

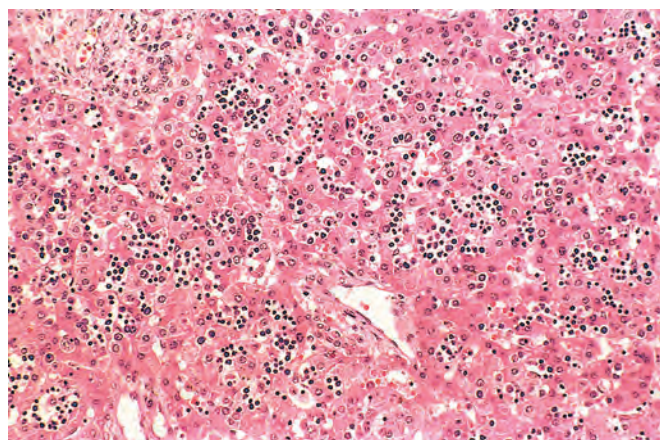


**Figure 10-11** Hydrops fetalis. **A**, There is generalized accumulation of fluid in the fetus. **B**, Fluid accumulation is particularly prominent in the soft tissues of the neck, and this condition has been termed *cystic hygroma*. Cystic hygromas are characteristically seen, but not limited to, constitutional chromosomal anomalies such as 45,X karyotypes. (Courtesy Dr. Beverly Rogers, Department of Pathology, University of Texas Southwestern Medical Center, Dallas, Texas.)

The most serious threat in fetal hydrops is CNS damage, known as **kernicterus** (Fig. 10-13). The affected brain is enlarged and edematous and, when sectioned, has a bright yellow color, particularly the basal ganglia, thalamus, cerebellum, cerebral gray matter, and spinal cord. The precise level of bilirubin that induces kernicterus is unpredictable, but neural damage usually requires a blood bilirubin level greater than 20 mg/dL in term infants; in premature infants this threshold may be considerably lower.

**Clinical Features.** The clinical manifestations of fetal hydrops vary with the severity of the disease and can be inferred from the preceding discussion. Minimally affected

infants display pallor, possibly accompanied by hepatosplenomegaly (to which may be added jaundice with more severe hemolytic reactions), whereas the most gravely ill neonates present with intense jaundice, generalized edema, and signs of neurologic injury. These infants may be supported by a variety of measures, including phototherapy (visual light oxidizes toxic unconjugated bilirubin to harmless, readily excreted, water-soluble dipyrroles) and, in severe cases, total exchange transfusion of the infant.



**Figure 10-12** Numerous islands of extramedullary hematopoiesis (small blue cells) are scattered among mature hepatocytes in the liver of this infant with nonimmune hydrops fetalis.



**Figure 10-13** Kernicterus. Note the yellow discoloration of the brain parenchyma due to bilirubin accumulation, which is most prominent in the basal ganglia deep to the ventricles.