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## Case Report

## Pediatric idiopathic retroperitoneal fibrosis

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## ABSTRACT

Retroperitoneal fibrosis (RPF) is a very rare disease that is even more rare in the pediatric population. Even less common are idiopathic pediatric cases of retroperitoneal fibrosis, with a majority of reported pediatric retroperitoneal fibrosis cases being associated with secondary etiologies. We present an 11-year-old Caucasian female that was diagnosed with idiopathic retroperitoneal fibrosis using magnetic resonance imaging (MRI) to work-up severe bilateral hydronephrosis that was identified with retroperitoneal ultrasound. Given the uncommon nature of this serious condition, we present this case to illustrate the importance for physicians to include retroperitoneal fibrosis in the differential diagnosis of a pediatric patient presenting with obstructive urinary findings and understand the utility of using MRI to diagnosis and monitor this disease.

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## Introduction

We present a case of idiopathic retroperitoneal fibrosis (RPF) in a pediatric patient. RPF is a rare condition with an estimated incidence of 1 in 200,000–500,000 annually, most commonly

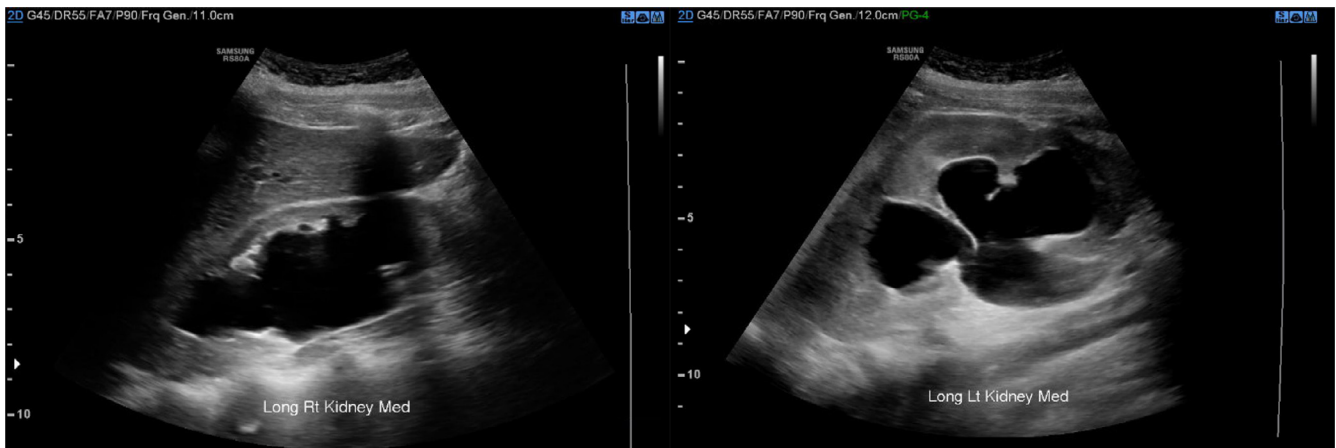
seen in 40–60-year-old males [1]. Pediatric RPF is a very rare entity with less than 40 reported cases, the majority of which have known secondary causes. Despite the uncommon occurrence of this disease in the pediatric population, we present this case to illustrate the similarities in the disease process and radiographic appearances of RPF in pediatric and adult

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**Fig. 1 – Ultrasound shows marked hydronephrosis bilaterally with relatively preserved cortical thickness.**

patients. It is important for RPF to be identified by radiologists in the pediatric population to prevent delay in diagnosis and management of this serious condition.

### Case report

An 11-year-old previously healthy Caucasian girl presented in outpatient consultation to the pediatric nephrology clinic for evaluation of elevated creatinine. At presentation, her creatinine measured 1.9 mg/dL, having increased from 0.6 mg/dL 1 year prior. Over the year prior to her consultation, she had experienced intermittent cramps but otherwise nonspecific abdominal and right flank pain, as well as an unexplained weight loss of 10 kg. Evaluation prior to nephrology consultation led to discovery of a normochromic normocytic anemia and an elevated erythrocyte sedimentation rate (ESR).

Her physical examination was unremarkable. A renal ultrasound was performed which demonstrated moderate to severe bilateral hydronephrosis (Fig. 1). She had no known history of urinary tract infections, vesicoureteral reflux, or voiding dysfunction. Subsequent magnetic resonance imaging (MRI) of the abdomen and pelvis again demonstrated bilateral hydronephrosis and proximal hydroureter, with nonvisualization of the distal ureters. A rim of soft tissue measuring up to 9 mm was visualized encasing the distal aorta with extension around the iliac vessels and ureters bilaterally (Fig. 2).

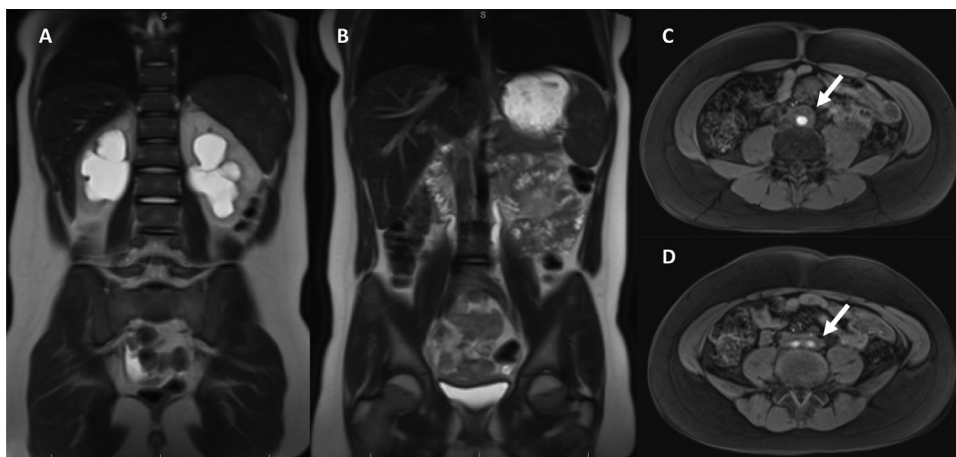
Three days following the MRI, this patient underwent bilateral nephrostomy tube placement, as well as laparoscopic biopsy of the retroperitoneal soft tissue. Hematoxylin and Eosin stained sections of retroperitoneal soft tissue showed dense fibrosis with admixed chronic inflammation with bland fibroblasts and areas of hypocellular (keloid-like) collagen. The inflammatory infiltrate was composed of an admixture of B (CD20+) and T lymphocytes (CD3+), plasma cells, histiocytes, and eosinophils. An immunohistochemical stain did not highlight a population of IGG4 positive plasma cells, and there was no staining for S100 or CD1a (Fig. 3). Final pathology

was consistent with fibrous tissue with chronic inflammation without evidence of lymphoma, and negative for IgG4 staining. Additional testing for oncologic, rheumatologic, and infectious processes was negative. Pathology and additional evaluation supported the radiographic diagnosis of idiopathic RPF (IRPF).

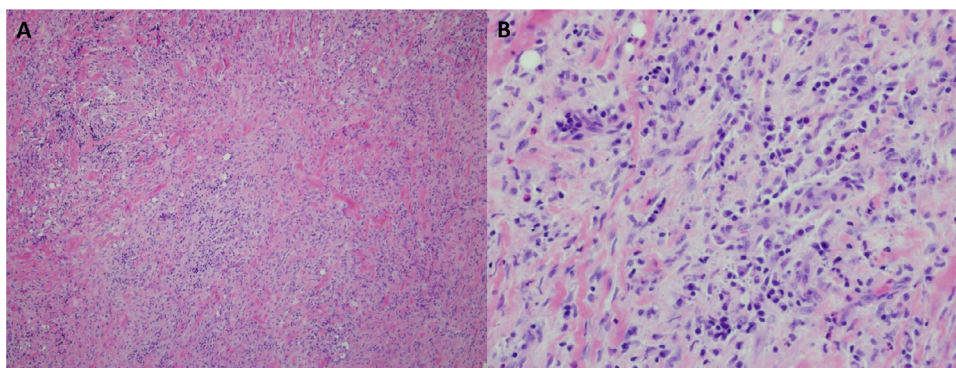
The patient was treated with a course of prednisolone for induction and mycophenolate mofetil for maintenance therapy. Her first follow-up MRI showed a decrease in size of the fibrosis by 33% (Fig. 4). Subsequent MRIs continued to show gradual decrease on soft tissue around the aorta with an 80% decrease 11 months after initial diagnosis. Ureteral compression also showed progressive improvement, and she was transitioned from bilateral nephrostomy tubes to bilateral indwelling ureteral stents. Her ESR normalized and anemia improved. She has continued to be managed medically with surgical ureterolysis deferred because of her continued radiographic and clinical response to mycophenolate mofetil.

### Discussion

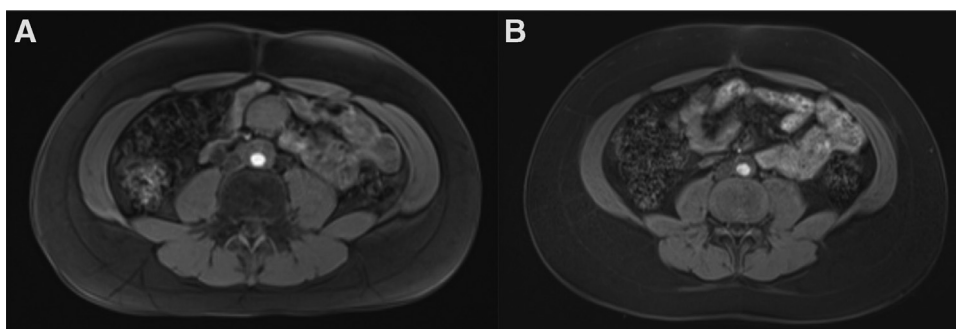
RPF is a rare diagnosis most commonly seen in middle-aged patients with a 3:1 male to female ratio [1]. It is characterized by the presence of a fibrous and/or inflammatory soft tissue in the retroperitoneum that often encases retroperitoneal structures like the great vessels and ureters [2]. Presentation and severity can vary widely based on the location and cause. RPF lacks clear clinical criteria for diagnosis, making this a challenging diagnosis. Causes of RPF can be divided into idiopathic and secondary RPF. So-called IRPF accounts for more than 70% of cases [3]. IRPF is associated with increased acute phase reactants and autoimmune disease. Immune staining of plasma cells is positive for IgG4 in up to 55% of patients (though not in our patient), suggesting that this could be an IgG4-mediated disease [4]. Causes for secondary RPF include prior surgery, malignancy, infection, autoimmune diseases (such as systemic lupus erythematosus [SLE] or juve-



**Fig. 2 – Initial MRI: Coronal T2 demonstrates bilateral hydronephrosis and hydroureter (A). Ureters were no longer visualized at the level of the iliacs (B). Axial T1 vibe images demonstrate a nonmass-like soft tissue collar (arrow) surrounding the distal aorta (C), and along the bilateral iliac arteries (D).**



**Fig. 3 – Hematoxylin and Eosin stained section of retroperitoneal soft tissue (A) displays dense fibrosis with admixed inflammation at 100 $\times$  magnification. Increased magnification to 400 $\times$  (B) reveals inflammatory infiltrate composed of lymphocytes, plasma cells, histiocytes, and rare eosinophils.**



**Fig. 4 – Initial imaging (A) demonstrates a soft tissue collar around the distal aorta, which demonstrated a significant decrease of soft tissue as demonstrated on initial follow-up imaging 2 months later (B).**

nile idiopathic arthritis (JIA)), and a variety of drugs including ergots, beta blockers, methyldopa, and hydralazine [1].

Pediatric RPF is quite rare with less than 40 reported cases. Within pediatric cases of RPF, it is more common to have secondary causes of RPF, such as those associated with pul-

monary hyalinizing granuloma, SLE or JIA. IRPF, such as the case we present, is more uncommon, with less than 10 reported cases in the pediatric literature [5].

Due to the varying presentations of RPF, the initial clinical features are often nonspecific. Disease is often advanced

when it becomes symptomatic, with patients presenting to their health care provider after the fibrosis has caused extrinsic compression of vessels or ureters. Pain in the lower back and/or flank is a common clinical manifestation, occurring in over 90% of patients at some point during the disease course [6]. Physical exam findings can include hypertension due to renal artery involvement, or lower extremity edema due to inferior vena cava or iliac vein involvement. Laboratory findings can often include elevated blood urea nitrogen (BUN) and creatinine correlating with obstructive uropathy. ESR is elevated about 75% of the time [7].

Because initial presentation is often concerning for urinary tract obstruction, retroperitoneal ultrasound is generally the first imaging study performed, especially in the pediatric patient. Ultrasound findings suggestive of RPF include a poorly marginated, hypoechoic mass [8]. However, due to ultrasound limitations, hydronephrosis may be the only detectable abnormality, so it is not the ideal imaging study for confirming this diagnosis.

Traditionally, the mainstay of imaging for RPF has involved a multiphase CT exam. This may be a reasonable option in older patients but is not appropriate in pediatric patients because of the amount of radiation exposure. We have demonstrated successful diagnosis and follow-up of pediatric RPF using MRI and believe this is the most appropriate imaging modality for diagnosis and disease monitoring in these patients. In the adult literature, it has been described that dynamic contrast-enhanced imaging can be useful for evaluation of RPF [9]. We found 15-minute delayed images very useful in delineating fibrosis from surrounding soft tissue. While some have suggested that T2 imaging is useful in diagnosing RPF, the variable appearance depending on the phase of fibrosis makes this a less reliable imaging tool.

Biopsy can be performed to confirm the diagnosis [1]. The gross appearance of RPF is a white plaque of varying thickness. Pathologically, the biopsy would show dense fibrous tissue with inflammatory infiltrate consisting of lymphocytes, macrophages, and plasma cells [10].

Medical treatment of idiopathic RPF is accomplished through the use of immunosuppressive agents [2]. The most well-documented treatment regimens exist for those with IgG4 positive disease, but in general, response to medical treatment is extremely variable. Urinary tract decompression with ureteral stents or nephrostomy tubes is often necessary. Surgical ureterolysis is often necessary unless an excellent response to medical therapy is achieved.

This case demonstrates the utility of MRI in making the diagnosis and evaluating the treatment response in pediatric patients with RPF. As is often the case, patients may initially present with renal impairment making noncontrast MRI an ideal initial imaging modality. Once renal function

normalizes, contrast can be utilized for further follow-up. We found that 15-minute delayed post-contrast imaging was very useful in distinguishing the margins of the fibrosis and evaluating treatment response.

While the presentation and clinical course of this patient were fairly typical for patients with RPF, it is very unusual to see this disease process in a pediatric patient. It is important for physicians to be aware that while rare, this disease process can occur in children, and should be considered in the differential diagnosis for patients who present with obstructive urinary tract findings.

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## Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:[10.1016/j.radcr.2019.01.006](https://doi.org/10.1016/j.radcr.2019.01.006).

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