Figure 21-2 Papillary transitional cell carcinoma extensively involving the ureter. (Courtesy Dr. Cristina Magi-Galluzzi, The Johns Hopkins Hospital, Baltimore, Md.)

Unilateral obstruction typically results from proximal causes, whereas bilateral obstruction arises from distal causes, such as nodular hyperplasia of the prostate. Only sclerosing retroperitoneal fibrosis is discussed further.

Sclerosing Retroperitoneal Fibrosis. This uncommon cause of ureteral narrowing or obstruction is characterized by a fibrotic proliferative inflammatory process encasing the retroperitoneal structures and causing hydronephrosis. The disorder occurs in middle to late age and is more common in males than females. At least a subset of these cases is related to *IgG4-related disease*, a recently described entity associated with elevated levels serum IgG4 and fibroinflammatory lesions rich in IgG4-secreting plasma cells (Chapter 6). In addition to the retroperitoneum, this disorder often involves other tissues as well, particularly exocrine organs such as the pancreas and salivary glands. Other cases of retroperitoneal fibrosis are associated with drug exposures (ergot derivatives, β-adrenergic blockers), adjacent inflammatory conditions (vasculitis, diverticulitis, Crohn disease), or malignant disease (lymphomas, urinary tract carcinomas). Most, however, have no obvious cause and are considered primary or idiopathic (Ormond disease).

Microscopic examination typically reveals fibrous tissue containing a prominent infiltrate of lymphocytes, often with germinal centers, plasma cells (frequently IgG4-positive), and eosinophils. Treatment initially involves corticosteroids, although many patients eventually become resistant and require ureteral stents or surgical extrication of the ureters from the surrounding fibrous tissue (ureterolysis).

Table 21-1 Major Causes of Ureteral Obstruction

Type of Obstruction	Cause
Intrinsic	
Calculi	Of renal origin, rarely more than 5 mm in diameter Larger renal stones cannot enter ureters Impact at loci of ureteral narrowing—ureteropelvic junction, where ureters cross iliac vessels, and where they enter bladder—and cause excruciating "renal colic"
Strictures	Congenital or acquired (inflammations)
Tumors	Transitional cell carcinomas arising in ureters Rarely, benign tumors or fibroepithelial polyps
Blood clots	Massive hematuria from renal calculi, tumors, or papillary necrosis
Neurogenic	Interruption of the neural pathways to the bladder
Extrinsic	
Pregnancy	Physiologic relaxation of smooth muscle or pressure on ureters at pelvic brim from enlarging fundus
Periureteral inflammation	Salpingitis, diverticulitis, peritonitis, sclerosing retroperitoneal fibrosis
Endometriosis	With pelvic lesions, followed by scarring
Tumors	Cancers of the rectum, bladder, prostate, ovaries, uterus, cervix; lymphomas, sarcomas

KEY CONCEPTS

Disorders of the Ureters

- Ureteral obstruction is clinically significant because it can subsequently involve the kidney (hydronephrosis or even pyelonephritis), compromising renal function.
- In children, congenital ureteropelvic junction (UPJ) obstruction is the most common obstructive lesion in the ureter.
- In adults, ureteral obstruction may be acute (e.g., due to obstructing calculi), or chronic (e.g., due to intrinsic or extrinsic tumors or rarely idiopathic conditions such as sclerosing retroperitoneal fibrosis).

Urinary Bladder

Diseases of the bladde are often disabling but rarely lethal. Cystitis is particularly common in young women of reproductive age. Tumors of the bladder are an important source of both morbidity and mortality.

Congenital Anomalies

- **Vesicoureteral reflux** is the most common and serious congenital anomaly. As a major contributor to renal infection and scarring, it was discussed earlier, in Chapter 20, in the consideration of pyelonephritis. Abnormal connections between the bladder and the vagina, rectum, or uterus may create *congenital vesicouterine fistulae.*
- **Diverticula** are pouchlike evaginations of the bladder wall that vary from less than 1 cm to 5 to 10 cm in diameter and may be congenital or acquired. *Congenital diverticula* may be due to a focal failure of development of the normal musculature or to some urinary tract obstruction during fetal development. *Acquired diverticula* are most often seen with prostatic enlargement (hyperplasia or neoplasia), producing obstruction to urine outflow and marked muscle thickening of the bladder wall. The increased intravesical pressure causes outpouching of the bladder wall and the formation of diverticula. They are frequently multiple and have narrow necks located