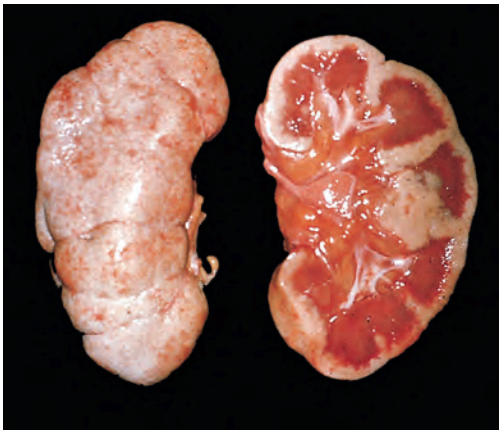

Figure 20-42 Diffuse cortical necrosis. The pale ischemic necrotic areas are confined to the cortex and columns of Bertin.

Diffuse Cortical Necrosis

This uncommon condition occurs most frequently after an obstetric emergency, such as abruptio placentae (premature separation of the placenta), septic shock, or extensive surgery. The cortical destruction has the features of ischemic necrosis. Glomerular and arteriolar microthrombi are often found and contribute to the necrosis and renal damage. The morphologic features considerably overlap with thrombotic microangiopathy and disseminated intravascular coagulation, but the pathogenic sequence of events remains obscure.

MORPHOLOGY

The gross alterations of massive ischemic necrosis are sharply limited to the cortex (Fig. 20-42). The histologic appearance is that of acute ischemic infarction. The lesions may be patchy, with areas of coagulative necrosis and apparently better preserved cortex. Intravascular and intraglomerular thromboses may be prominent but are usually focal, and acute necroses of small arterioles and capillaries may occasionally be present. Hemorrhages occur into the glomeruli, together with the formation of fibrin plugs in the glomerular capillaries.

Massive acute cortical necrosis is of grave significance, since it gives rise to sudden anuria, terminating rapidly in uremic death. Instances of unilateral or patchy involvement are compatible with survival.

Renal Infarcts

The kidneys are common sites for the development of infarcts. Contributing to this predisposition is the extensive blood flow to the kidneys (one fourth of the cardiac output), but probably more important is the limited collateral circulation from extrarenal sites (small blood vessels penetrating from the renal capsule supply only the very outer rim of the cortex). Although thrombosis in advanced atherosclerosis and the acute vasculitis of polyarteritis nodosa may occlude arteries, most infarcts are due to embolism. A major source of such emboli is mural thrombosis in the left atrium and ventricle as a result of myocardial infarction. Vegetative endocarditis, aortic aneurysms, and aortic atherosclerosis are less frequent sources of emboli.

MORPHOLOGY

Because of the lack of a collateral blood supply, most renal infarcts are of the "white" anemic variety. Within 24 hours infarcts become sharply demarcated, pale, yellow-white areas that may contain small irregular foci of hemorrhagic discoloration. They are usually ringed by a zone of intense hyperemia.

The infarcts are wedge-shaped, with the base against the cortical surface and the apex pointing toward the medulla. In time these acute areas of ischemic necrosis undergo progressive fibrous scarring, giving rise to depressed, pale, gray-white scars that assume a V-shape on section. The histologic changes in renal infarcts are those of ischemic coagulative necrosis, described in Chapter 2.

Many renal infarcts are clinically silent. Sometimes, pain with tenderness localized to the costovertebral angle occurs, associated with showers of red cells in the urine. Large infarcts of one kidney are probably associated with narrowing of the renal artery or one of its major branches, which in turn may cause hypertension.

Congenital and Developmental Anomalies

About 10% of people are born with significant malformations of the urinary system. Renal dysplasias and hypoplasias account for 20% of chronic kidney disease in children.

Congenital renal disease can be hereditary but most often results from an acquired developmental defect during gestation. As discussed in Chapter 10, defects in genes involved in normal renal development, including the Wilms tumor-associated genes, understandably cause urogenital anomalies. As a rule, the resulting developmental abnormalities involve structural components of both the kidney and urinary tract. Other genetic defects primarily produce functional abnormalities in tubular transport, such as cystinuria and renal tubular acidosis. Here, we restrict the discussion to structural anomalies involving primarily the kidney. All except horseshoe kidney are uncommon. Anomalies of the lower urinary tract are discussed in Chapter 21.

Agenesis of the Kidney. Bilateral agenesis is incompatible with life, and usually encountered in stillborn infants. It is often associated with other congenital disorders (e.g., limb defects, hypoplastic lungs). Unilateral agenesis is uncommon and compatible with normal life if no other abnormalities exist. The solitary kidney enlarges as a result of compensatory hypertrophy. Some patients eventually develop progressive glomerular sclerosis in the remaining kidney as a result of the adaptive changes in hypertrophied nephrons, discussed earlier in the chapter, and in time, chronic kidney disease ensues.

Hypoplasia. Hypoplasia refers to failure of the kidneys to develop to a normal size. This anomaly may occur bilaterally, resulting in renal failure in early childhood, but it is more commonly encountered as a unilateral defect. True renal hypoplasia is observed in low birth weight infants and may contribute to their increased lifetime risk for chronic kidney disease. Differentiation between