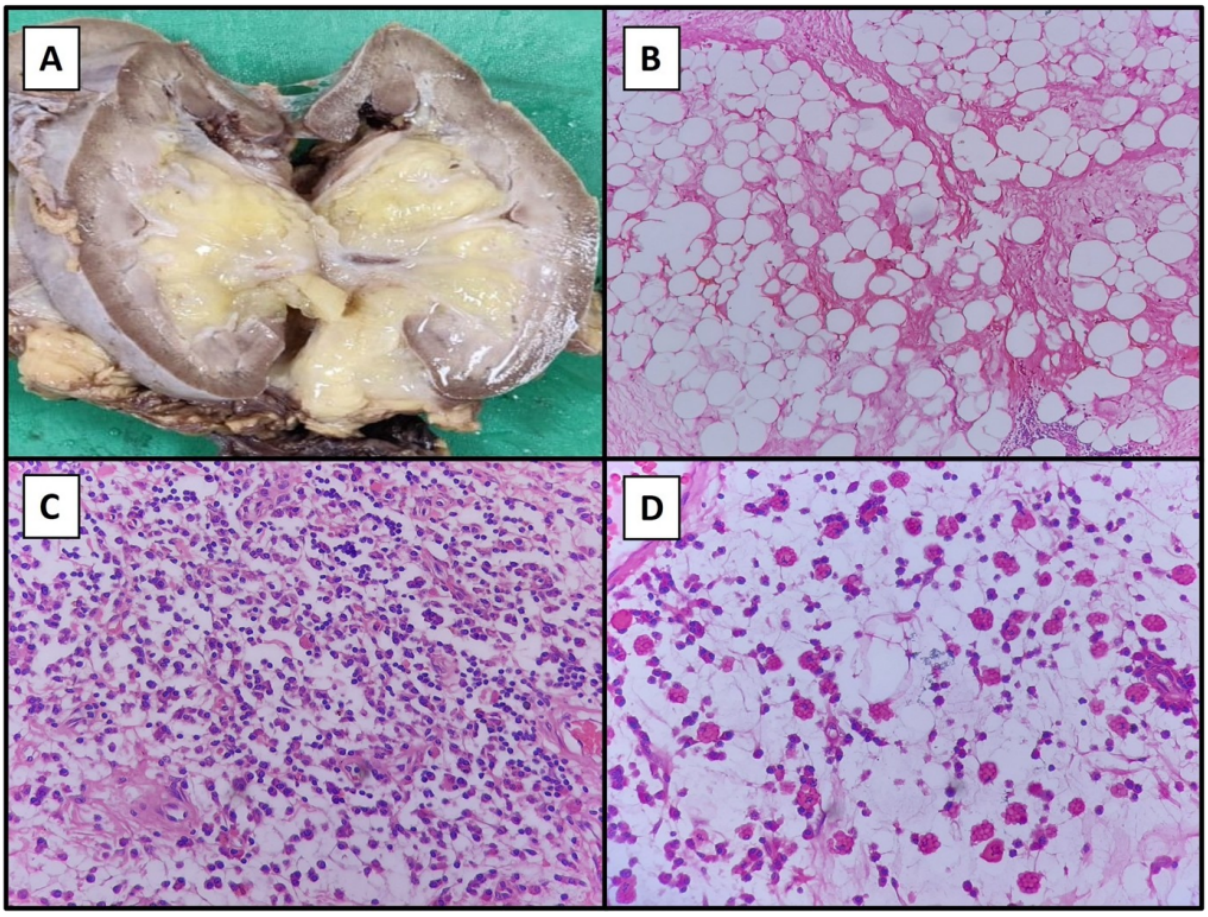

Figura 1. Renal replacement lipomatosis with interstitial nephritis and secondary degenerative changes

Note. (A) Gross image of left kidney with cut surface showing dilated pelvicalyceal system and entirely replaced by fat; (B) Microscopic image showing mature adipocytes separated by fibrous septae (H&E stain, 100X); (C) Microscopic image showing interstitial aggregates of increased plasma cells and few lymphocytes (H&E stain, 400X); (D) Microscopic image showing numerous plasma cells in the form of mott cells and morula cells on myxoid background (H&E stain, 400X).
Source: Own elaboration.

No tumor or growth was identified. A single brown-colored stone was noted. Sections were taken from all the representative areas of the kidney. On histopathological examination, it was found that the dilated pelvicalyceal system showed excessive myxoid degeneration and the presence of mature adipocytes (Figure 1B). The interstitium showed a dense mixed inflammatory cell collection comprising polymorphs, lymphocytes, plasma cells, eosinophils, granulation tissue, and hemosiderin-laden macrophages, along with excessive congestion and

oedema. Lymphoid aggregates were also present. Additionally, plasma cells were increased in number and present in various forms like Mott cells, flame cells and morula cells (Figure 1C and 1D). Glomerulus and renal tubules were unremarkable. Blood vessels were thick-walled and congested. Alcian blue with PAS stain highlighted mucoid collection. Mucicarmine was patchy positive, and Congo red stain was negative. Features of malignancy were not observed. The final diagnosis was renal replacement lipomatosis with interstitial nephritis and secondary degenerative changes. The patient remains under follow-up.

The differential diagnosis of RRL includes malakoplakia, xanthogranulomatous pyelonephritis, lipoma, angiomyolipoma, and liposarcoma. Lipoma is a well-circumscribed, benign, encapsulated tumor composed of mature adipocytes, while malakoplakia is characterized by PAS (Periodic Acid Schiff)-positive foamy macrophages and the presence of Michaelis-Gutmann bodies [4]. Angiomyolipoma is comprised of dysmorphic thick-walled blood vessels, adipocytes, and smooth muscle. Xanthogranulomatous pyelonephritis displays lipid-laden foamy macrophages, giant cells, inflammation, and cholesterol clefts. Liposarcoma consists of three variants, namely, well differentiated, myxoid and pleomorphic, and has the presence of pathognomonic atypical spindle cells with multivacuolated lipoblasts on microscopic examination [5]. Jesrani G. *et al.* [5] reported a case of bilateral RRL in a known case of obstructive uropathy.

In view of the raised interstitial plasma cells, IgG4-related disorder or plasma cell dyscrasia could also be a possibility, which came negative in our case. Nephrectomy remains the treatment of choice, with histopathology serving as the gold standard for confirmatory diagnosis. Because of lack of suspicion and rarity, RRL is often not suspected pre-operatively [1]. The purpose of this letter is to raise awareness about this rare entity and to help prevent its overdiagnosis or misdiagnosis.

Conflicts of interest

The authors have no conflicts of interest related to this article.

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Declaration of patient consent

The authors declare that patient consent has been taken.

Author contributions

All authors contributed to the study conception and design. Material preparation and first draft of the manuscript was written by Mousmi Agrawal; data collection and analysis were performed by Deepak Kumar Biswal; editing of manuscript was done by Amit Kumar Chowhan. All authors read and approved the final manuscript.

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