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Idiopathic Retroperitoneal Fibrosis Presenting as a Podagra-Like Condition: A Case Report

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Abstract

Retroperitoneal fibrosis (RPF) is a rare fibroinflammatory disorder in which fibrous tissue forms around the infrarenal aorta and iliac vessels, often compressing the ureters and causing obstructive uropathy and renal impairment. It typically presents insidiously with back, flank, or abdominal pain and constitutional symptoms. We describe a 52-year-old man with a background of hypertension who presented on three occasions over 10 months with an acutely painful, swollen first metatarsophalangeal (MTP) joint, initially on the left and subsequently on the right. The first two episodes were managed as presumed gout, with naproxen and then a short course of oral prednisolone, and his symptoms settled between attendances. His serum urate and creatinine rose across these attendances. At his third presentation, he was incidentally found to be hypertensive with severe renal impairment and was admitted. Imaging demonstrated bilateral hydronephrosis secondary to ureteric compression by a confluent para-aortic soft tissue mass. Following further investigation and specialist multidisciplinary team review, a working diagnosis of idiopathic RPF was made. Bilateral nephrostomies relieved the obstruction, and he was commenced on prednisolone and mycophenolate mofetil. He remains under urology and rheumatology follow-up, with serial renal function monitoring and radiological surveillance of the mass, nine months after diagnosis. This case illustrates a podagra-like presentation of idiopathic RPF and highlights the value of maintaining a broad differential diagnosis in recurrent monoarthritis with declining renal function and the importance of excluding malignancy before an idiopathic label is applied, particularly when tissue diagnosis is not feasible.

Categories: Urology, Rheumatology, Internal Medicine
Keywords: gout disease, idiopathic retroperitoneal fibrosis, igg 4 disease, podagra, retroperitoneal fibrosis

Introduction

Retroperitoneal fibrosis (RPF) is a rare fibroinflammatory disorder characterized by the formation of fibrous tissue around the infrarenal aorta and iliac vessels, frequently resulting in ureteric compression, obstructive uropathy, and renal impairment [1]. It typically presents insidiously with back, flank, or abdominal pain and constitutional symptoms [1]. Approximately two-thirds of cases are idiopathic, while the remainder are secondary to malignancy, medications, infection, radiotherapy, or previous surgery [2]. The reported incidence ranges from 0.1 to 1.3 cases per 100,000 people per year, with a male predominance and peak incidence between the fifth and seventh decades of life [3]. A substantial proportion of cases previously classified as idiopathic are now recognized as belonging to the spectrum of immunoglobulin G4-related disease (IgG4-RD) [4].

Because RPF presents with non-specific symptoms and its radiological features overlap with those of retroperitoneal malignancies and lymphoma, diagnosis may be delayed. Secondary causes must therefore be actively investigated before a diagnosis of idiopathic RPF is established [5]. We describe a 52-year-old man with hypertension whose disease manifested not through the typical insidious onset of pain but through recurrent episodes of acute pain and swelling of a first metatarsophalangeal (MTP) joint, initially managed as presumed gout. Severe renal impairment was identified incidentally at his third presentation. To our knowledge, based on a targeted literature search, recurrent podagra-like episodes preceding the diagnosis of idiopathic RPF have rarely been reported. This case highlights the importance of considering a broad differential diagnosis in patients presenting with recurrent monoarthritis and deteriorating renal function, as well as the need to exclude malignancy before diagnosing idiopathic RPF, particularly when tissue confirmation is not feasible.

Case Presentation

A 52-year-old man with a history of hypertension presented to the emergency department in early 2025 with a painful, swollen left first metatarsophalangeal (MTP) joint. His regular medications were amlodipine 10 mg, doxazosin 4 mg, and ramipril 5 mg, each taken once daily. Blood tests showed a serum urate level of 471 µmol/L (reference range, 200-430 µmol/L), a serum creatinine level of 110 µmol/L, and a urea level of 5.1 mmol/L. He was treated for presumed gout with a course of naproxen and discharged. Six months later, he re-presented with similar symptoms affecting the left first MTP joint. Treatment was escalated to prednisolone 30 mg once daily for seven days. On this occasion, his serum urate level was 551 µmol/L, and his renal function had deteriorated, with a serum creatinine level of 169 µmol/L and a urea level of 6.9

mmol/L. On both occasions, gout was diagnosed presumptively on clinical grounds, supported by elevated serum urate levels; joint aspiration was not performed. His symptoms resolved between episodes.

Four months later, he presented to the urgent treatment center with a painful right first MTP joint, where markedly impaired renal function was incidentally identified. His estimated glomerular filtration rate (eGFR) was 16 mL/min/1.73 m^2, serum creatinine was 343 µmol/L, and urea was 15.3 mmol/L (Table *1*). His blood pressure was 189/108 mmHg despite treatment with three antihypertensive agents. He was referred to the general medical team and admitted for further investigation. On further questioning, he denied symptoms suggestive of systemic autoimmune disease, including sicca symptoms, skin rashes, and oral ulceration. He also denied back, flank, or abdominal pain, weight loss, fever, and night sweats. He had no history of smoking or asbestos exposure.

Given his deteriorating renal function, ultrasonography of the urinary tract was performed and demonstrated mild-to-moderate right-sided and moderate left-sided hydronephrosis. A non-contrast computed tomography (CT) scan of the urinary tract was subsequently performed because of his impaired renal function. It revealed severe bilateral hydronephrosis secondary to compression of both ureters by a confluent para-aortic soft-tissue mass (Figure *1*).



FIGURE 1: Coronal non-contrast CT of the urinary tract demonstrating a confluent para-aortic soft-tissue mass (arrow)

Confluent para-aortic soft-tissue mass (arrow) compressing both ureters, with severe bilateral hydronephrosis.

As both imaging studies were performed on the same day, the difference in the reported severity of hydronephrosis likely reflects differences in the imaging modalities. Ultrasonography is operator-dependent and provides a more limited assessment of the collecting systems and ureters, whereas CT offers a more comprehensive cross-sectional evaluation.

Blood tests revealed mild microcytic anemia, with a hemoglobin level of 115 g/L and a mean corpuscular volume (MCV) of 79 fL, but no other cytopenias or hematological abnormalities. The hematology team was consulted regarding the possibility of lymphoma. Further investigations included lactate dehydrogenase (LDH), paraprotein screening, serum free light chains, beta-2 microglobulin, and erythrocyte sedimentation rate (ESR).

The key results are presented in Table *1*. The ESR (64 mm/h), C-reactive protein (37 mg/L), beta-2 microglobulin (5.4 mg/L), and LDH (421 IU/L) levels were elevated. Serum kappa free light chains were mildly elevated at 24.19 mg/L (reference range, 9.6-21.6 mg/L), whereas lambda free light chains were within the reference range at 16.74 mg/L (reference range, 9.8-22.6 mg/L). The kappa-to-lambda ratio was 1.45, at

the upper limit of the reference range (0.87-1.45). Immunoglobulin G (IgG) subclasses, including IgG4, were within their respective reference ranges, and no paraproteins were detected.

Investigation	Patient values	Reference range
Hemoglobin	115 g/L	130-180 g/L
MCV	79 fL	83-101 fL
Creatinine	343 µmol/L	49-90 µmol/L
Urea	15.3 mmol/L	2.5-7.8 mmol/L
eGFR	16 mL/min/1.73 m²	>60 mL/min/1.73 m²
ESR	64 mm/hr	<20 mm/hr
Beta-2-microglobulin	5.4 mg/L	0.7-2.5 mg/L
LDH	421 IU/L	140-280 IU/L
C-reactive protein	37 mg/L	<5 mg/L
IgG4	0.39 g/L	0.04-0.86 g/L
Serum kappa free light chain	24.19 mg/L	9.6-21.6 mg/L
Serum lambda free light chain	16.74 mg/L	9.8-22.6 mg/L
Serum kappa/lambda light chain ratio	1.45	0.87-1.45

TABLE 1: Summary of laboratory investigations of the patient

MCV: mean corpuscular volume, eGFR: estimated glomerular filtration rate, ESR: erythrocyte sedimentation rate, LDH: lactate dehydrogenase.

To relieve hydronephrosis, bilateral nephrostomy tubes were inserted. Following multidisciplinary team (MDT) discussion, biopsy of the para-aortic mass was considered necessary for diagnosis. However, the procedure was technically challenging because of the mass's proximity to the aorta. Ultimately, the MDT concluded that the risk of aortic injury and associated bleeding was too high to permit safe biopsy.

A half-body fluorodeoxyglucose positron emission tomography-computed tomography (FDG PET-CT) scan was performed instead. This demonstrated circumferential increased tracer uptake in the retroperitoneum and around the proximal common iliac arteries, suggestive of RPF.

Autoimmune screening was negative for antinuclear antibodies (ANA) and antineutrophil cytoplasmic antibodies (ANCA). Further CT imaging of the neck and thorax revealed no additional masses or lymphadenopathy. Ultrasonography of the neck identified a cystic lesion in the right parotid gland, and ultrasound-guided aspiration was planned to investigate possible salivary gland involvement by immunoglobulin G4-related disease (IgG4-RD) or lymphoma. However, repeat ultrasonography at the time of the procedure, one month later, showed only tiny hyperechoic lesions consistent with normal intraparotid lymph nodes. No cystic or focal lesion was identified, and the cervical lymph nodes, parotid glands, submandibular glands, and thyroid gland appeared normal. Consequently, aspiration was not performed.

The case was subsequently discussed at a national IgG4-RD MDT meeting. In the absence of a tissue diagnosis, a working diagnosis of idiopathic RPF was established. Treatment was initiated with prednisolone 40 mg daily for seven days, followed by a taper over one month, alongside mycophenolate mofetil 1 g twice daily.

Following nephrostomy insertion, renal function improved, with the estimated glomerular filtration rate (eGFR) increasing from 16 to 34 mL/min/1.73 m² over three months. The patient has experienced no further episodes of joint pain and remains under urological and rheumatological follow-up, with serial monitoring of renal function and radiological surveillance of the mass. He remains under follow-up nine months after diagnosis.

Discussion

Ureteric obstruction and the resulting renal impairment are among the most common clinical consequences of idiopathic RPF [6]. Approximately two-thirds of RPF cases are considered idiopathic, while the remainder

are secondary to malignancy, medications, infection, surgery, or radiation exposure [2]. Malignancy accounts for approximately 8% of RPF cases [7]. Idiopathic RPF is increasingly recognized as part of the spectrum of IgG4-RD [2]. In this patient, IgG4-RD was considered less likely because of the normal serum IgG4 concentration and the absence of salivary gland abnormalities on neck ultrasonography. However, it cannot be definitively excluded without histological confirmation.

The pathogenesis of idiopathic RPF remains unclear, although current evidence suggests that genetic susceptibility, environmental exposures, and autoimmune mechanisms contribute to disease development [1]. Histopathological studies have demonstrated infiltration by T lymphocytes, B lymphocytes, plasma cells, macrophages, and eosinophils, leading to fibroblast activation and excessive extracellular matrix deposition [1]. Some cases exhibit features of IgG4-RD, including elevated serum IgG4 concentrations and tissue infiltration by IgG4-positive plasma cells [3]. Chronic periaortic inflammation triggered by atherosclerotic antigens has also been proposed as a contributing mechanism [8].

The patient's age and sex are consistent with the typical demographic profile of idiopathic RPF. However, because the disease is rare and often presents with non-specific symptoms, its true incidence may be underestimated.

Several risk factors have been associated with idiopathic RPF. Cigarette smoking and asbestos exposure are among the strongest environmental associations, with a synergistic effect reported when both exposures are present [9-11]. However, this patient had no history of smoking or asbestos exposure. None of his antihypertensive medications belonged to the drug classes traditionally associated with secondary RPF, such as beta-blockers, hydralazine, and methyldopa [1,12]. Autoimmune conditions, including autoimmune thyroid disease, systemic vasculitis, and other immune-mediated disorders, are also more common among affected individuals. In addition, genetic susceptibility associated with HLA-DRB1 alleles has been reported [10].

The diagnosis of idiopathic RPF is based on a combination of clinical, laboratory, radiological, and, where available, histopathological findings. Patients typically present with non-specific symptoms, including back or abdominal pain, weight loss, fatigue, and features of obstructive uropathy [12]. In contrast, this patient presented with recurrent gout-like monoarthritis. We hypothesize that these episodes may have been related to reduced renal urate excretion secondary to obstructive uropathy. Krishnan [13] reported an increasing prevalence of gout with declining eGFR, which is consistent with this hypothesis, given the patient's eGFR of 16 mL/min/1.73 m^2. However, joint aspiration was not performed, and gout was therefore not confirmed by crystal identification. Accordingly, these episodes are more appropriately described as podagra-like presentations.

Serum urate concentrations were elevated at both initial attendances, measuring 471 and 551 µmol/L, respectively. Both values exceeded the 360 µmol/L threshold cited in national guidance to support a clinical diagnosis of gout [14]. These findings are consistent with hyperuricemia in the context of recurrent podagra-like symptoms, although hyperuricemia alone does not establish a diagnosis of gout.

Two additional factors may have influenced the patient's clinical course. First, naproxen was prescribed at the initial presentation while the patient was taking ramipril. This combination can increase the risk of acute kidney injury and may have contributed to the subsequent deterioration in renal function, although this cannot be established from the available data. Second, glucocorticoids are a mainstay of treatment for idiopathic RPF [12]. We therefore hypothesize that the short course of prednisolone prescribed for presumed gout may have temporarily suppressed the underlying inflammation and delayed recognition of RPF. Both proposed mechanisms remain speculative and cannot be confirmed from a single case.

Hydronephrosis and ureteric obstruction have been reported in approximately 33%-67% of patients with idiopathic RPF, with similar rates in patients with and without IgG4-RD [15].

Inflammatory markers, including the ESR and C-reactive protein, are typically elevated in RPF. Contrast-enhanced CT and MRI are the principal imaging modalities and generally demonstrate a periaortic retroperitoneal soft-tissue mass encasing adjacent structures [10]. PET-CT may help assess disease activity and treatment response. Biopsy is recommended when malignancy cannot be excluded, imaging findings are atypical, or the response to treatment is suboptimal [3]. In this patient, intravenous contrast was avoided because of impaired renal function, and fluorodeoxyglucose PET-CT was used to further characterize the mass.

Glucocorticoids remain the first-line and most widely accepted treatment for idiopathic RPF. Prednisolone is typically initiated at 0.6-1.0 mg/kg/day and gradually tapered according to clinical, biochemical, and radiological responses [16]. Glucocorticoid treatment achieves remission in most patients and often reduces the fibrotic mass and relieves ureteric obstruction [12,16]. For patients with relapse, steroid dependence, or contraindications to glucocorticoids, immunosuppressive agents such as azathioprine, mycophenolate mofetil, methotrexate, cyclophosphamide, and rituximab may be considered [16]. Ureteric stenting or surgical ureterolysis may be required when obstructive uropathy persists [16]. Although no universally

accepted treatment gold standard exists, current evidence and international practice support glucocorticoids as the cornerstone of therapy [17].

In this patient, mycophenolate mofetil was initiated as a steroid-sparing immunosuppressive agent. Although mycophenolate has been used to treat idiopathic RPF [16], the evidence supporting its use remains limited. The randomized trial cited here evaluated methotrexate combined with low-dose prednisone rather than mycophenolate mofetil [17].

This case required an extensive investigation to exclude malignant mimics before a working diagnosis of idiopathic RPF could be established. This distinction is important because, although malignancy accounts for a minority of RPF cases, it has been identified in patients initially diagnosed with RPF [5].

To determine whether similar presentations had previously been reported, we searched PubMed/MEDLINE and Google Scholar from inception to October 2026 using combinations of the terms “retroperitoneal fibrosis”, “Ormond's disease”, “gout”, “podagra”, “hyperuricaemia”, and “monoarthritis”. We also screened the reference lists of relevant articles. Two relevant reports were identified. In a small case series, Cullen described two patients with idiopathic RPF who had been treated for gout before their RPF diagnosis. The report also described other patients with hyperuricemia and unusually numerous urate crystals within fibrotic tissue, leading the author to propose that urate crystal deposition might initiate the fibrotic process [18]. More recently, Benmoussa et al. reported radiation-associated RPF in a patient with a history of gout. The authors suggested that this case, together with previously reported associations with hyperuricemia, might indicate an underlying fibro-inflammatory predisposition [19].

In both reports, gout was an established diagnosis rather than the presenting feature of RPF. In our patient, serum urate was already elevated when the serum creatinine level was 110 µmol/L. However, the available evidence cannot establish whether hyperuricemia preceded or contributed to RPF, or instead resulted from impaired renal function associated with the disease. We found no previous reports describing recurrent podagra-like episodes preceding a diagnosis of idiopathic RPF.

This report has several limitations. First, the diagnosis of idiopathic RPF was made without histological confirmation because the risk of hemorrhage precluded biopsy. Consequently, malignancy and IgG4-RD could not be definitively excluded. Nevertheless, the available clinical, laboratory, and radiological findings, together with multidisciplinary assessment, made these alternative diagnoses less likely. Continued clinical and radiological surveillance is therefore essential.

Conclusions

Although the patient's age and sex were typical of idiopathic RPF, this case was notable for recurrent podagra-like monoarthritis, treated as presumed gout, preceding the incidental detection of severe obstructive renal impairment. Such presentations preceding an RPF diagnosis appear to be rarely reported. The absence of back pain, abdominal pain, and constitutional symptoms may have contributed to delayed recognition of the underlying disease. This case highlights the importance of investigating deteriorating renal function in patients presenting with recurrent podagra-like episodes, even when gout is clinically suspected.

As histological confirmation was not feasible, malignancy and IgG4-RD could not be definitively excluded, although they were considered less likely based on the available clinical, laboratory, and radiological findings. The patient was therefore managed under a working diagnosis of idiopathic RPF, supported by specialist multidisciplinary assessment and ongoing clinical and radiological surveillance. When tissue diagnosis is not feasible, this approach, alongside careful evaluation for secondary causes, may provide a pragmatic basis for clinical management.

Additional Information

Author Contributions

All authors have reviewed the final version to be published and agreed to be accountable for all aspects of the work.

Concept and design: Salma A. Abdirizak

Acquisition, analysis, or interpretation of data: Salma A. Abdirizak

Drafting of the manuscript: Salma A. Abdirizak

Critical review of the manuscript for important intellectual content: Salma A. Abdirizak

Disclosures

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