

# HOSPITAL PHYSICIAN®

## UROLOGY BOARD REVIEW MANUAL

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The *Hospital Physician Urology Board Review Manual* is a peer-reviewed study guide for residents and practicing physicians preparing for board examinations in urology. Each manual reviews a topic essential to the current practice of urology.

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## Retroperitoneal Fibrosis

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# Retroperitoneal Fibrosis

David A. Duchene, MD

## INTRODUCTION

Retroperitoneal fibrosis (RPF) is defined as an inflammatory and fibrotic mass that develops in the retroperitoneum. These masses can be malignant, benign, or iatrogenic. Malignant conditions are usually classified by the primary malignancy and considered metastatic lesions with a desmoplastic reaction as opposed to RPF. The remaining cases of RPF are either a primary condition or secondary to a previous condition or treatment, such as radiation therapy to the retroperitoneum. The most common primary cause of benign RPF is termed *idiopathic RPF*.

Idiopathic RPF is an uncommon disease of unclear etiology in which a fibrotic process causes some degree of compression of the retroperitoneal organs. Albarán<sup>1</sup> first described the disease in 1905, but it was not until Ormond<sup>2</sup> reported cases of RPF in 1948 that it became a recognized clinical entity. A variety of terms have been used to describe the disorder, including Ormond's disease, periureteritis fibrosa, periureteritis plastica, chronic periureteritis, sclerosing retroperitoneal granuloma, and fibrous retroperitonitis. The disease became acknowledged as RPF in the 1960s, and this remains the current preferred term.

Because of the typical patient presentation, urologists have an integral role in the diagnosis and management of patients with RPF. Urologists usually become involved when the mass extends laterally to envelop the ureters and causes extrinsic compression or interference with peristalsis.<sup>3,4</sup> Idiopathic RPF most often occurs in patients aged 40 to 60 years, although it has been reported in children. RPF tends to afflict men 2 to 3 times more frequently than women.

## ETIOLOGY AND PATHOLOGY

### CASE 1

A 67-year-old man with a previously diagnosed 4.3-cm abdominal aortic aneurysm (AAA) undergoes a scheduled surveillance computed tomography (CT)

scan, which demonstrates a 4.3 cm × 4 cm infrarenal AAA with a 2-cm circumferential layer of soft tissue mass surrounding it and encompassing the ureters. The CT scan also demonstrates moderate hydronephrosis of the right collecting system and minimal hydronephrosis of the left collecting system. The radiologist's interpretation of these findings is RPF, inflammatory aneurysm, or a small aneurysmal leak.

- **What are the identifiable causes of RPF? What is the proposed etiology of idiopathic RPF?**

### IDENTIFIABLE CAUSES OF A RETROPERITONEAL MASS Malignancy

Because it can account for ureteral obstruction in approximately 10% of cases,<sup>5</sup> malignancy (primary or metastatic) must be excluded as the etiology of the retroperitoneal mass prior to assigning a diagnosis of RPF. Lymphoma is the most common primary neoplasm in the differential diagnosis, with carcinoid, multiple myeloma, and sarcoma less commonly identified.<sup>6,7</sup> Metastatic lesions from pancreatic, prostatic, rectal, colon, breast, and gastric cancer can also involve the retroperitoneum. Malignancy can usually be identified by the appearance or association of retroperitoneal lymphadenopathy, but occasionally the malignancy will have the flat, infiltrating mass appearance of RPF.

### Benign Retroperitoneal Fibrosis

An identifiable cause of benign RPF is found in only approximately 30% of patients.<sup>4</sup> All other patients fall under the category of idiopathic RPF. The most common known cause of benign RPF is prolonged use of medications, such as methysergide and other ergot alkaloids (ie, lysergic acid diethylamide [LSD]). Methysergide was once used to prevent recurrent migraine headaches but now is not commonly prescribed. The reported incidence of RPF in long-term methysergide users is 1%.<sup>8</sup> Other medications thought to be involved in the development of RPF include  $\beta$ -blockers,  $\alpha$ -blockers (ie, methyldopa, hydralazine), dopaminergic agonists (ie, pergolide, pramipexol), haloperidol, amphetamines, and phenacetin.<sup>9-11</sup> The pathophysiology of drug-induced RPF is unknown. Ergot alkaloids