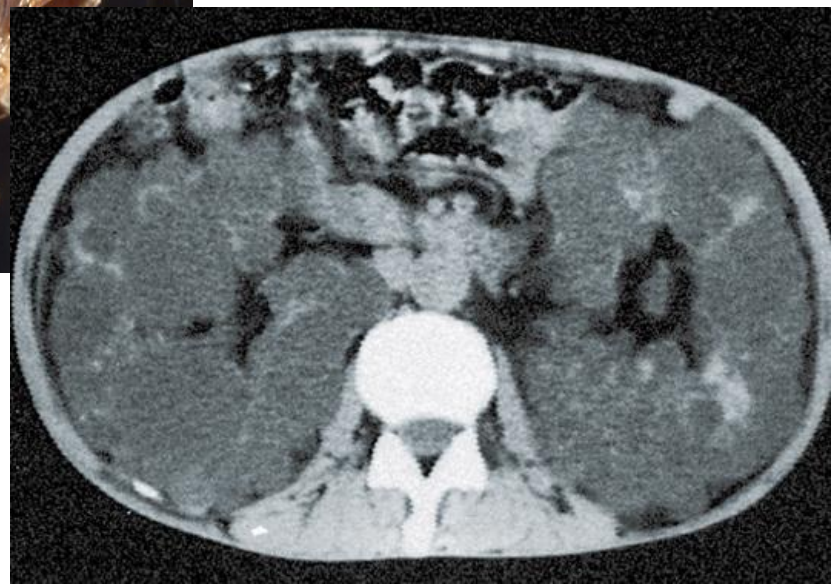
Gross appearance of polycystic kidney diseases

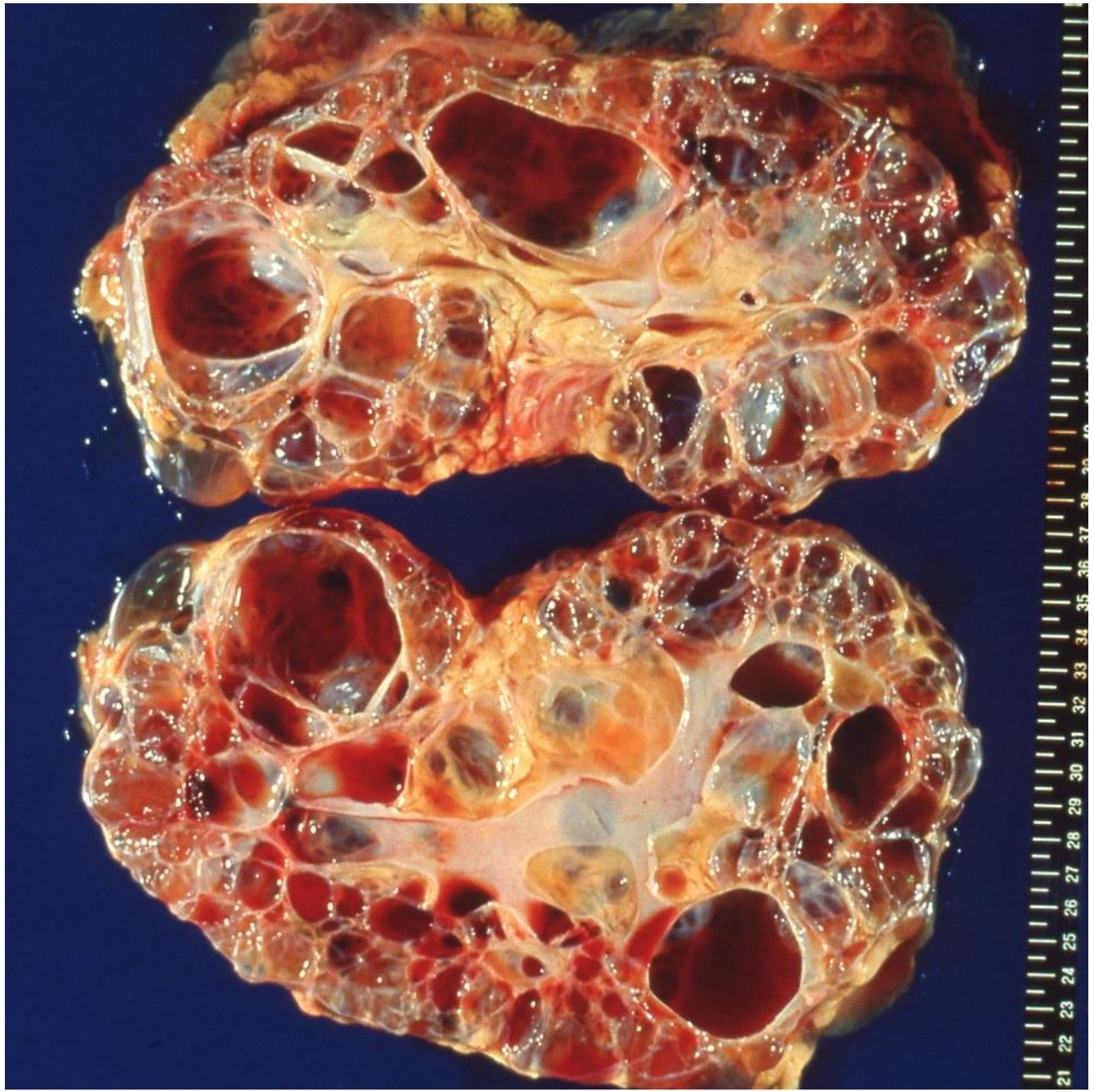
Polycystic kidney disease (PKD) with an autosomal dominant trait get numerous cysts to grow in the markedly enlarged kidneys. The kidneys reduce their function, leading to hypertension and chronic renal failure. Back or side pain is common. Most patients do not develop symptoms until the age of 30 to 40 years. In contrast, infantile or autosomal recessive PKD presents with symptoms in the newborn or even in the uterus. The disease rapidly progresses to be fatal in the first few months of life. The involved kidneys are not enlarged, and may have spongy appearance. It is also called as Potter syndrome or Potter sequence. Classical form of Potter syndrome is, however, caused by bilateral renal agenesis. Acquired polycystic kidney disease happens in the end-stage kidneys on hemodialysis or peritoneal dialysis for years. Multiple small-sized cysts are seen in the atrophic renal parenchyma. Renal cell carcinoma may occur in association with acquired polycystic kidney disease.

Ref.: Harris PC, Torres VE. Polycystic kidney disease. Annu Rev 2009; 60: 321-337. doi: 10.1146/annurev.med.60.101707.125712

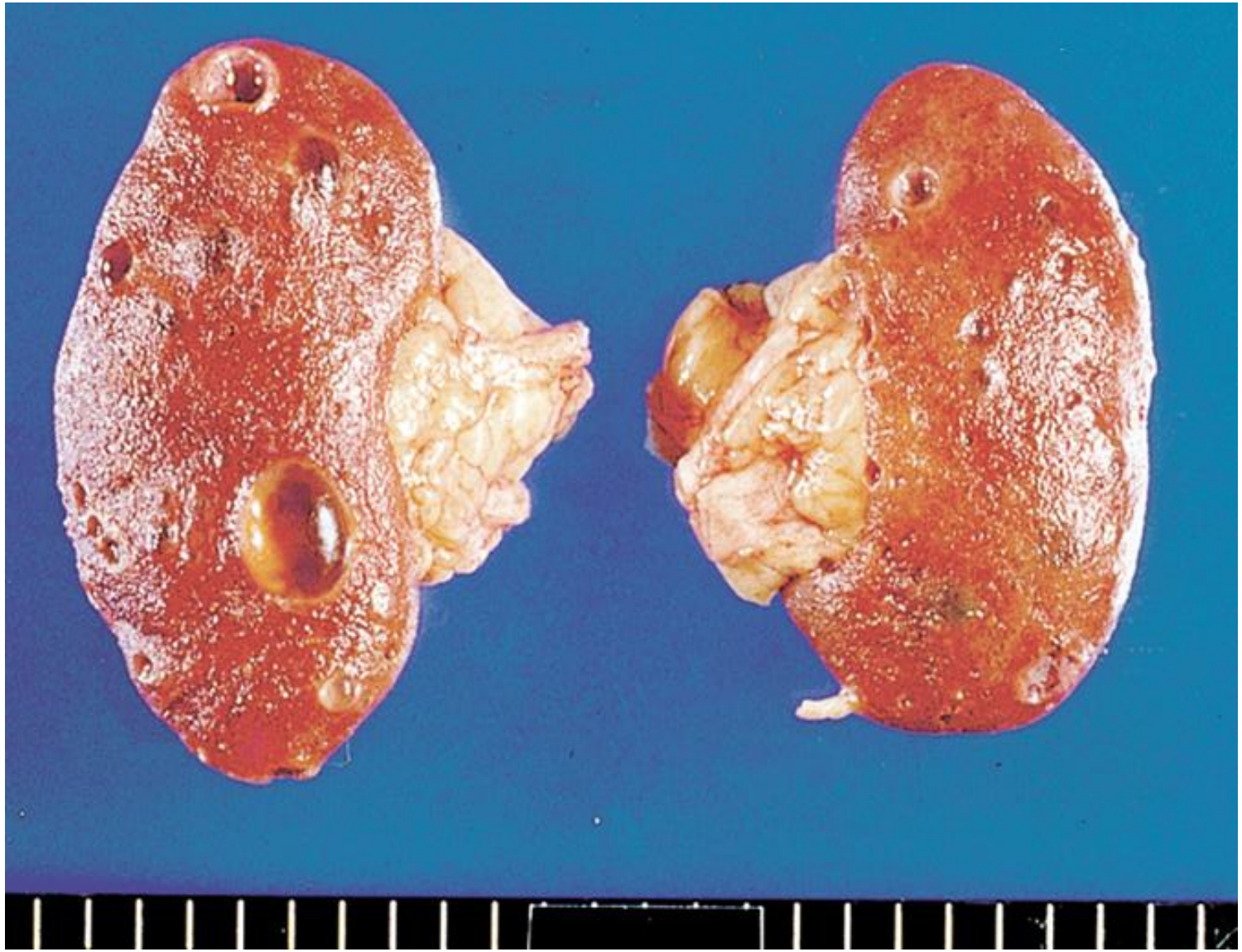
Polycystic kidney disease with autosomal dominant trait presents with huge-sized kidneys replaced by multiple cysts (gross appearance after formalin fixation). The kidney surgically removed from a 40 y-o male patient measures 22 cm in length.

Abdominal CT scan demonstrates markedly enlarged kidneys with numerous cystic structures.

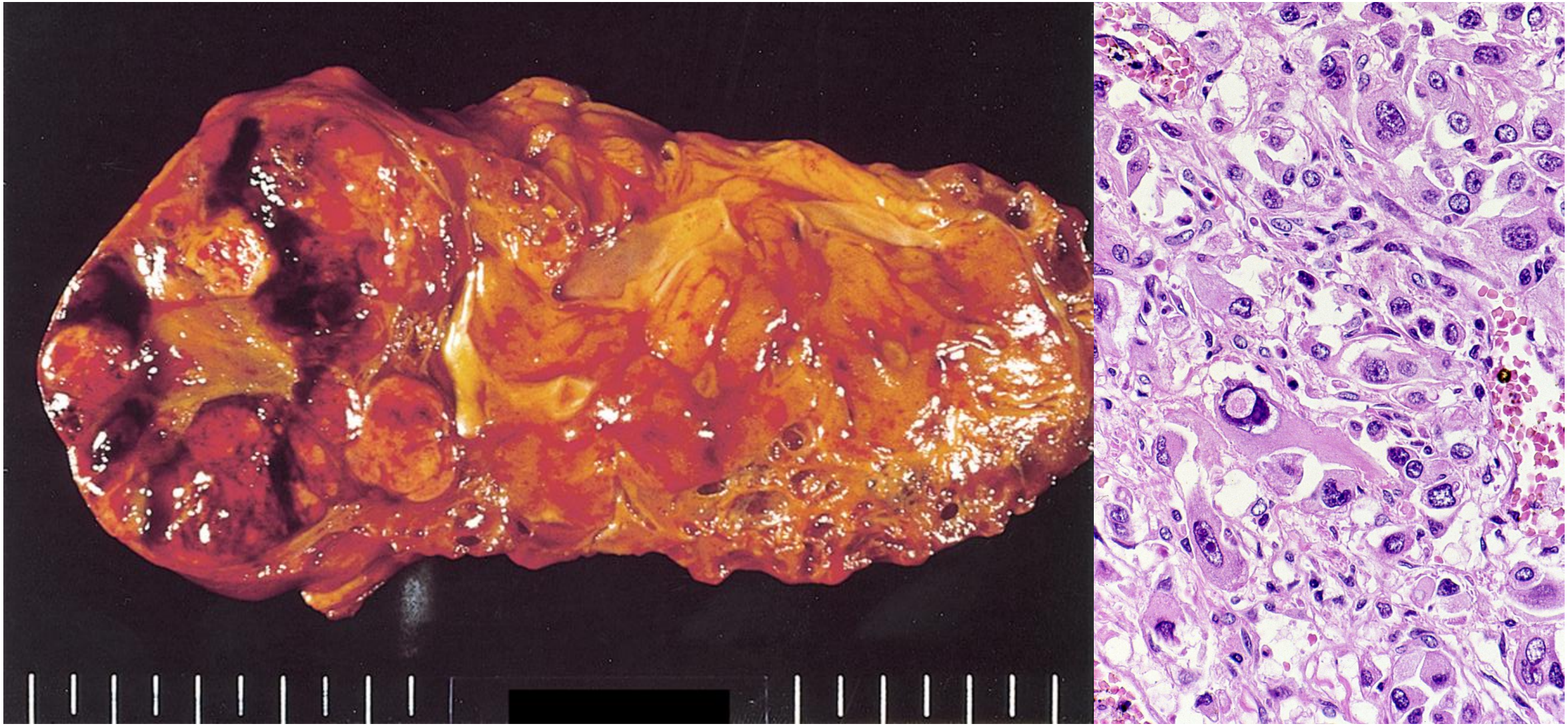




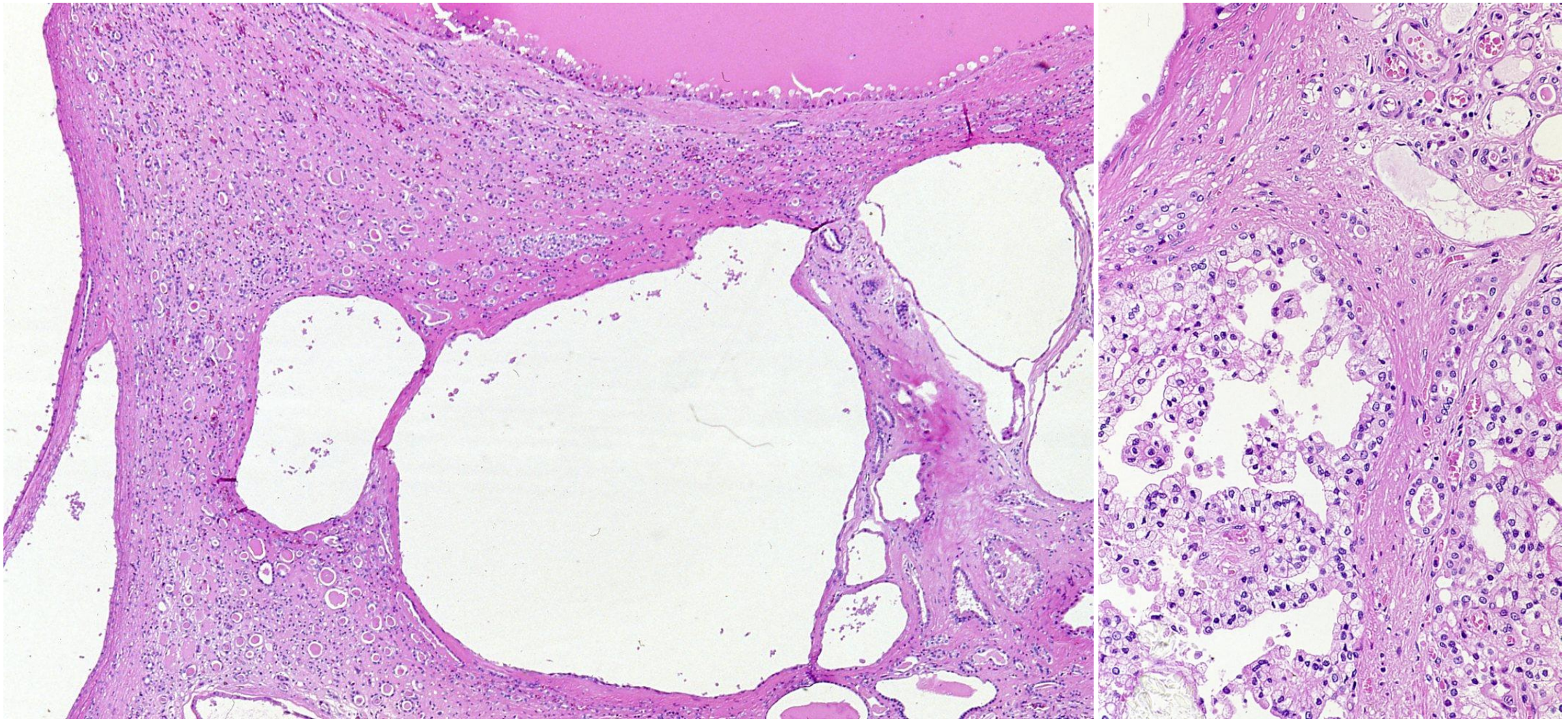
Polycystic kidney disease with autosomal dominant trait presents with huge-sized kidneys replaced by multiple cysts (cut surfaces of bilateral kidneys at autopsy). The kidneys of a female patient aged 50's measures 27 cm in length. Polycystic liver was also observed.



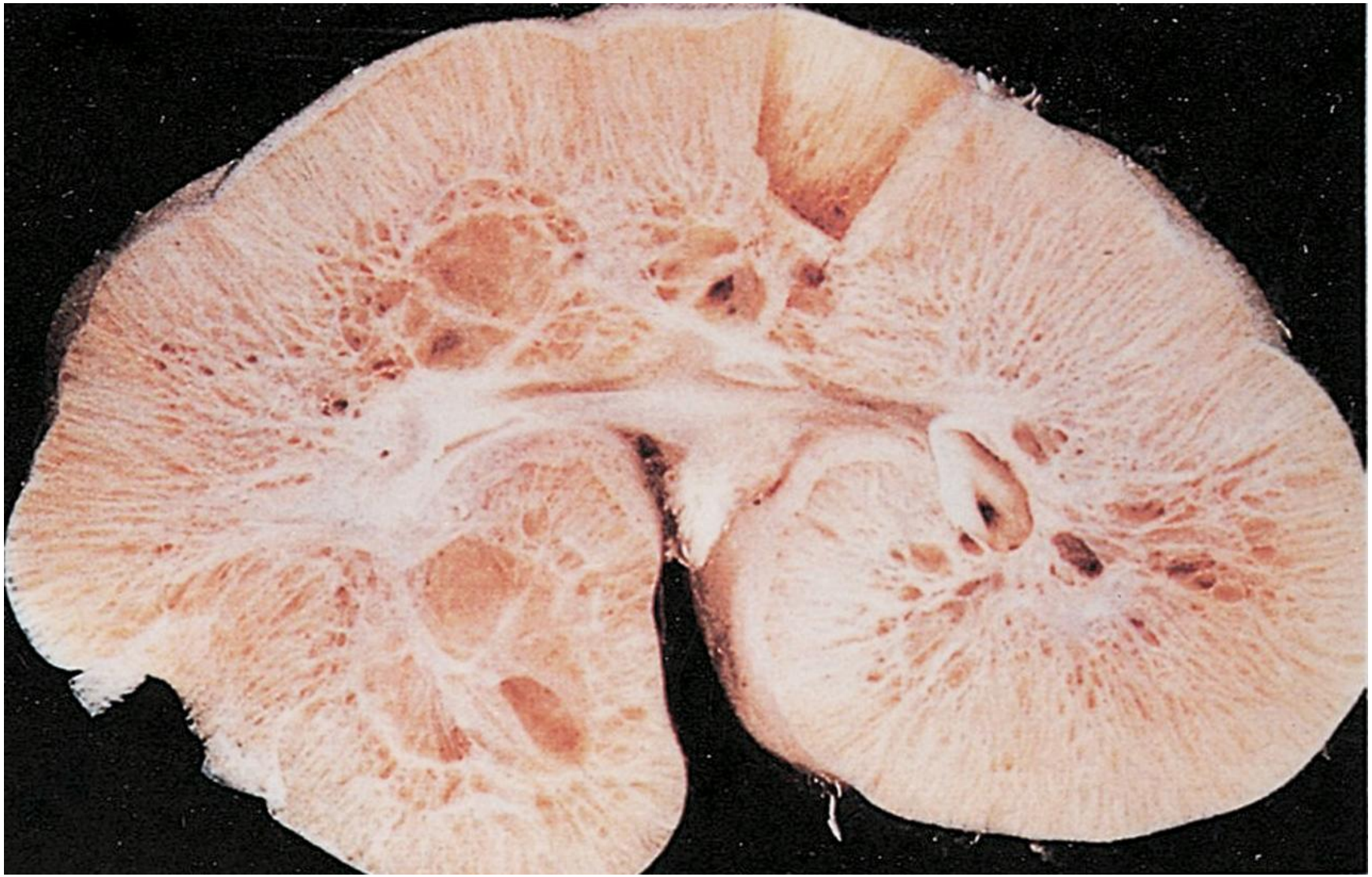
Acquired polycystic kidney seen in the end-stage kidney of a 63 y-o male patient. The patient was on hemodialysis for 13 years. The kidneys weigh 80 g (left) and 70 g (right).



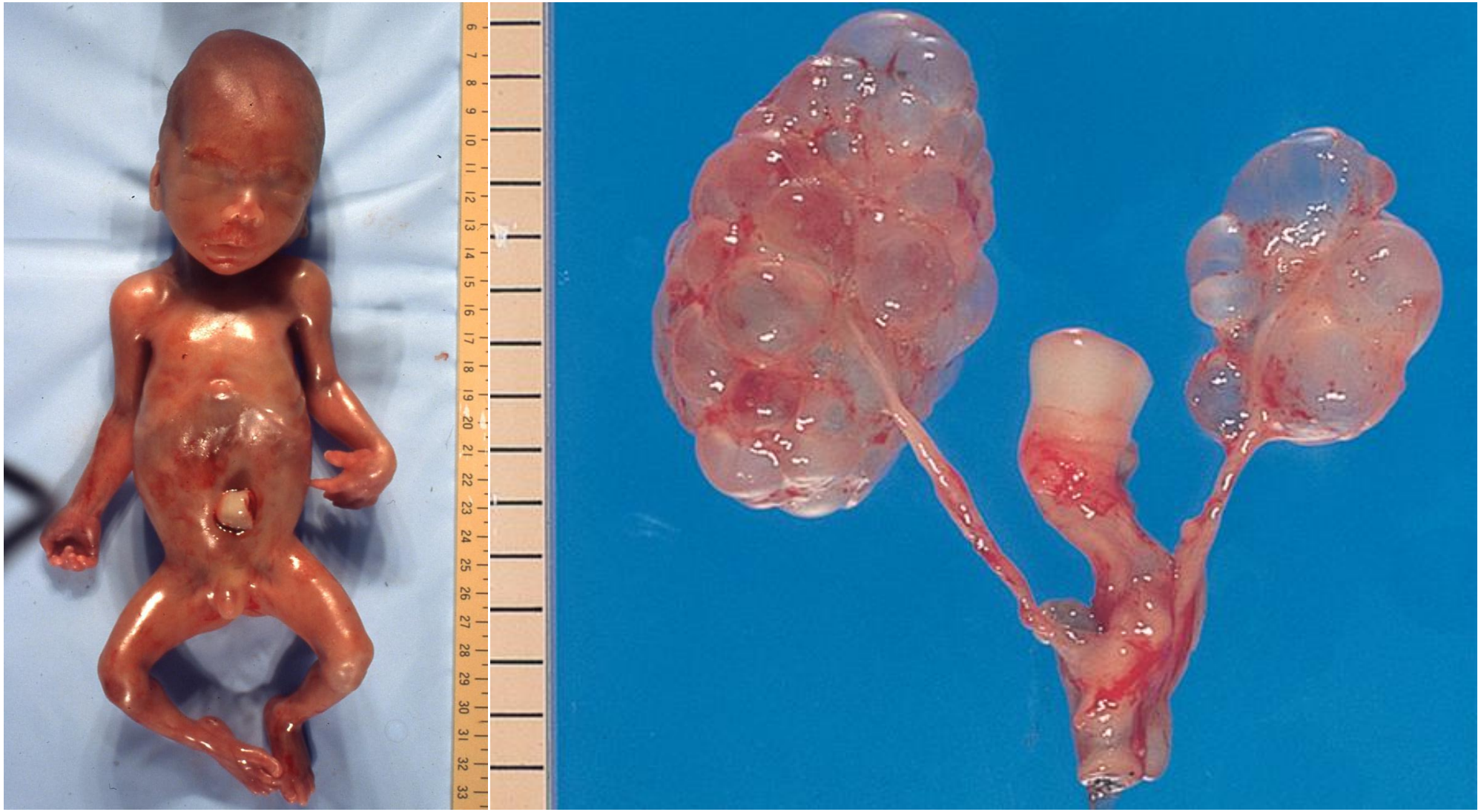
Acquired polycystic kidney seen in the end-stage kidney of a 58 y-o female patient. The patient was on hemodialysis for 16 years. Lethal renal cell carcinoma was complicated in the upper pole of the right kidney (left: gross appearance). Microscopically, high-grade atypia is recognized in renal cell carcinoma (right: H&E). Systemic metastasis killed the patient.



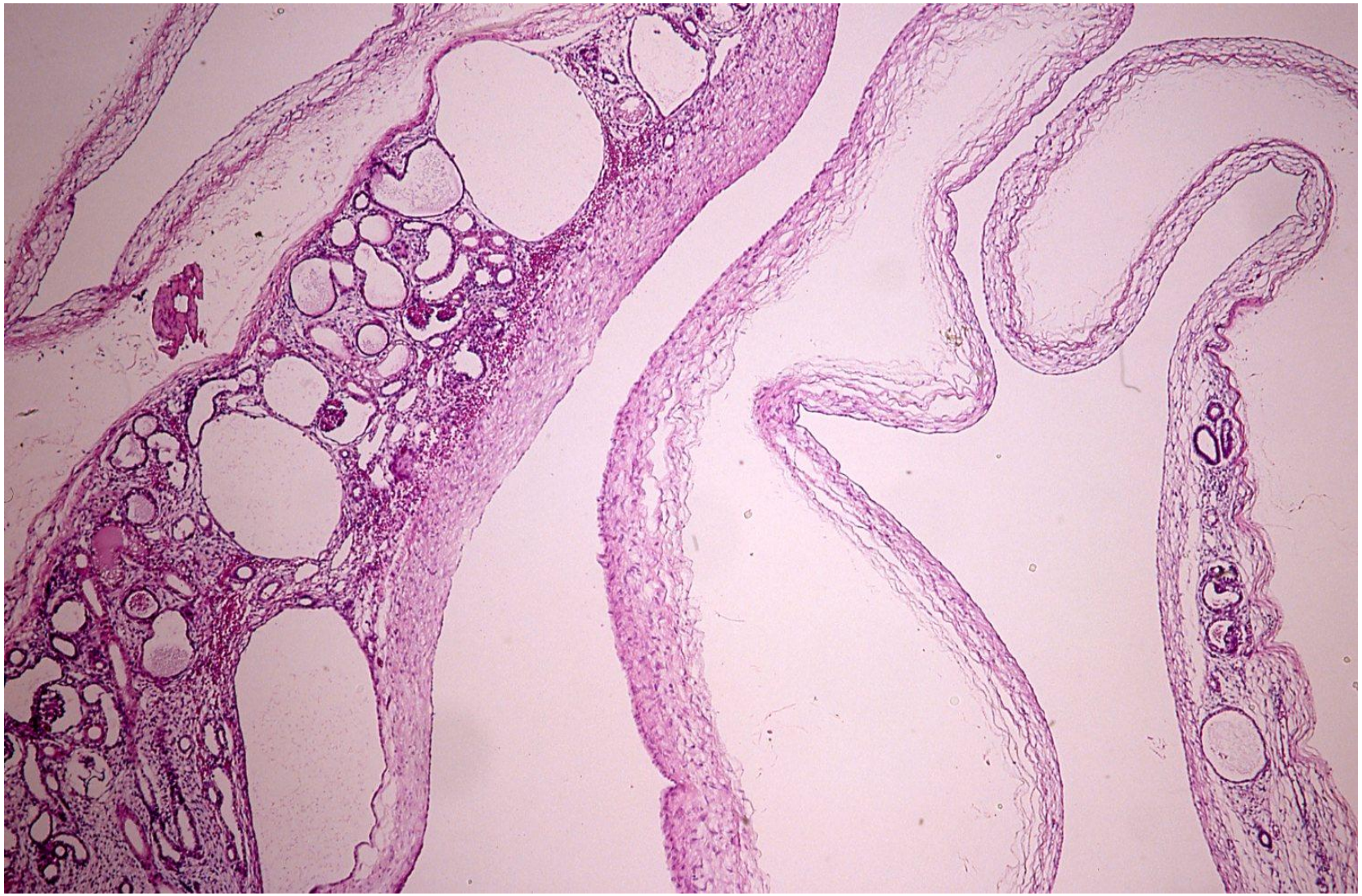
Acquired polycystic kidney seen in the end-stage kidney of a 58 y-o female patient. The patient was on hemodialysis for 16 years. Dilatation of the renal tubules is multifocally observed (left). Papillary hyperplasia of the clear epithelial cells may be seen in the proximal renal tubules (right). Lethal renal cell carcinoma was complicated in the kidney in the present case (H&E).



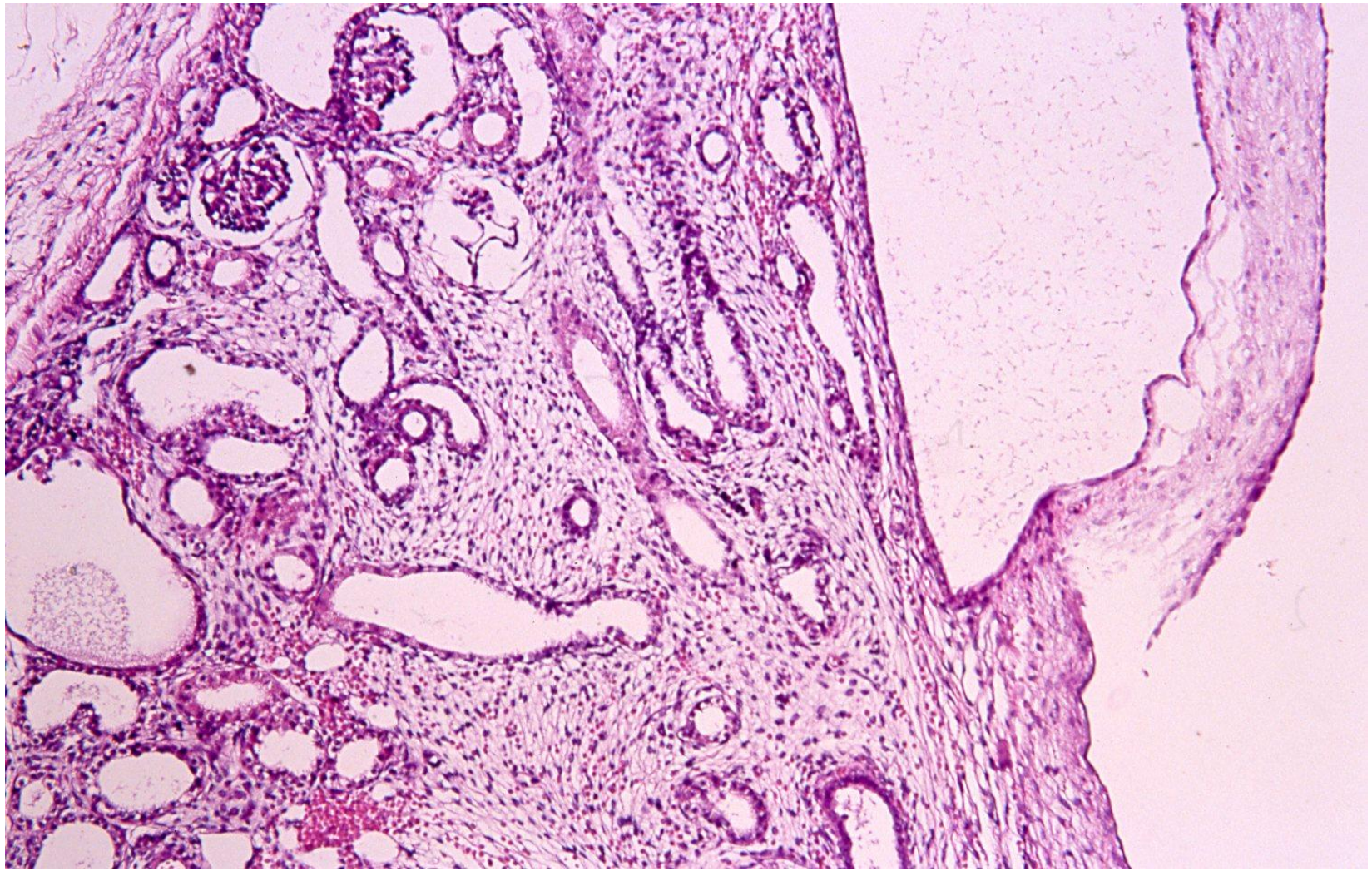
Infantile polycystic kidney (also classified in Potter syndrome type I) seen in a male newborn. It is also called as “sponge kidney” or autosomal recessive polycystic kidney disease (see Uro-78-2-kidney). The kidneys are not enlarged. Renal failure is lethal in the newborn period.



Potter syndrome (type IIa) in a male fetus at the 20th gestational week (body weight: 490 g). This non-hereditary condition accompanies renal dysplasia, causing oligohydramnios due to the inability of urine production. Oligohydramnios causes deformations in morphogenesis, including the Potter's face with parrot beak nose, redundant skin, and skin fold of tissue extending from the medial canthus across the cheek. The extremities show flexion contracture and clubbed feet. The both kidneys reveal polycystic changes (see Uro-81-kidney).



Potter syndrome (type IIa) in a male fetus at the 20th gestational week (body weight: 490 g). Renal dysplasia causes oligohydramnios due to the inability of urine production. Polycystic changes extensively replace the renal parenchyma. Only a few glomeruli are distributed (H&E-1).



Potter syndrome (type IIa) in a male fetus at the 20th gestational week (body weight: 490 g). Renal dysplasia causes oligohydramnios due to the inability of urine production. Polycystic changes extensively replace the renal parenchyma. Only a few glomeruli are distributed (H&E-2).