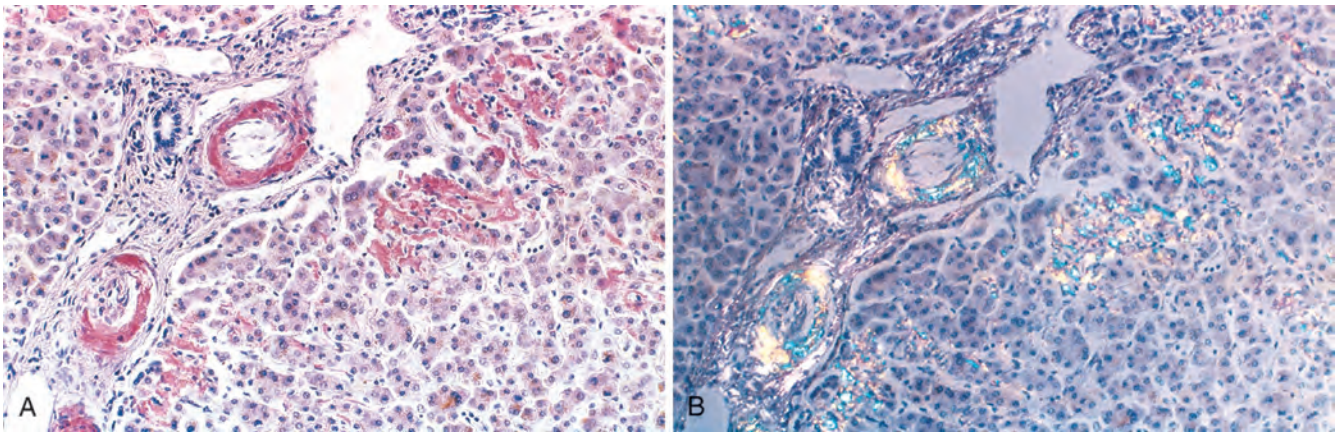

Figure 6-46 Amyloidosis. **A,** A section of the liver stained with Congo red reveals pink-red deposits of amyloid in the walls of blood vessels and along sinusoids. **B,** Note the yellow-green birefringence of the deposits when observed by polarizing microscope. (Courtesy Dr. Trace Worrell and Sandy Hinton, Department of Pathology, University of Texas Southwestern Medical School, Dallas, Texas.)

The pattern of organ involvement in different clinical forms of amyloidosis is variable.

Kidney. Amyloidosis of the kidney is the most common and potentially the most serious form of organ involvement. Grossly, the kidneys may be of normal size and color, or, in advanced cases, they may be shrunken because of ischemia caused by vascular narrowing induced by the deposition of amyloid within arterial and arteriolar walls.

Histologically, the amyloid is deposited primarily in the glomeruli, but the interstitial peritubular tissue, arteries, and arterioles are also affected. The glomerular deposits first appear as subtle thickenings of the mesangial matrix, accompanied usually by uneven widening of the basement membranes of the glomerular capillaries. In time the mesangial depositions and the deposits along the basement membranes cause capillary narrowing and distortion of the glomerular vascular tuft. With progression of the glomerular amyloidosis, the capillary lumens are obliterated, and the obsolescent glomerulus is flooded by confluent masses or interlacing broad ribbons of amyloid (Fig. 6-47).

Spleen. Amyloidosis of the spleen may be inapparent grossly or may cause moderate to marked splenomegaly (up to 800 g). For completely mysterious reasons, one of two patterns of deposition is seen. In one, the deposits are largely limited to the splenic follicles, producing tapioca-like granules on gross inspection, designated sago spleen. In the other pattern, the amyloid involves the walls of the splenic sinuses and connective tissue framework in the red pulp. Fusion of the early deposits gives rise to large, maplike areas of amyloidosis, creating what has been designated lardaceous spleen.

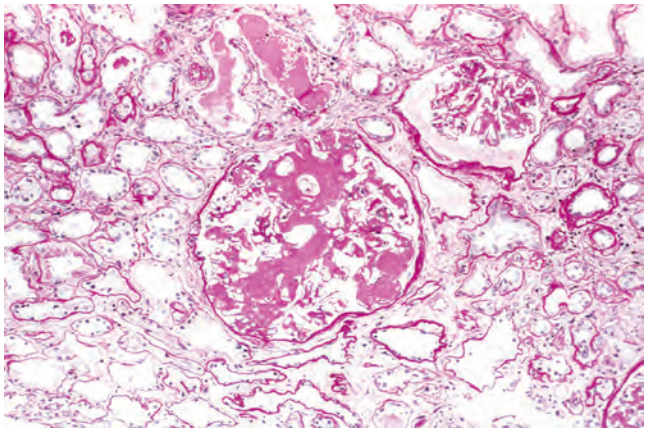

Figure 6-47 Amyloidosis of the kidney. The glomerular architecture is almost totally obliterated by the massive accumulation of amyloid.

Liver. The deposits may be inapparent grossly or may cause moderate to marked hepatomegaly. Amyloid appears first in the space of Disse and then progressively encroaches on adjacent hepatic parenchymal cells and sinusoids (Fig. 6-46). In time, deformity, pressure atrophy, and disappearance of hepatocytes occur, causing total replacement of large areas of liver parenchyma. Vascular involvement and deposits in Kupffer cells are frequent. Normal liver function is usually preserved despite sometimes quite severe involvement of the liver.

Heart. Amyloidosis of the heart (Chapter 12) may occur in any form of systemic amyloidosis. It is also the major organ involved in senile systemic amyloidosis. The heart may be enlarged and firm, but more often it shows no significant changes on gross inspection. Histologically the deposits begin as focal subendocardial accumulations and within the myocardium between the muscle fibers. Expansion of these myocardial deposits eventually causes pressure atrophy of myocardial fibers. When the amyloid deposits are subendocardial, the conduction system may be damaged, accounting for the electrocardiographic abnormalities noted in some patients.

Other Organs. Nodular depositions in the tongue may cause macroglossia, giving rise to the designation tumor-forming amyloid of the tongue. The respiratory tract may be involved focally or diffusely from the larynx down to the smallest bronchioles. As mentioned earlier, a distinct form of amyloid is found in the brains of patients with Alzheimer disease. It may be present in so-called plaques as well as blood vessels (Chapter 28). Amyloidosis of peripheral and autonomic nerves is a feature of several familial amyloidotic neuropathies. Depositions of amyloid in patients on long-term hemodialysis are most prominent in the carpal ligament of the wrist, resulting in compression of the median nerve (carpal tunnel syndrome). These patients may also have extensive amyloid deposition in the joints.