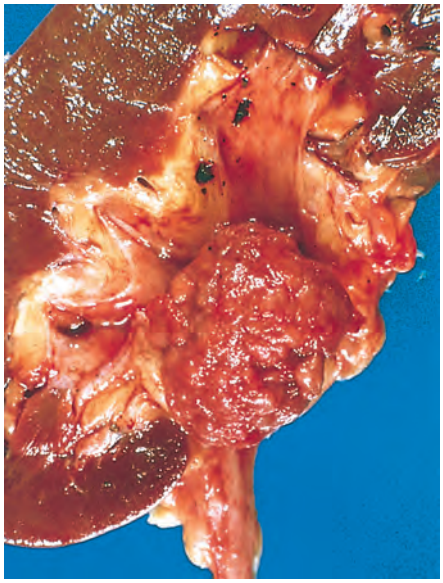

Figure 20-53 Urothelial carcinoma of the renal pelvis. The pelvis has been opened to expose the nodular irregular neoplasm, just proximal to the ureter.

pelvis and, by fragmentation, produce noticeable hematuria. They are almost invariably small when discovered. These tumors may block the urinary outflow and lead to palpable hydronephrosis and flank pain. On histologic examination, pelvic tumors are the exact counterpart of those found in the urinary bladder; further details are in Chapter 21.

Urothelial tumors may occasionally be multiple, involving the pelvis, ureters, and bladder. In 50% of renal pelvic tumors there is a preexisting or concomitant bladder urothelial tumor. On histologic examination, there are also foci of atypia or carcinoma in situ in grossly normal urothelium remote from the pelvic tumor. There is an increased incidence of urothelial carcinomas of the renal pelvis in individuals with Lynch syndrome and analgesic nephropathy.

Infiltration of the wall of the pelvis and calyces is common. For this reason, despite their apparently small, deceptively benign appearance, the prognosis for these tumors is not good. Reported 5-year survival rates vary from 50% to 100% for low-grade noninvasive lesions to 10% with high-grade infiltrating tumors.

KEY CONCEPTS

Kidney Neoplasms

- Clear cell renal cell carcinoma is the most common subtype of malignant renal neoplasms, which often involves *VHL*, a tumor suppressor gene.
- Papillary renal cell carcinoma is the second most common subtype of malignant renal neoplasms, which may involve the *MET* proto-oncogene.
- Hereditary forms of renal cell carcinoma have led to the discovery of important genes (e.g., *VHL*, *BHD*) in renal carcinogenesis.
- Urothelial tumors resembling similar tumors in the urinary bladder can also originate in the renal pelvis. These tumors have a poor prognosis.

SUGGESTED READINGS

Pathogenesis of Immune-Mediated Glomerular Injury

Couser WG: Basic and translational concepts of immune-mediated glomerular diseases. *J Am Soc Nephrol* 23:381–99, 2012. *[An outstanding and comprehensive review of the immunopathogensis of glomerular diseases.]*

Pickering M, Cook HT: Complement and glomerular disease: new insights. *Curr Opin Nephrol Hypertens* 20:271–7, 2011. *[An insightful review into newly emerging concepts of the contribution of complement to renal disease, with an emphasis on dysregulation of the alternate pathway of complement activation.]*

Mechanisms of Progression in Glomerular Diseases

Schlondorff DO: Overview of factors contributing to the pathophysiology of progressive renal disease. *Kidney Int* 74:860–6, 2008. *[An excellent summary of complex and interacting pathways that lead to progressive chronic kidney injury.]*

Nephrotic Syndrome

D'Agati VD, Kaskel FJ, Falk RJ: Focal segmental glomerulosclerosis. *N Engl J Med* 365:2398–411, 2011. *[An excellent and well-illustrated review of pathologic and clinical characteristics of this group of diseases.]*

Jefferson JA, Nelson PJ, Najafian B, et al: Podocyte disorders: Core Curriculum 2011. *Am J Kidney Dis* 58:666–77, 2011. *[An excellent clinically oriented synopsis of alternations in podocytes that underly multiple types of glomerular disease.]*

Ronco P, Debiec H: Pathogenesis of membranous nephropathy: recent advances and future challenges. *Nat Rev Nephrol* 8:203–13, 2012. *[Comprehensive review of the immunopathology of membranous nephropathy by two of the foremost investigators in the identification of the target antigens in this disease.]*

Sethi S, Fervenza FC: Membranoproliferative glomerulonephritis–a new look at an old entity. *N Engl J Med* 366:1119–31, 2012. *[An excellent reassessment of traditional classifications of this entity with a review of new classification schema that are based on underlying pathogenesis.]*

Nephritic Syndrome

Nast CC: Infection-related glomerulonephritis: changing demographics and outcomes. *Adv Chronic Kidney Dis* 19:68–75, 2012. *[A thoughtful review that integrates long established concepts in infection related glomerulonephritis with changing patterns of disease expression in both first and third world societies.]*

Rapidly Progressive (Crescentic) Glomerulonephritis

Jennette JC, Falk RJ, Gasim AH: Pathogenesis of antineutrophil cytoplasmic autoantibody vasculitis. *Curr Opin Nephrol Hypertens* 20:263–70, 2011. *[A short but thorough review of this major form of vasculitis with an emphasis on underlying pathophysiology.]*

Kambham N: Crescentic Glomerulonephritis: an update on Pauci-immune and Anti-GBM diseases. *Adv Anat Pathol* 19(2):111–24, 2012. *[A current review of the clinico-pathologic manifestations of these diseases.]*

Tarzi RM, Cook HT, Pusey CD: Crescentic glomerulonephritis: new aspects of pathogenesis. *Semin Nephrol* 31:361–8, 2011. *[An excellent consideration of pathogenesis of crescentic glomerulonephritis that complements the review of vasculitis by Jennette, et al.]*

Isolated Urinary Abnormalities

Gubler MC: Inherited diseases of the glomerular basement membrane. *Nat Clin Pract Nephrol* 4:24–37, 2008. *[A detailed review of the pathophysiology of hereditary nephritides and a clear guide to their identification and differential diagnosis.]*

Suzuki H, Kiryluk K, Novak J, et al: J The pathophysiology of IgA nephropathy. *Am Soc Nephrol* 22:1795–803, 2011. *[An excellent review that details emerging concepts of the pathophysiology of this disorder.]*

Acute Tubular Injury/Necrosis

Bellomo R, Kellum JA, Ronco C: Acute kidney injury. *Lancet* 380:756–66, 2012. *[An excellent review of acute kidney injury with an emphasis on the clinical aspects and management issues.]*