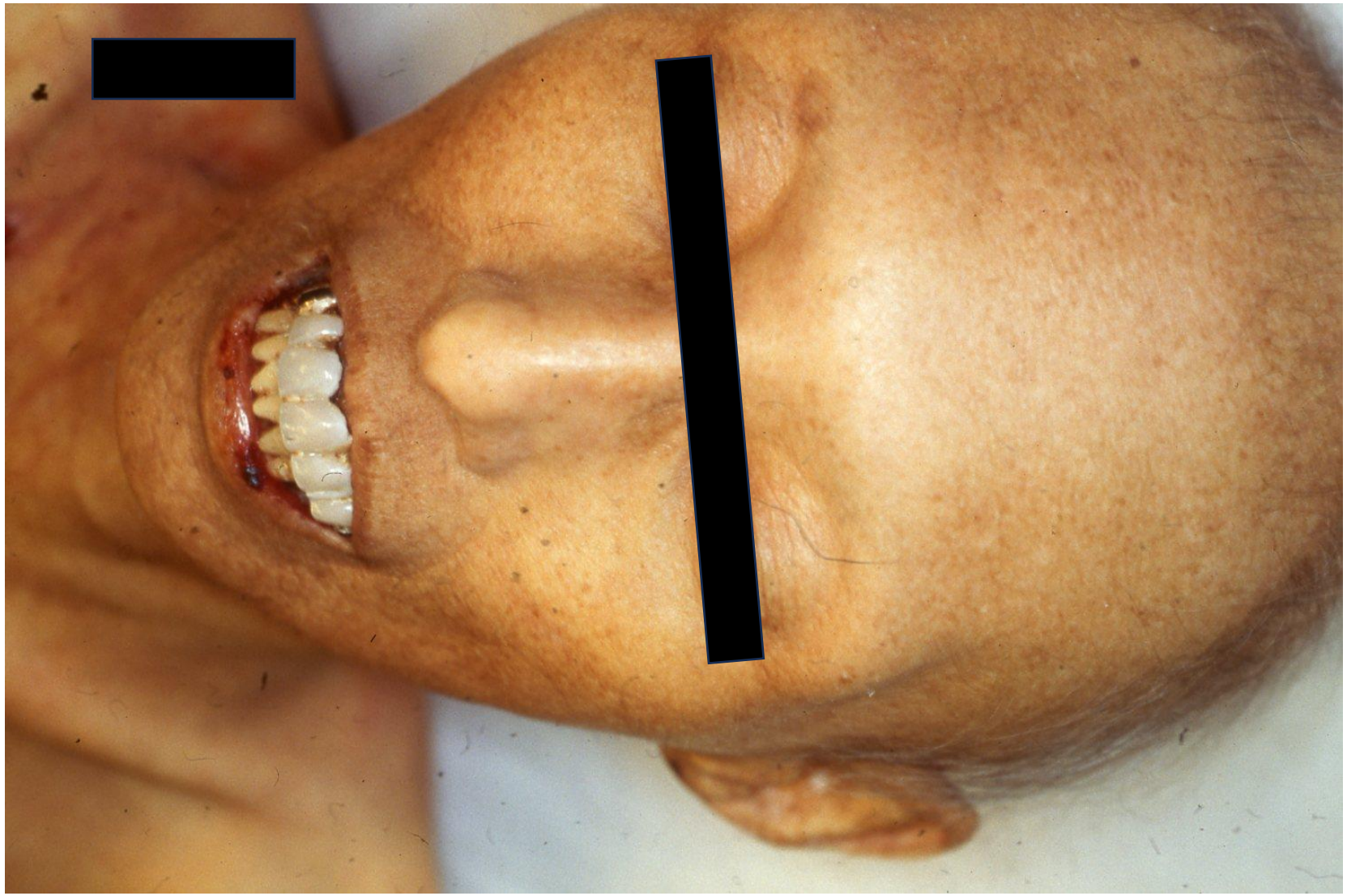
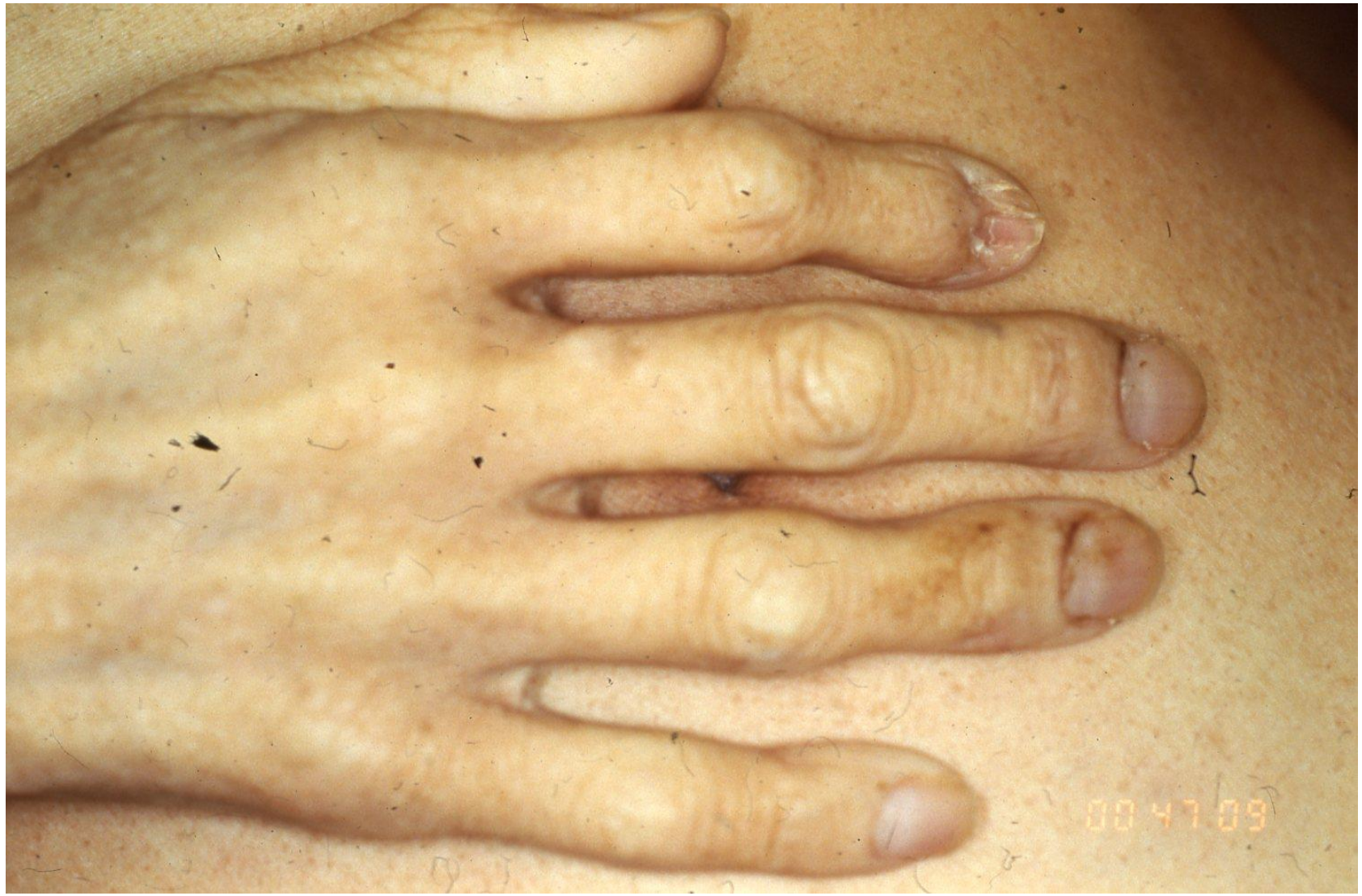
# Werner syndrome (adult progeria)

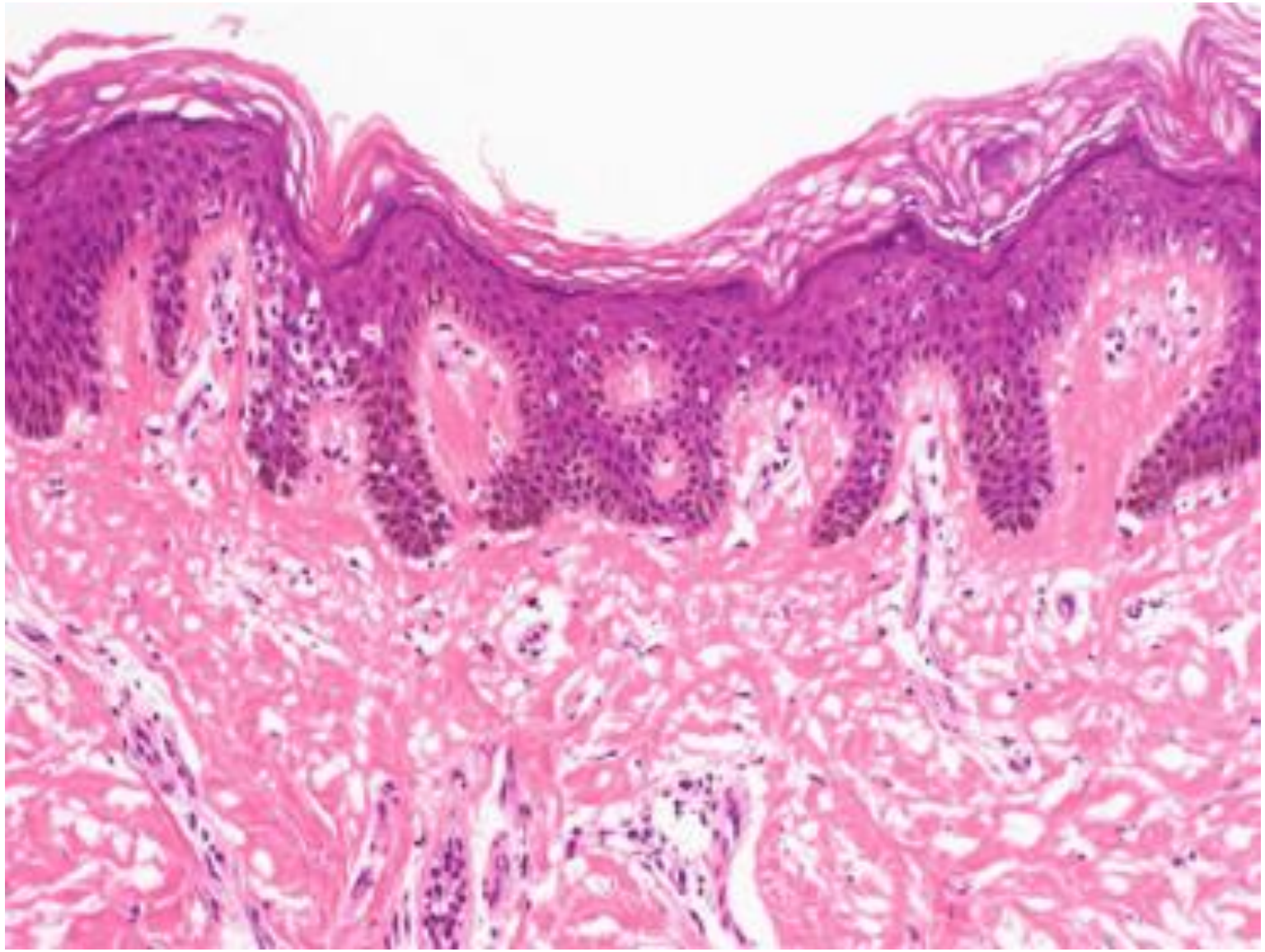
Werner syndrome is a rare autosomal recessive-inherited disorder, characterized by the appearance of unusually accelerated aging (progeria). The disorder is typically recognized by the third or fourth decades of life. The affected individuals have an abnormally slow growth rate and growth stops at puberty, resulting in a short stature and low weight. By age 25, the hair shows early graying and alopecia. As the disease progresses, the patient manifests loss of subcutaneous adipose tissue, atrophy of muscle tissue and skin aging, particularly on the face and distal extremities, with prominent eyes and a beaked or pinched nose. High-pitched voice, calcification of Achilles tendon, bilateral senile cataracts, atherosclerosis, diabetes mellitus and impaired functioning of the ovaries or testes are associated. Intractable skin ulceration is often seen. Malignant tumors may also develop. Two thirds of the cases have been reported from Japan.



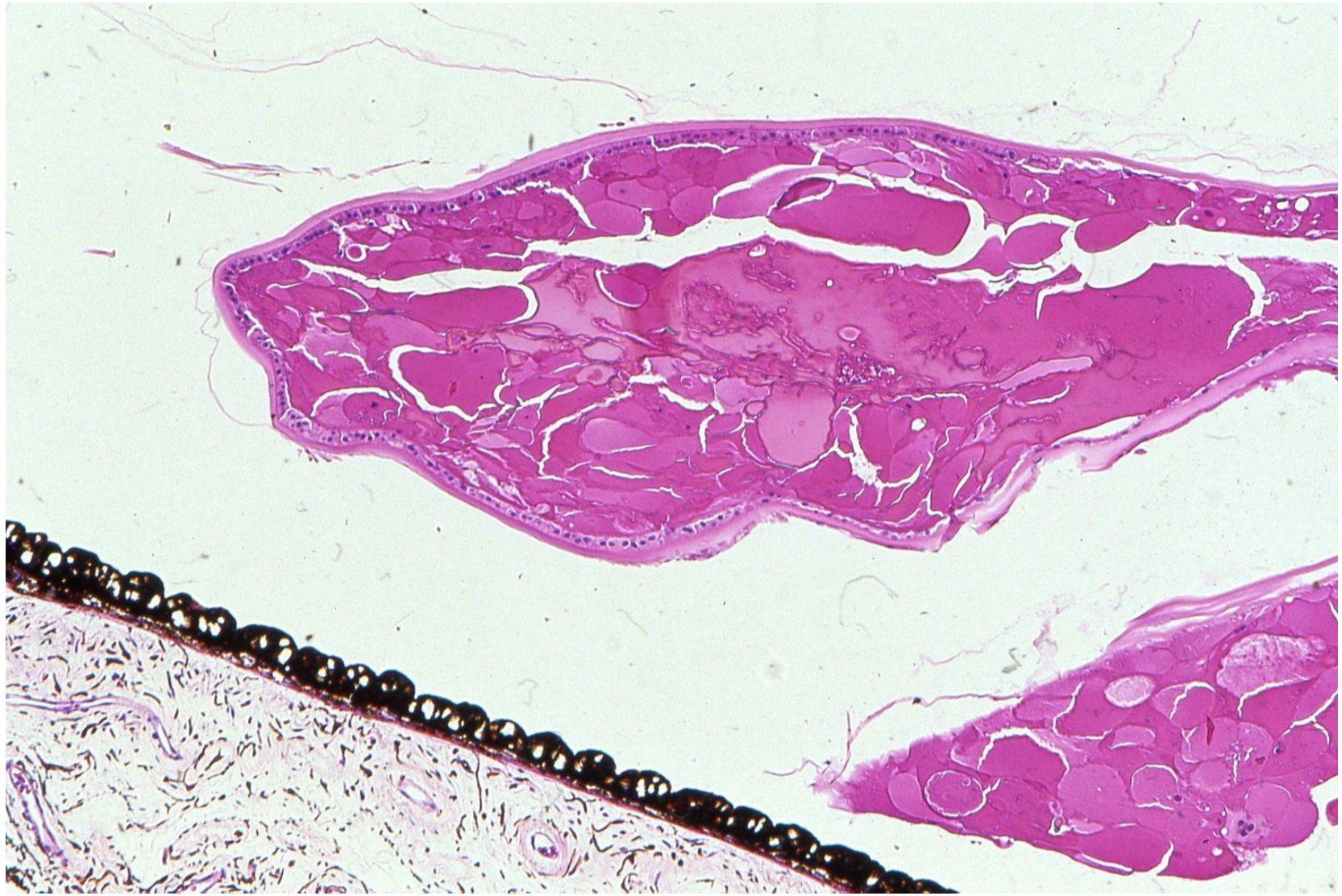
Werner syndrome (adult progeria) in a 42-year-old Japanese male patient.
Skin atrophy/pigmentation and alopecia are observed.  Dentures are put on.



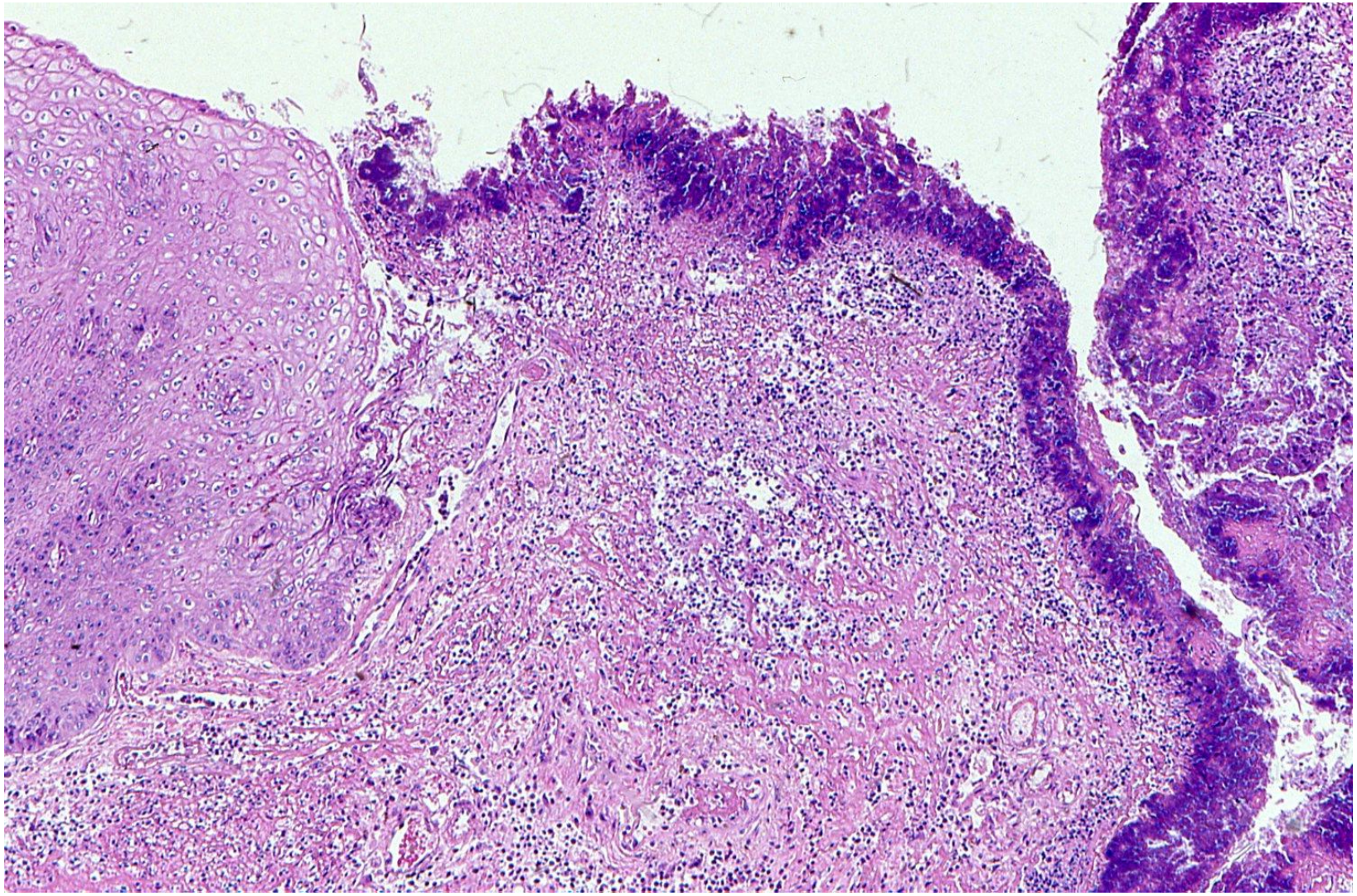

Werner syndrome (adult progeria) in a 42-year-old Japanese male patient. Skin of the hand is atrophic and pigmented, and the nails are deformed.



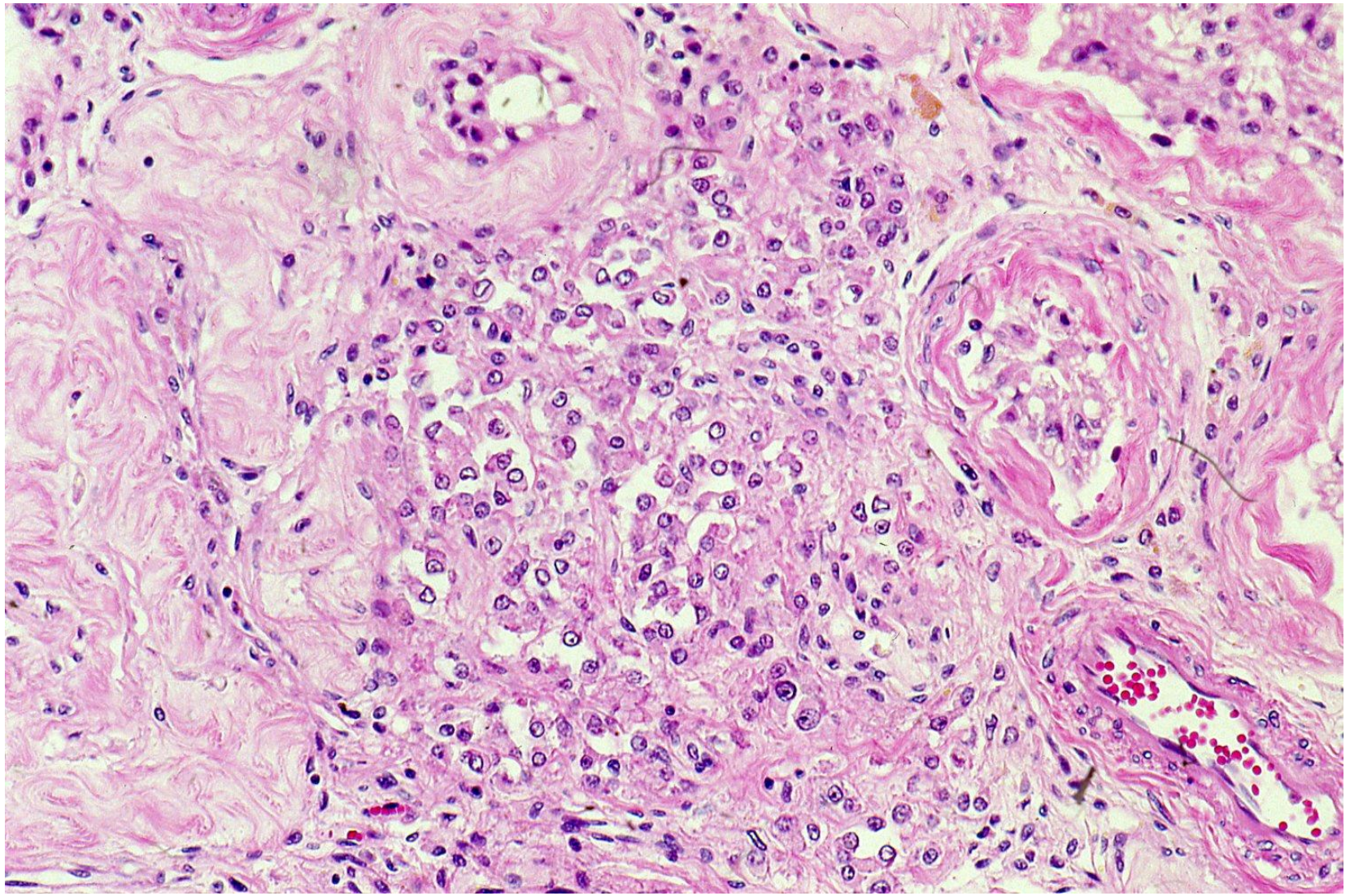
Histology of the skin in Werner syndrome (adult progeria, a 42-year-old Japanese male patient). Increase of melanocytes in the epidermal basal layer and hyaline fibrosis in the dermis are observed. H&E-1



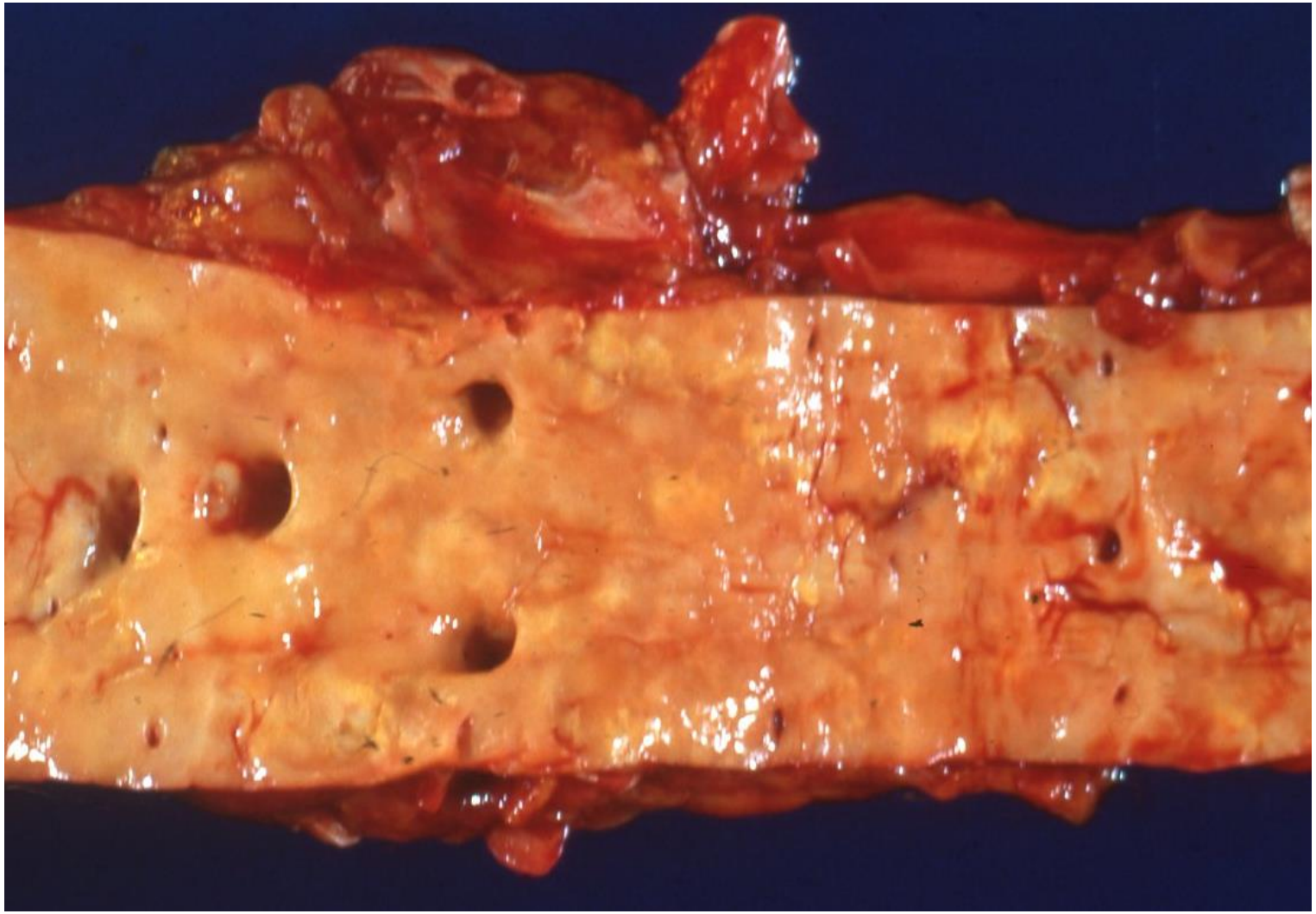
Cataracta is advanced in Werner syndrome (a 42-year-old Japanese male patient). The pigmented iris is seen. The patient has lost the vision. H&E-2



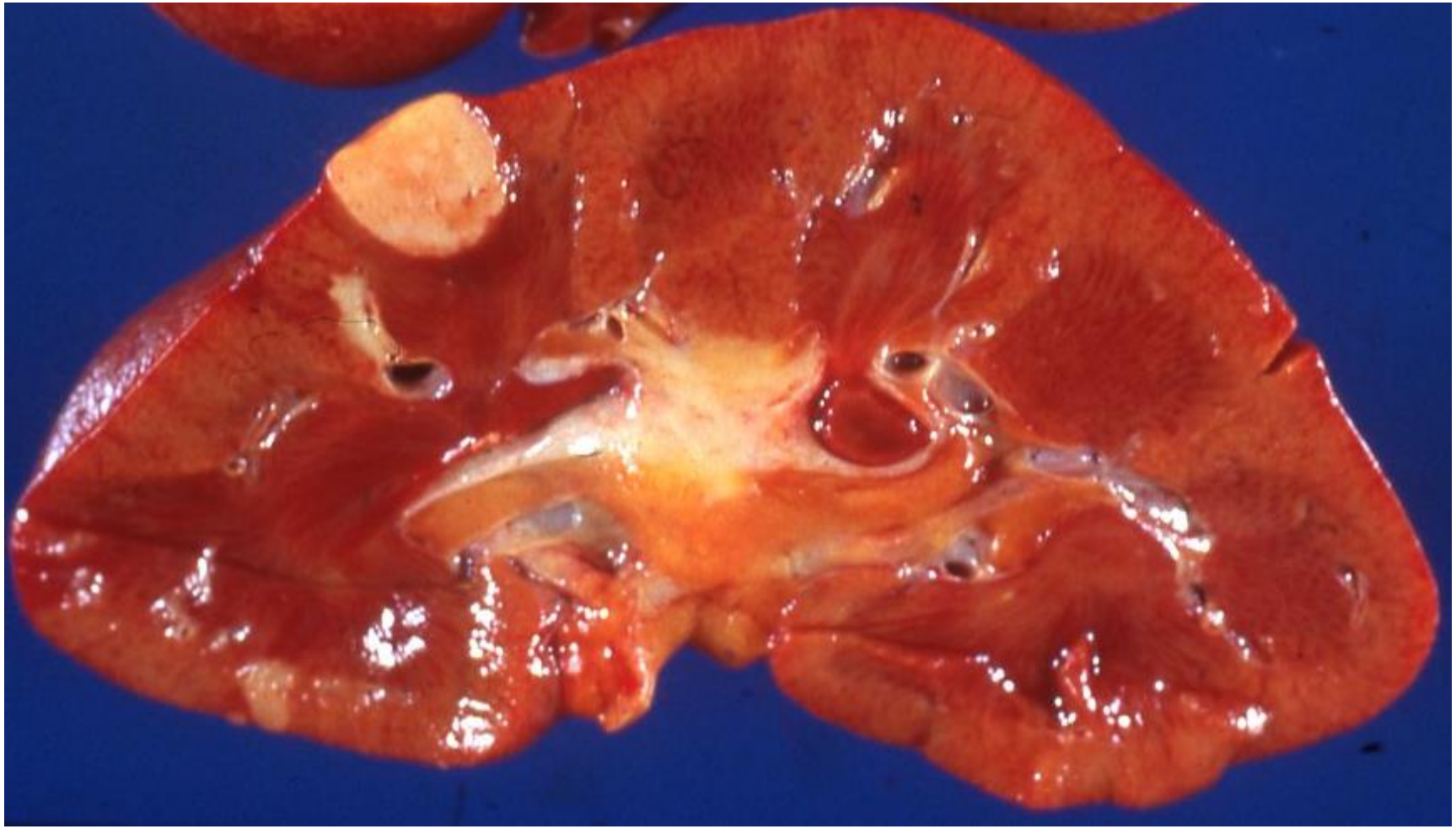
Pharyngeal ulcer in Werner syndrome (adult progeria, a 42-year-old Japanese male patient). The ulcerated pharyngeal mucosa is heavily infected with Gram-positive streptococci. H&E-3



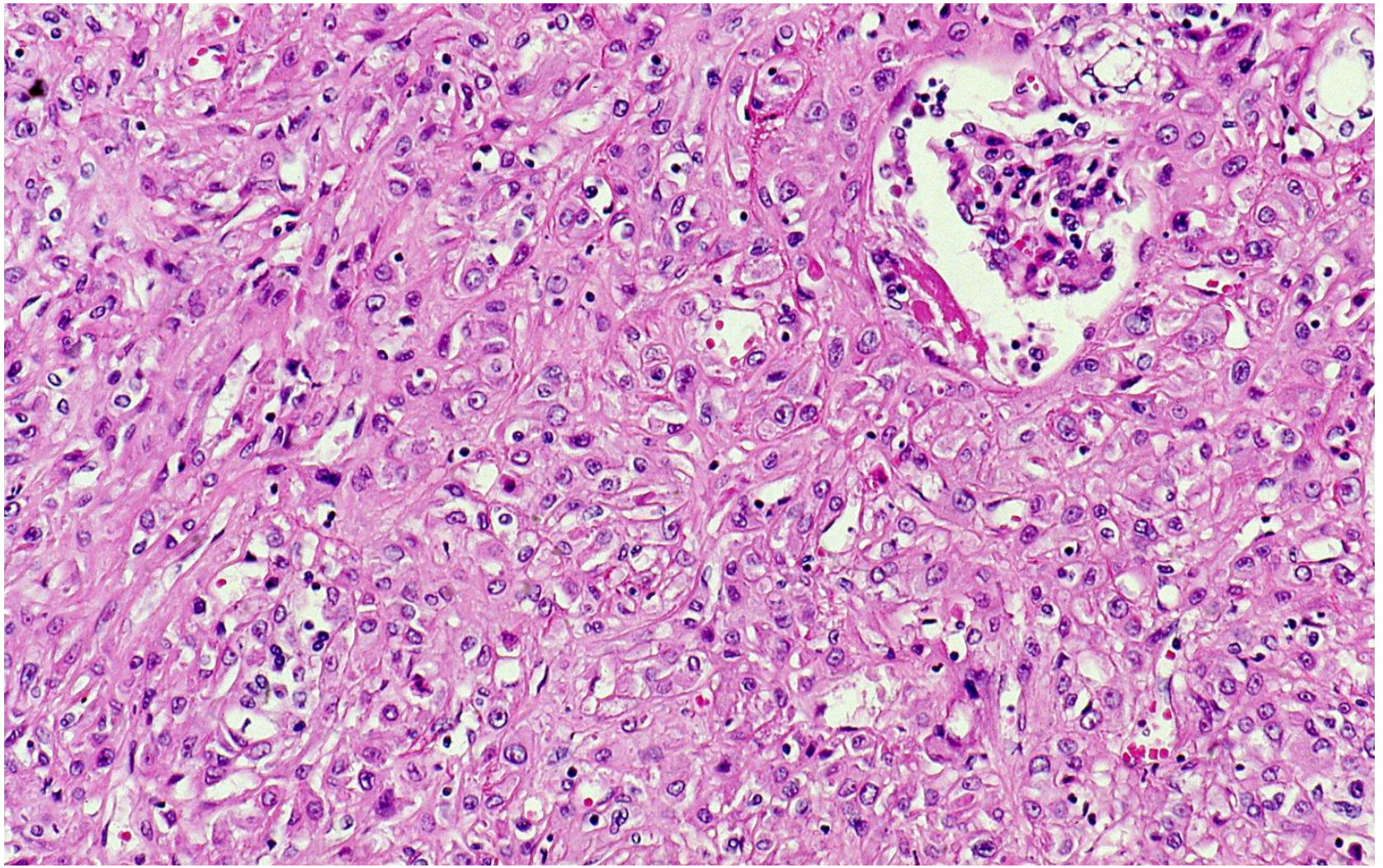
Testis in Werner syndrome (adult progeria, a 42-year-old Japanese male patient). Marked atrophy of the testis is evident. Hyperplastic Leidig's cells are clustered. H&E-4



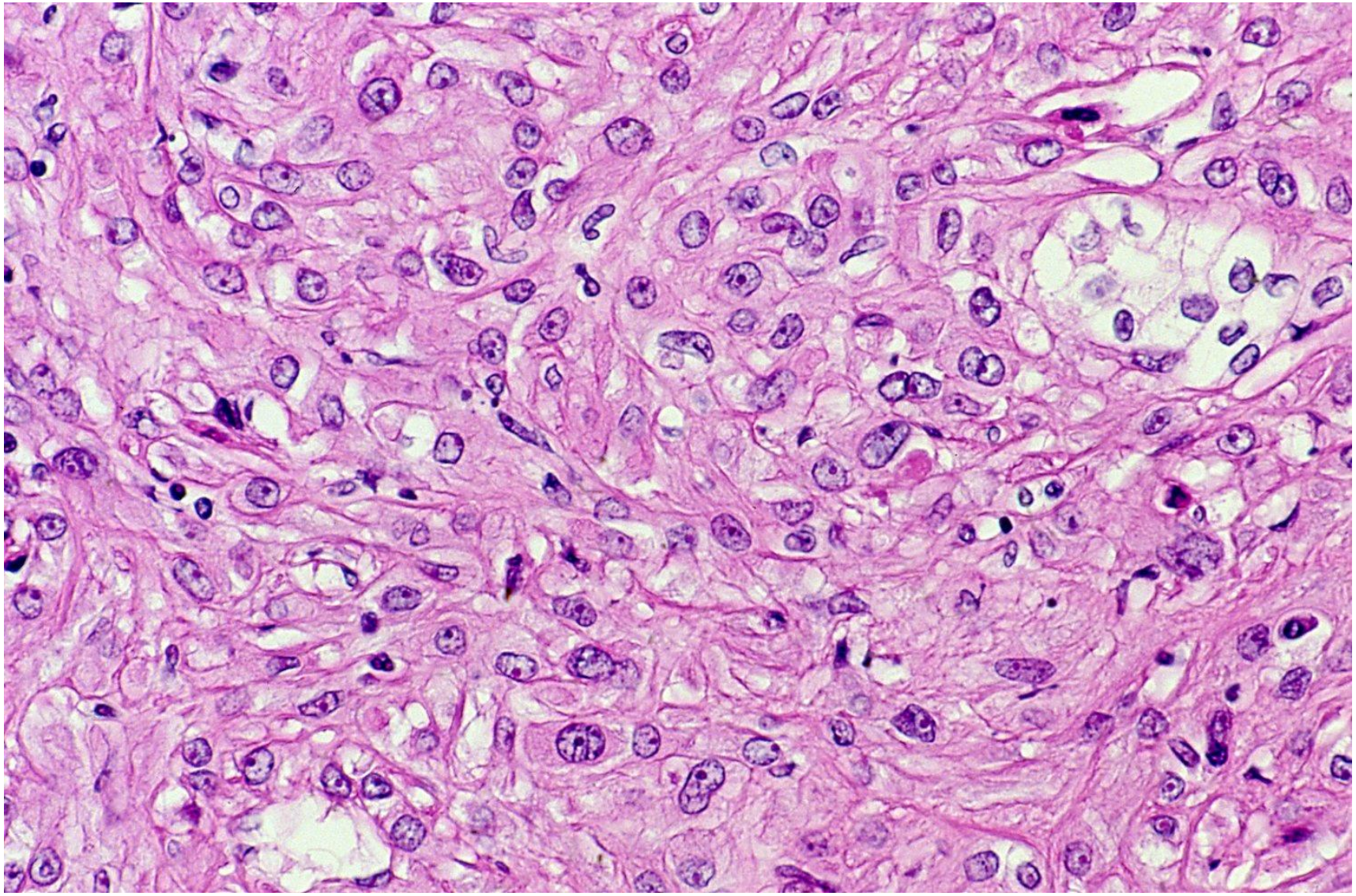
Gross appearance of aorta in Werner syndrome (adult progeria, a 42-year-old Japanese male patient).  Atheromatous plaques are.dispersed.



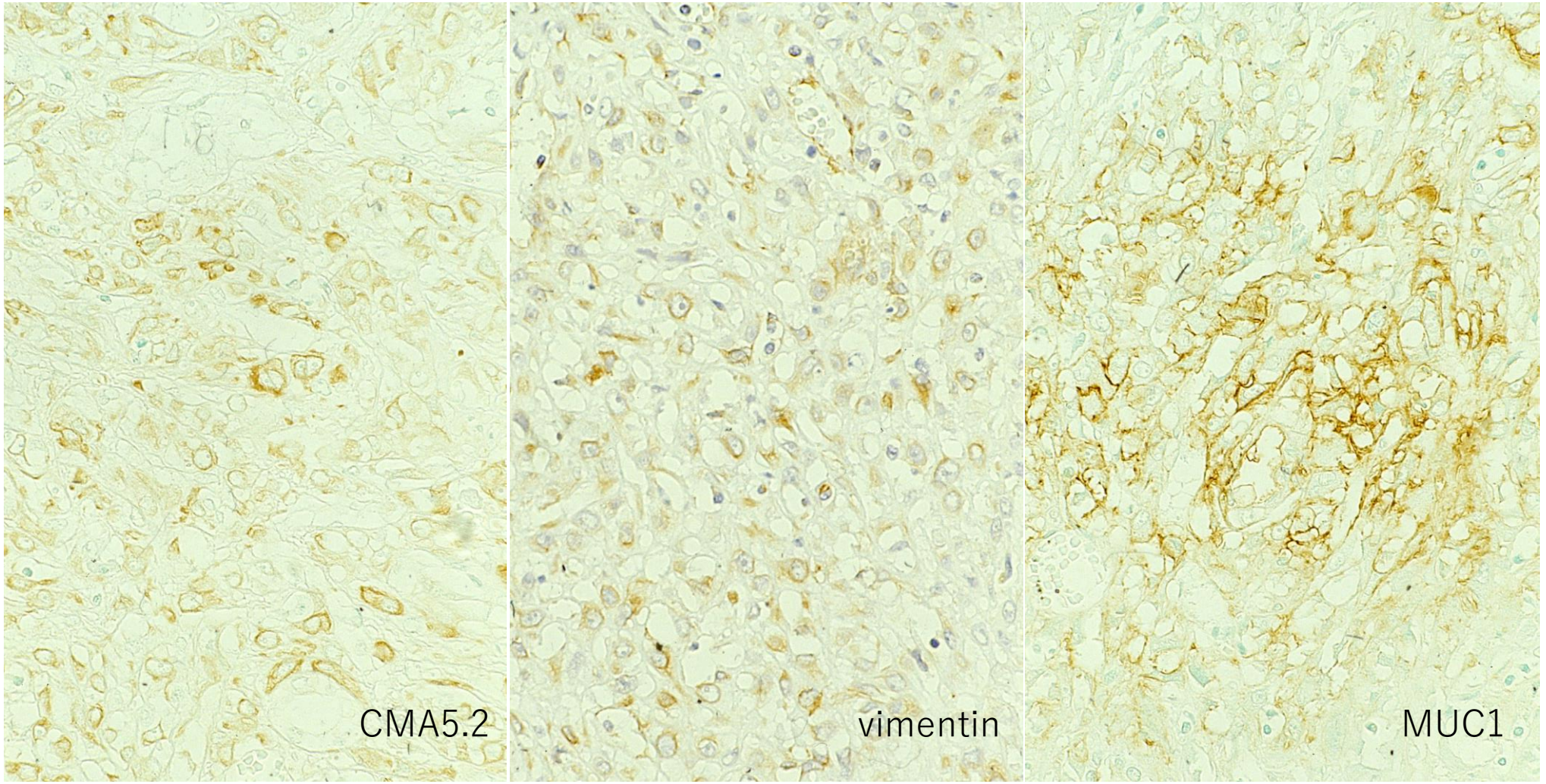
Gross appearance of the left kidney in Werner syndrome (adult progeria, a 42-year-old Japanese male patient). Grayish white tumor is seen in the subcapsular zone of the kidney.



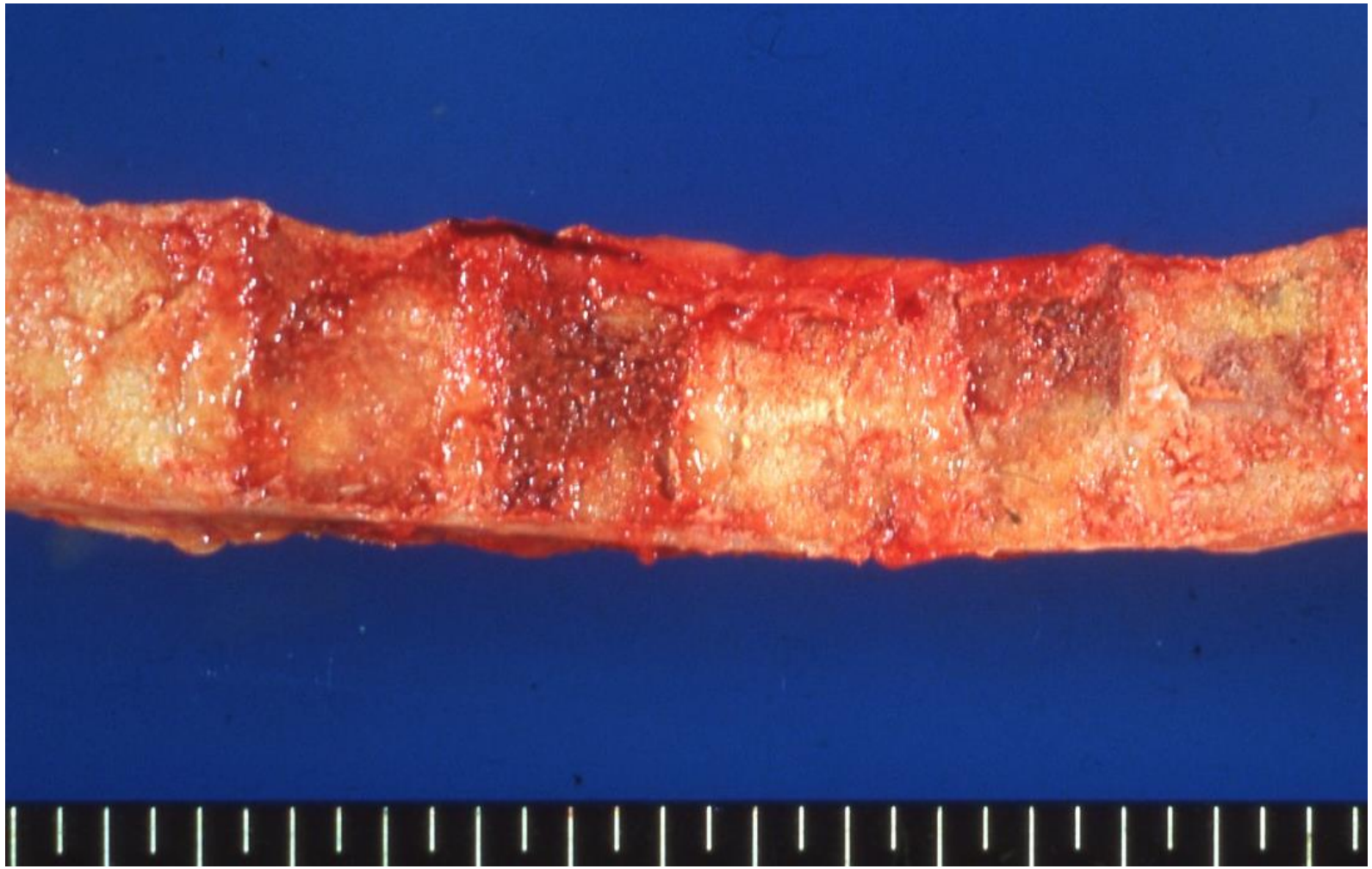
Microscopic appearance of the renal tumor in Werner syndrome (adult progeria, a 42-year-old Japanese male patient). The malignant cells are poorly differentiated, mainly composed of spindled cells. H&E-5



Higher-powered microscopic appearance of the renal tumor in Werner syndrome (adult progeria, a 42-year-old Japanese male patient). The malignant cells are poorly differentiated, mainly composed of spindled cells. H&E-6




Immunohistochemical features of the renal tumor in Werner syndrome (adult progeria, a 42-year-old Japanese male patient). The malignant cells are immunoreactive for low-molecular weight cytokeratin (CAM5.2), vimentin and CA15-3 (MUC1), being compatible with poorly differentiated renal cell carcinoma. Immunostaining for CAM5.2 (left), vimentin(center) and CA15-3 (MUC1, right).



Spine metastasis of the renal tumor in Werner syndrome (adult progeria, a 42-year-old Japanese male patient). The renal tumor was metastatic to systemic organs, including the bone, liver, adrenals and jejunum. Jejunal perforation resulting in purulent peritonitis was the cause of death.