Cutaneous involvement in immunotactoid glomerulopathy (immunotactoid dermatopathy)

Immunotactoid glomerulopathy (Uro-29-2-kidney) seen in adults around 60 years of age presents with nephrotic proteinuria, reduced glomerular filtration rate and hypocomplementemia in half of affected patients. Cryoglobulins are negative, while a circulating monoclonal paraprotein and/or lymphoplasmacytic malignancy is present in about two-thirds. The glomeruli commonly reveals a membranoproliferative pattern. Immunofluorescence demonstrates chunky deposition of IgG (often monoclonal) and C3 in the mesangium and capillary wall. Electron microscopy identified the deposition of microtubular structures, often in parallel arrays, with the diameter of 20-90 nm. The affected patients achieve remission with therapy directed against malignancy. However, the disease may result in end-stage kidney disease. Extrarenal presentations have been reported in the cornea, liver and skin. Henoch-Schönlein-like syndrome with leukocytoclastic vasculitis may be associated with immunotactoid glomerulopathy.

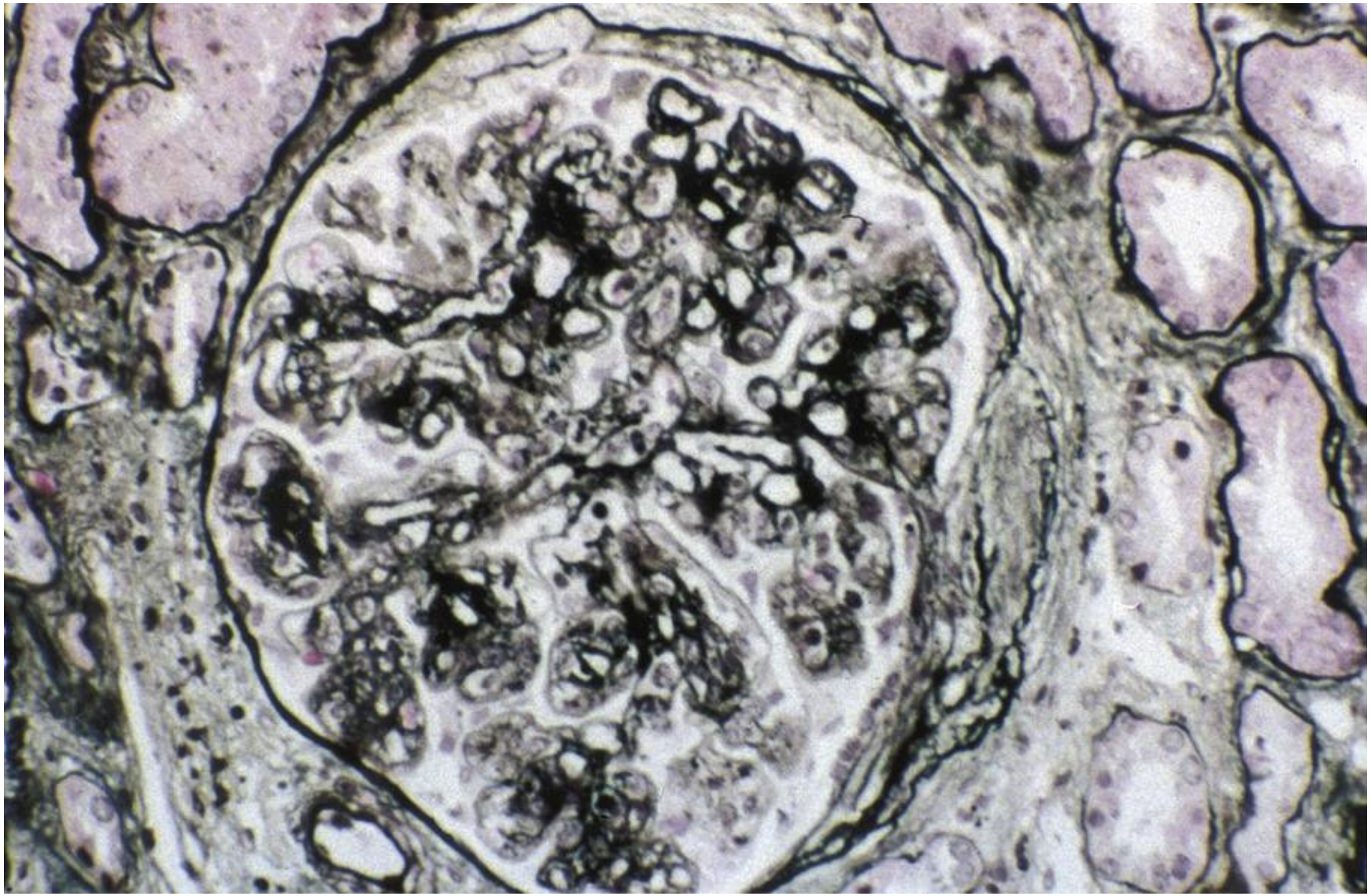
Ref.-1: Fogo AB, et al. AJKD atlas of renal pathology: immunotactoid glomerulopathy. Am J Kid Dis 2015; 66(4): e29-e30. doi 10.1053/j.ajkd.2015.08.003

Ref.-2: Orfila C, et al. Immunotactoid glomerulopathy and cutaneous vasculitis. Am J Nephrol 1991; 11(1): 67-72. doi: 10.1159/000168276

Case presentation

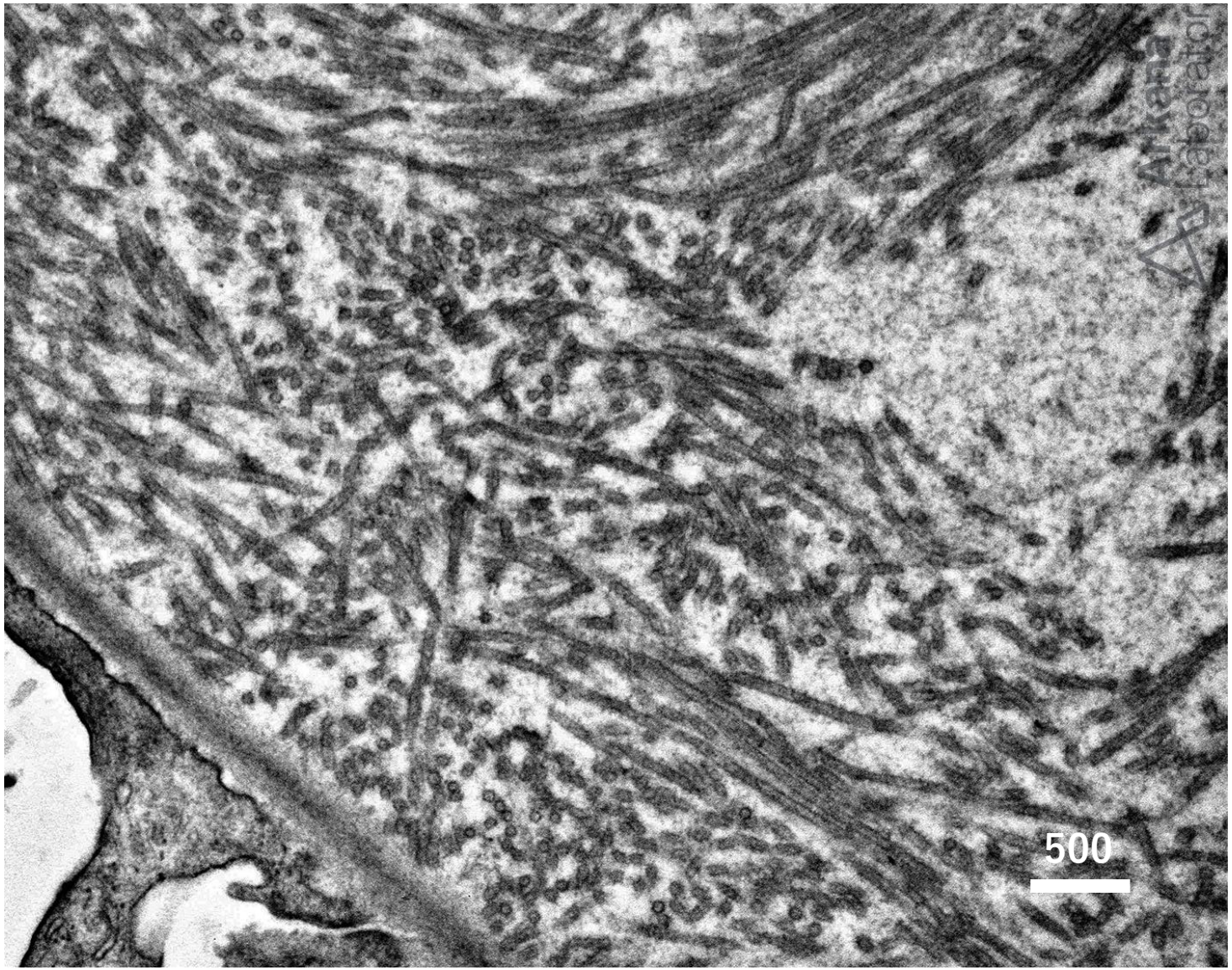
A 61 y-o male patient was diagnosed as immunotactoid glomerulopathy 7 years ago. He had manifested nephrotic syndrome and renal biopsy had revealed deposition of IgA-positive microtubule-like structures ultrastructurally. Because of rapidly progressive renal dysfunction, the patient eventually received hemodialysis. Three years ago, hemorrhagic necrosis of the jejunum abruptly happened, and intravascular deposition of IgA-positive material was microscopically proven in the surgical specimen. This time, he presented purpuric skin ulcers on bilateral lower legs. Intravascular thrombotic deposition of fibrillar material was proven. The jejunal and cutaneous lesions should be closely related to intravascular deposition of the IgA-related immunotactoid material.

Kidney Bx



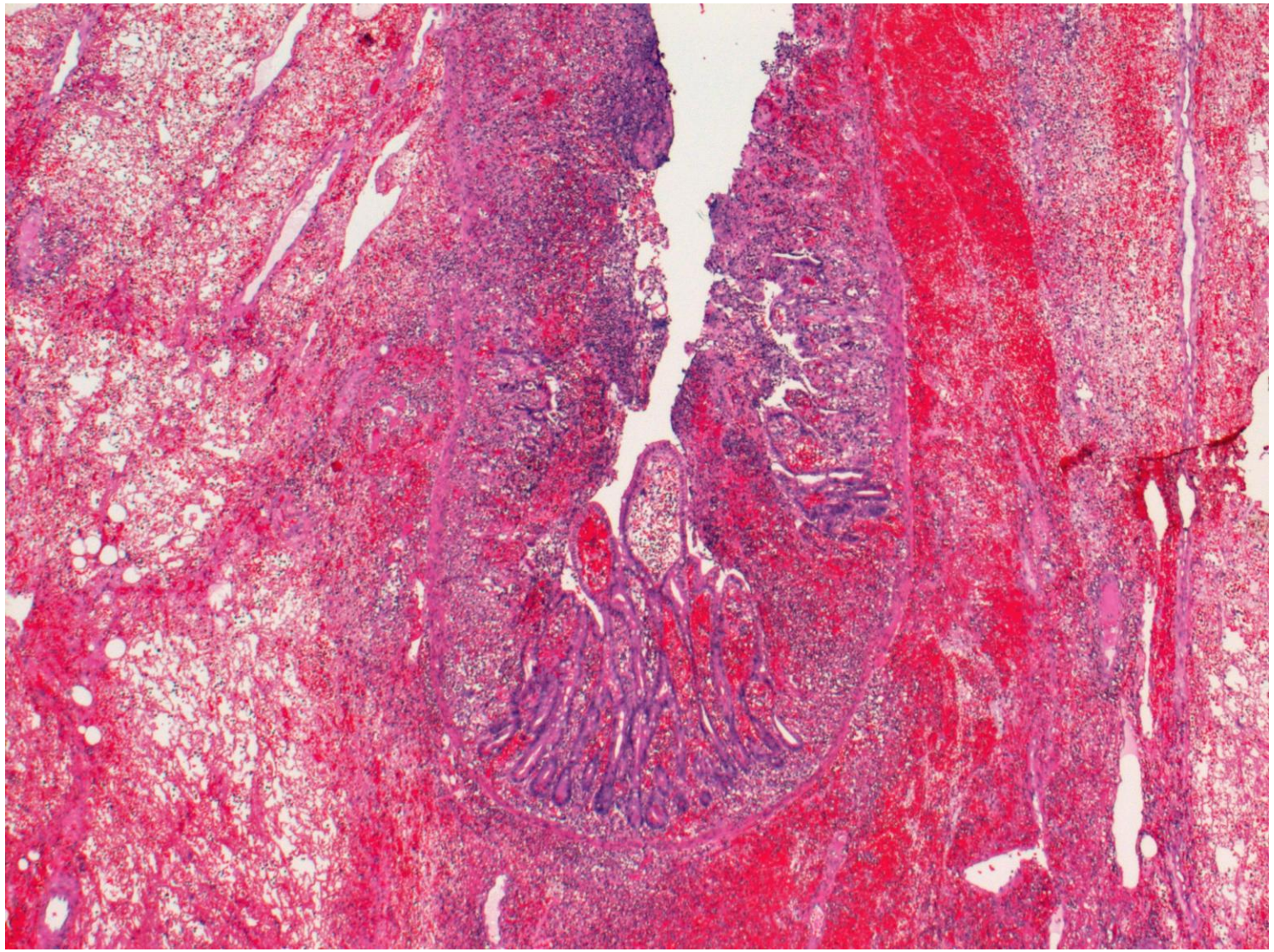
Immunotactoid glomerulopathy with a membranoproliferative pattern and crescent formation (renal biopsy 7 years earlier, PAM)

Kidney Bx



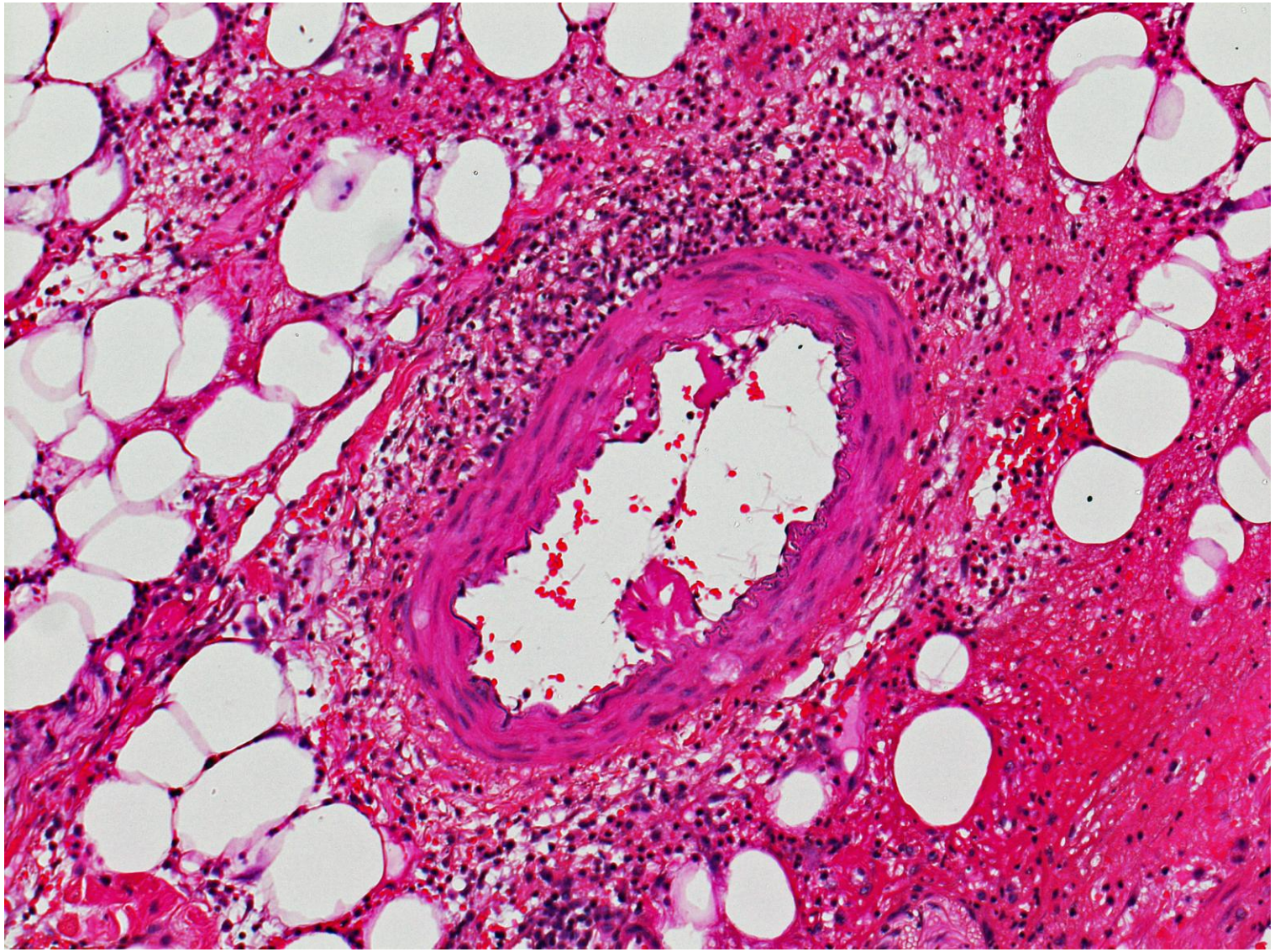
Ultrastructure of immunotactoid glomerulopathy with deposition of microtubular material in the glomerulus (renal biopsy 7 years earlier, TEM)

Jejunum Rx



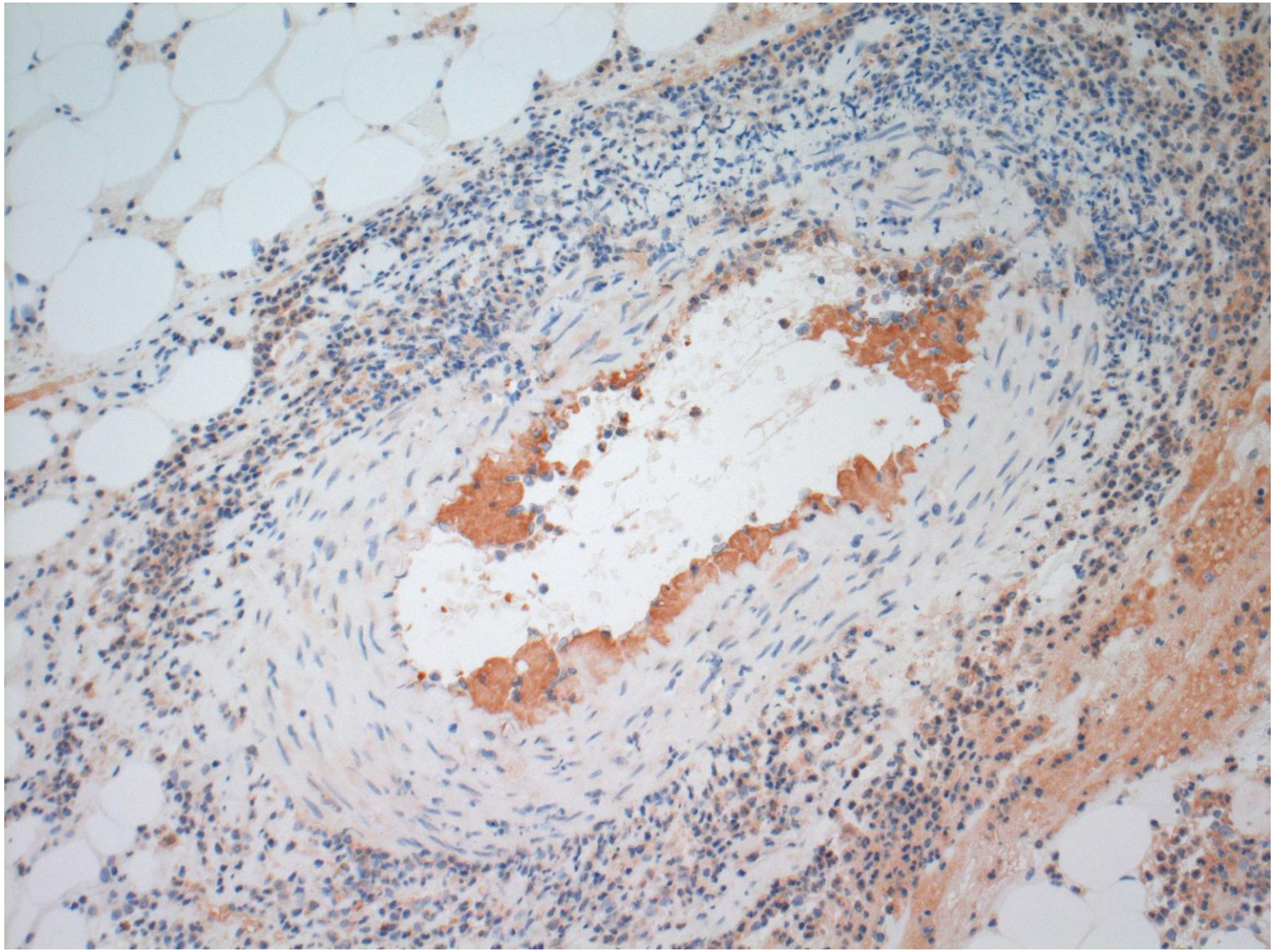
Hemorrhagic necrosis of the jejunum, surgical specimen 3 years earlier. Transmural hemorrhagic necrosis is observed (immunotactoid enteropathy, H&E-a).

Jejunum Rx



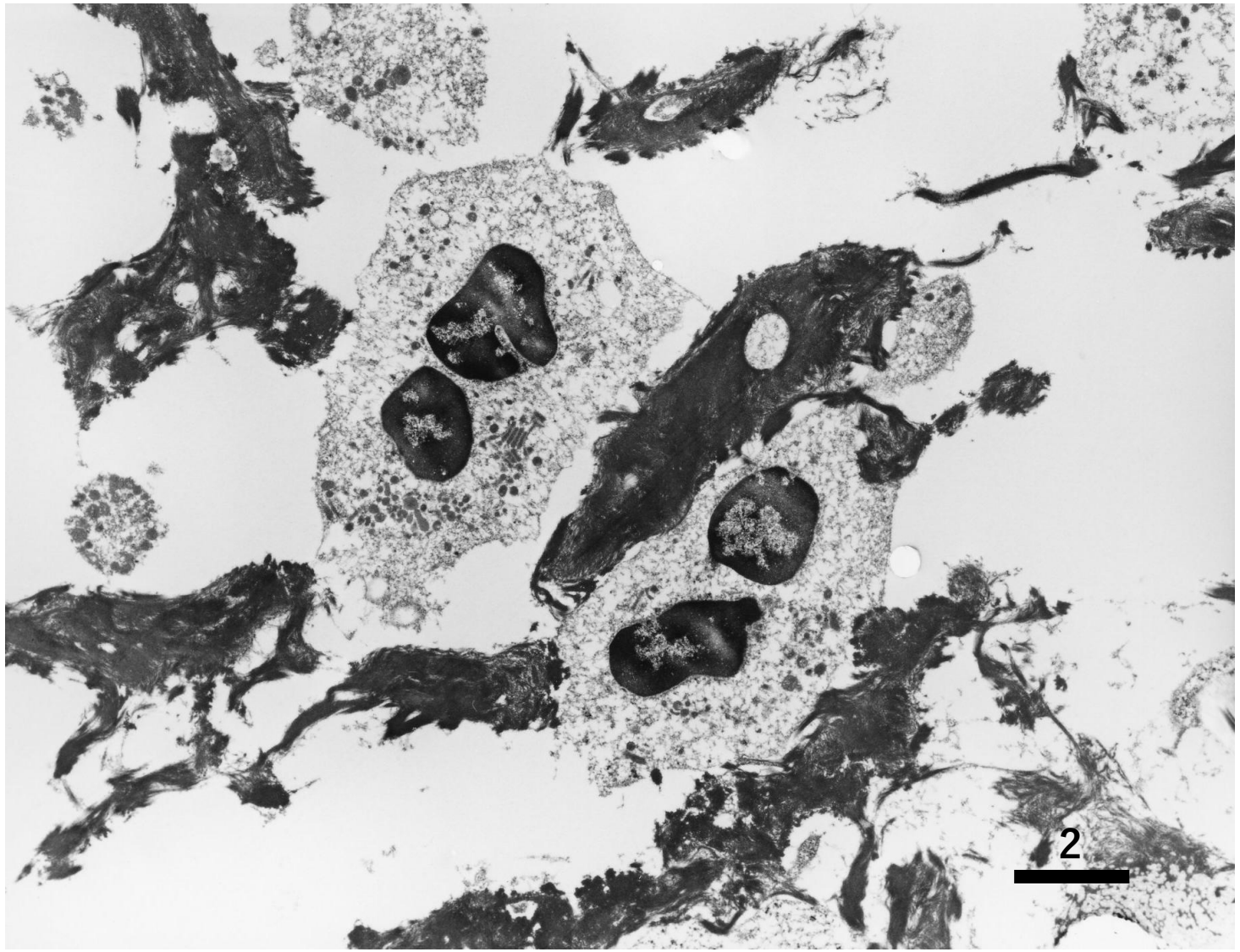
Hemorrhagic necrosis of the jejunum, surgical specimen 3 years earlier. A submucosal artery contains an eosinophilic thrombotic material in the hemorrhagic background (immunotactoid enteropathy, H&E-b).

Jejunum Rx



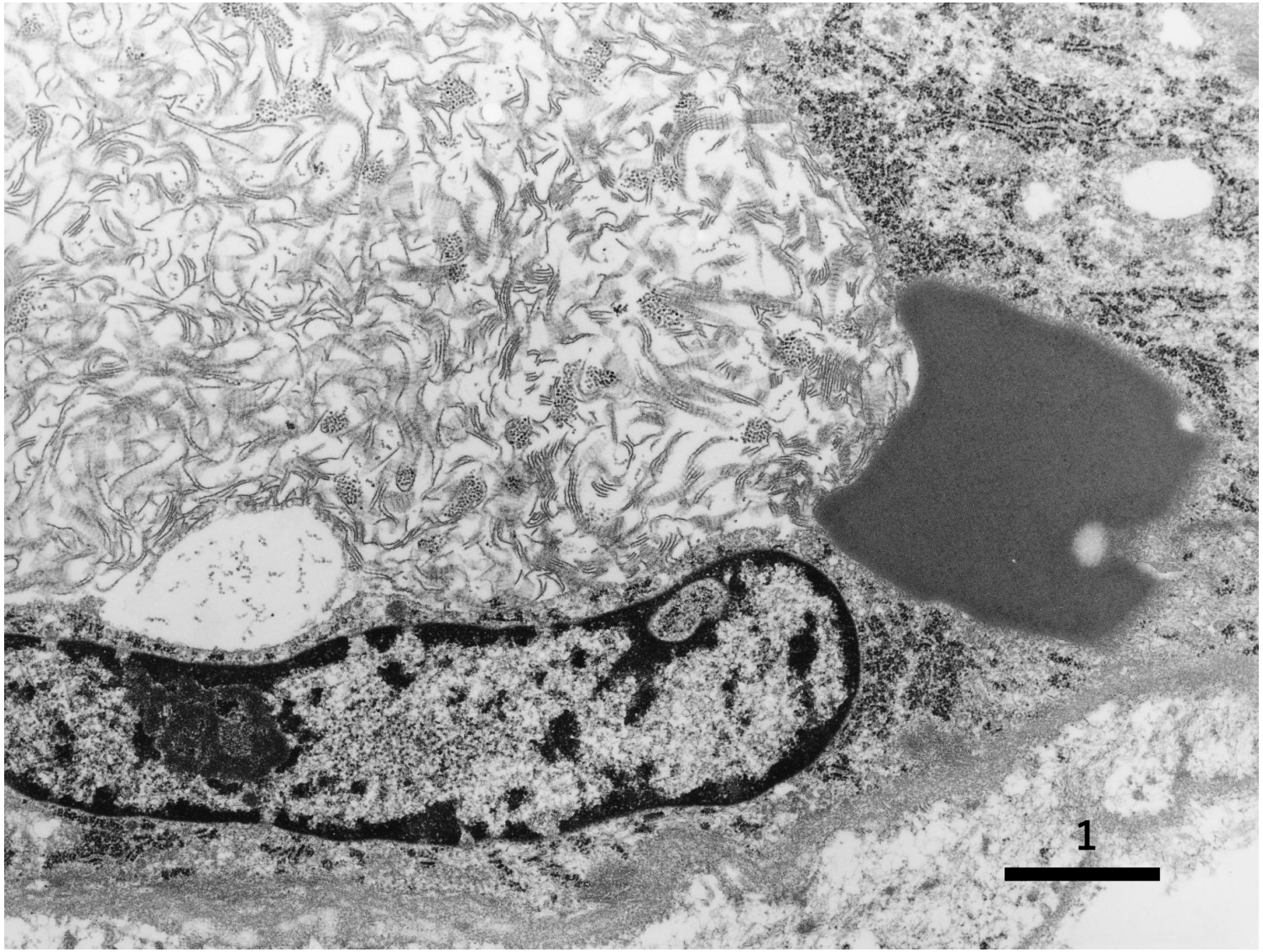
Hemorrhagic necrosis of the jejunum, surgical specimen 3 years earlier. The intraluminal thrombotic material on the endothelial cells is immunoreactive for IgA (immunotactoid enteropathy, immunostaining for IgA).

Jejunum Rx



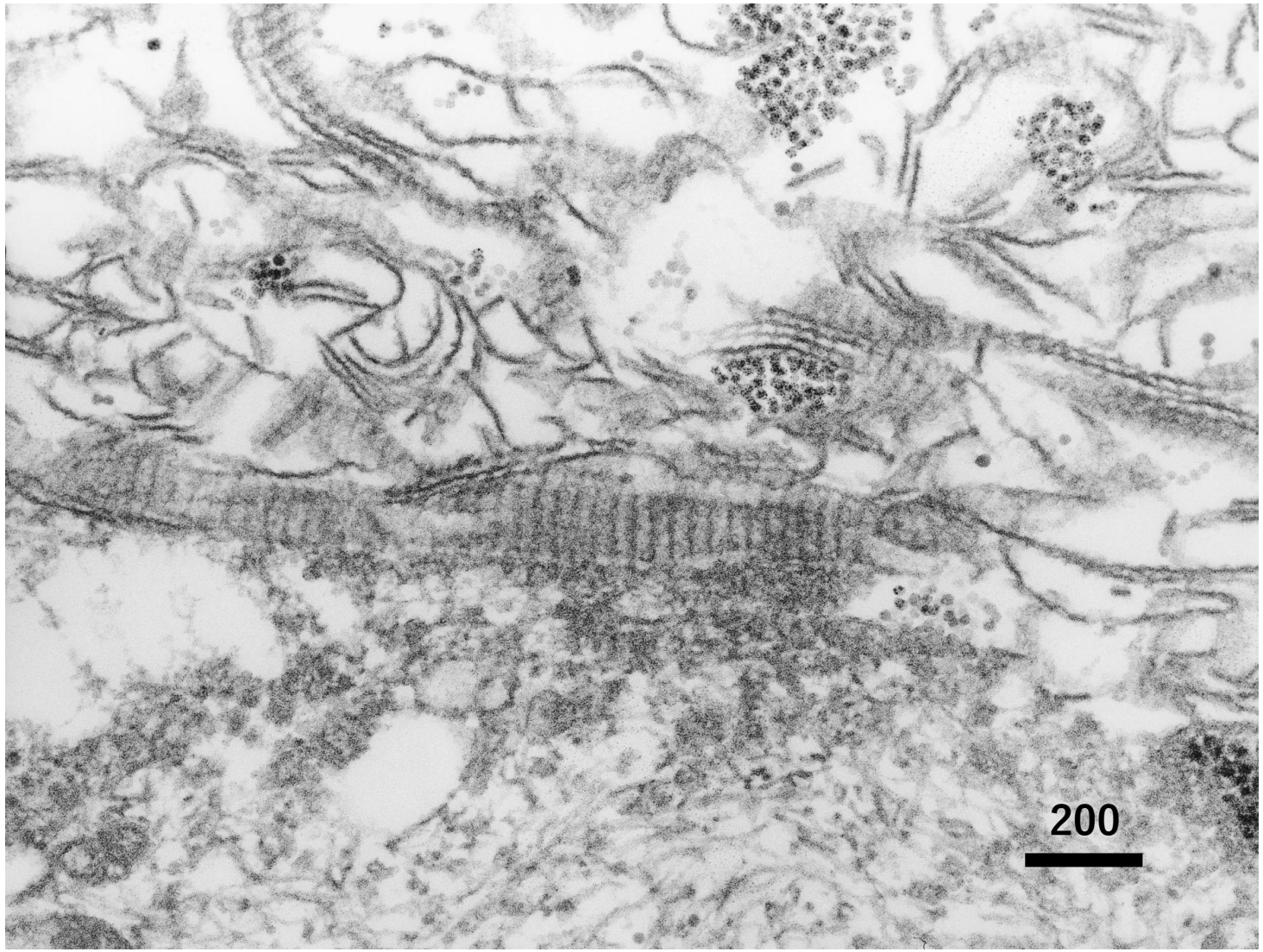
Ultrastructure of immunotactoid enteropathy. The jejunum with hemorrhagic necrosis shows exudation of electron-dense fibrin fibrils and neutrophils (TEM-1, using a block dug from formalin-fixed, paraffin-embedded surgical specimen).

Jejunum Rx



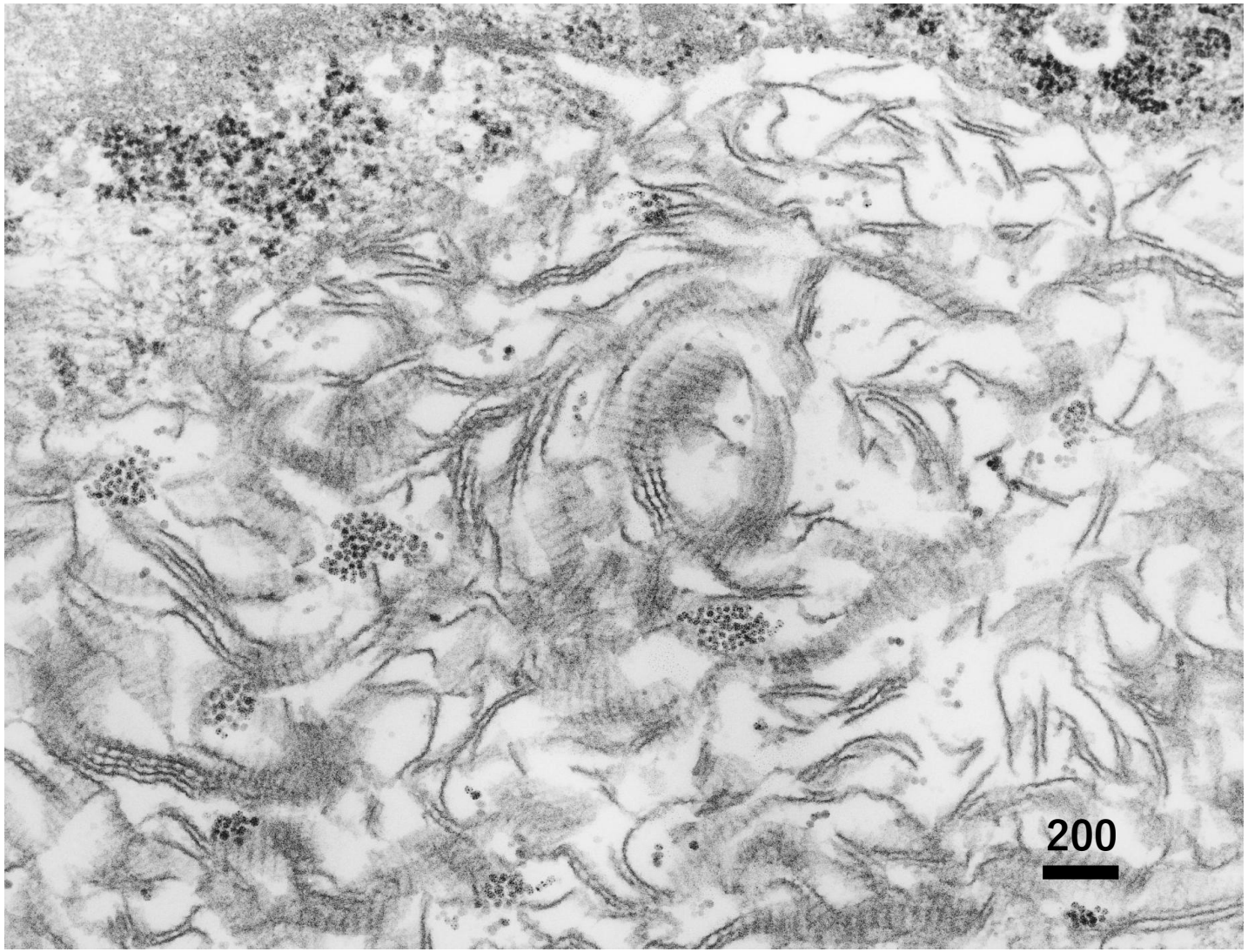
Ultrastructure of immunotactoid enteropathy. The vascular lumen contains a cluster of fibrillar material (TEM-2, using a block dug from formalin-fixed, paraffin-embedded surgical specimen).

Jejunum Rx



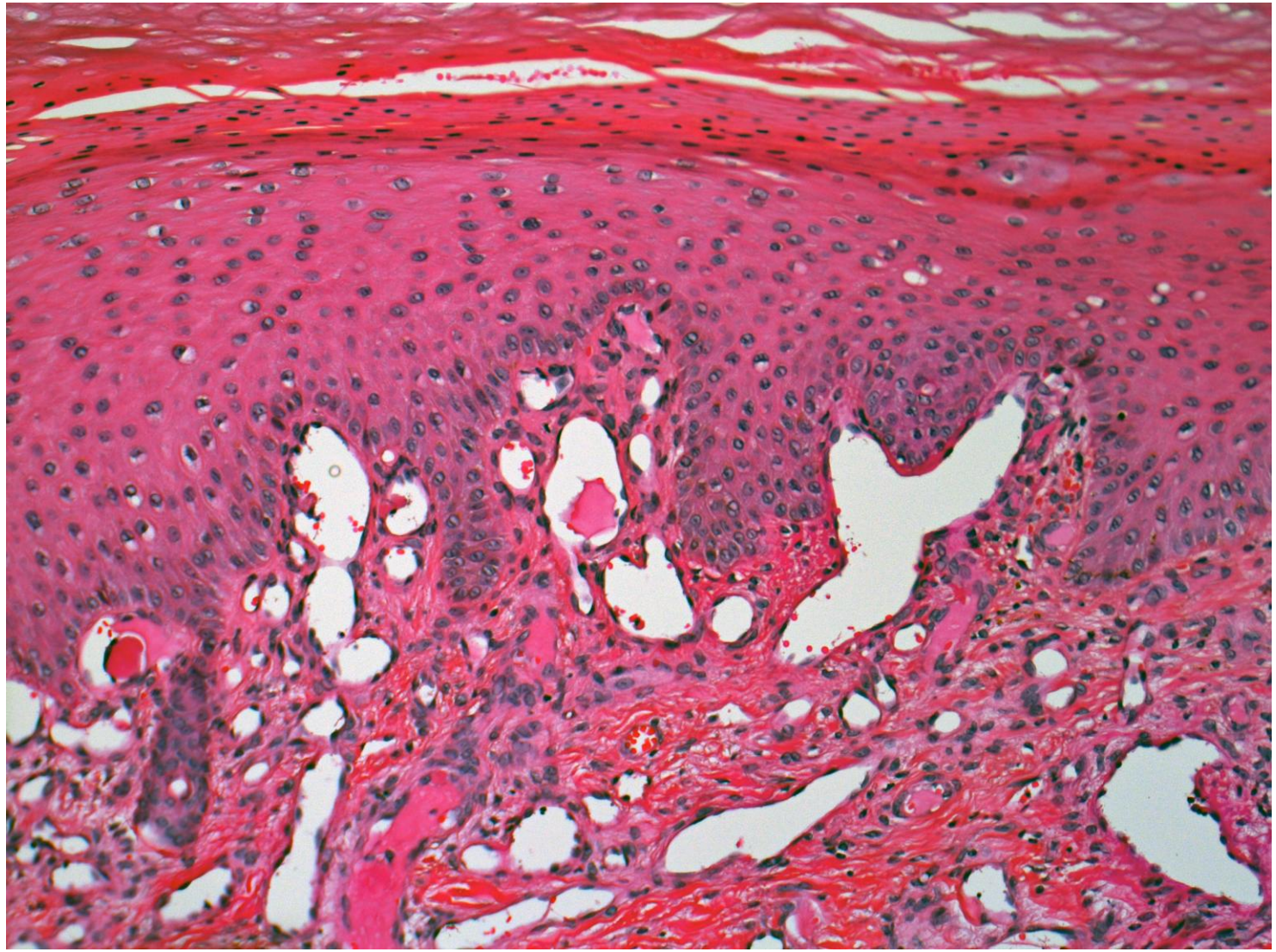
Ultrastructure of immunotactoid enteropathy. The vascular lumen contains a cluster of fibrillar material with fine cross striations (TEM-3, using a block dug from formalin-fixed, paraffin-embedded surgical specimen).

Jejunum Rx



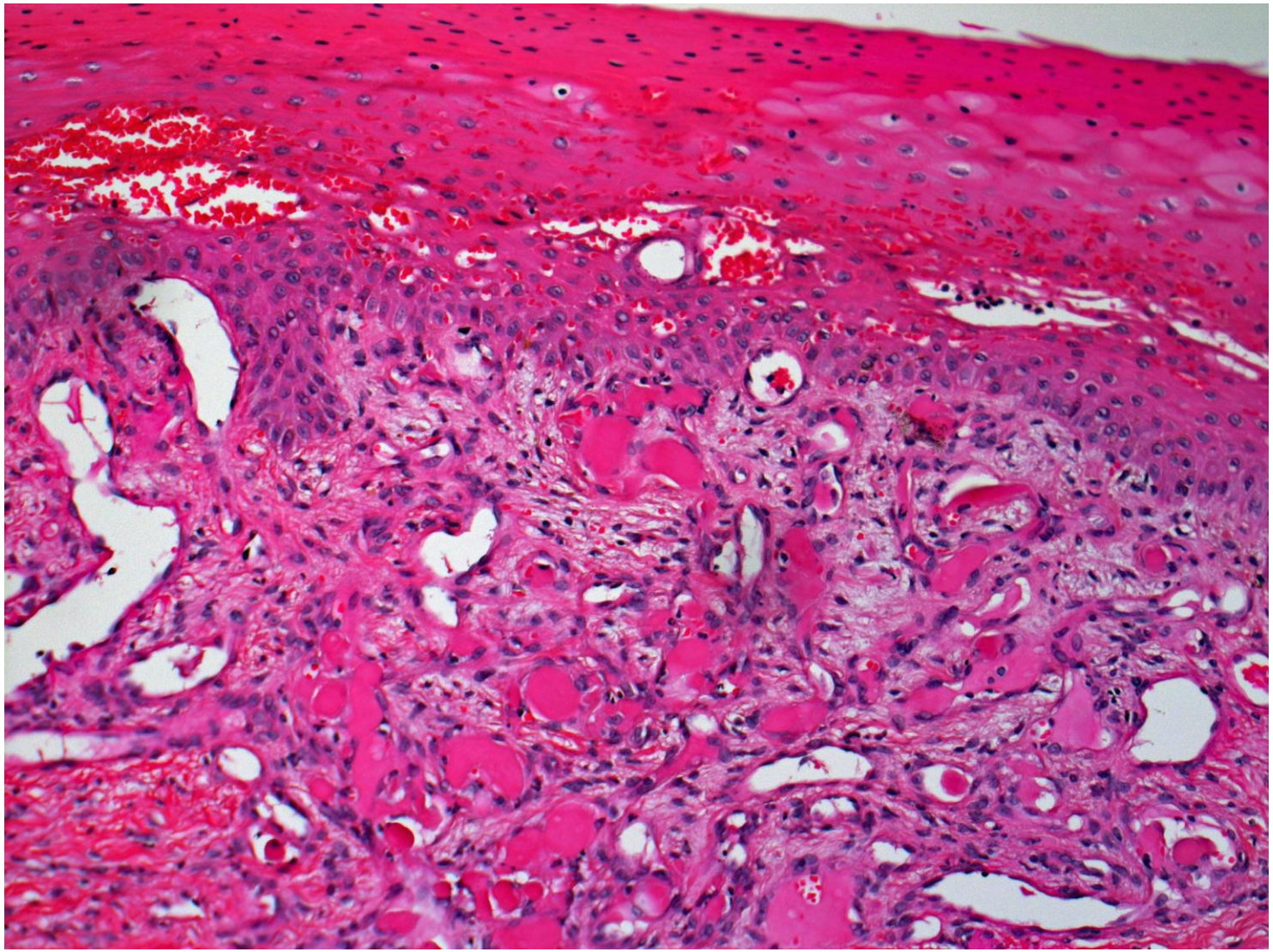
Ultrastructure of immunotactoid enteropathy. The vascular lumen contains a cluster of fibrillar material with fine cross striations. Elongated or whorled arrangements are discerned (TEM-4, using a block dug from formalin-fixed, paraffin-embedded surgical specimen).

Skin Bx



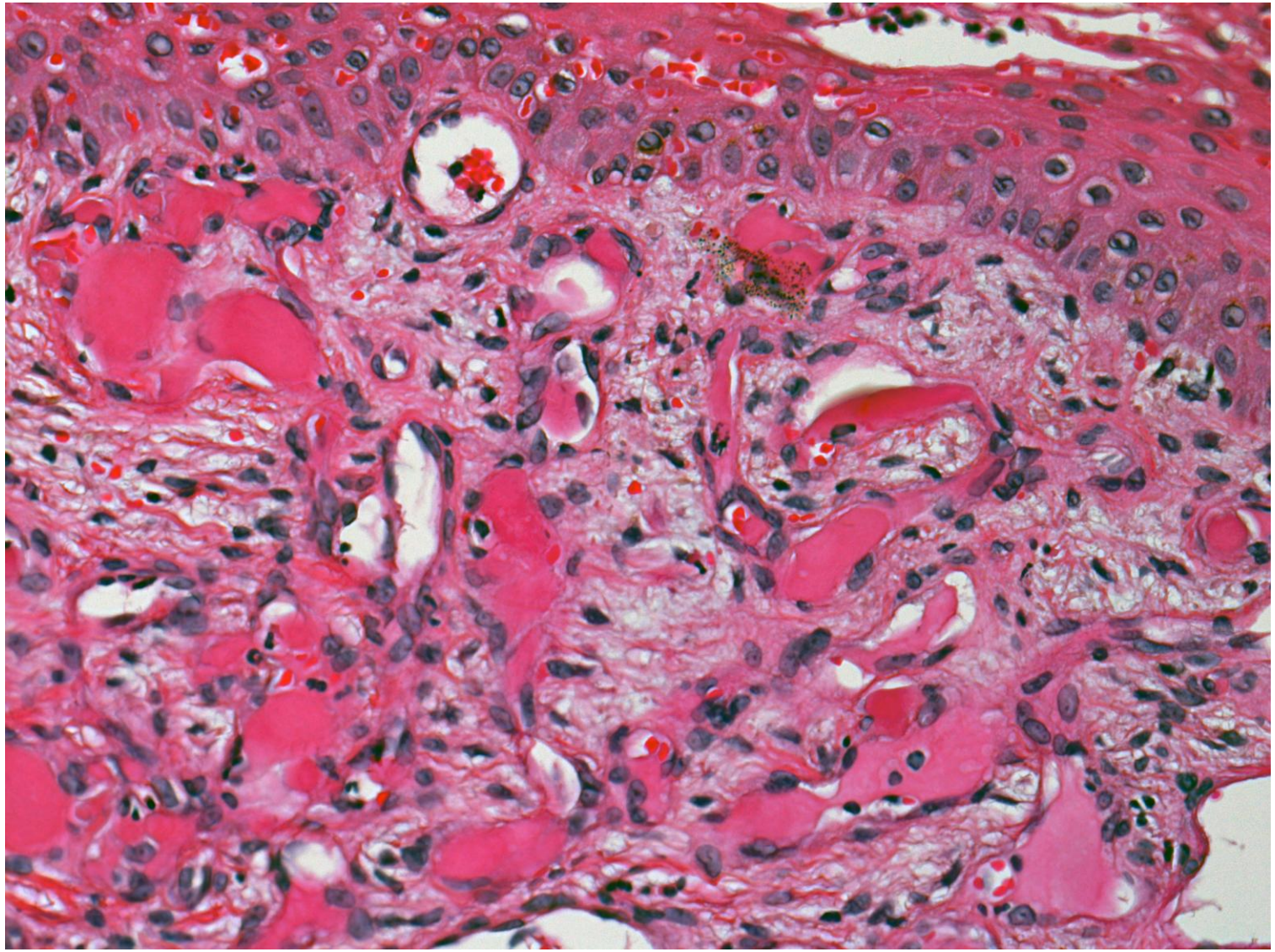
Immunotactoid dermatopathy seen on the lower leg of a 61 y-o male patient. The purpuric skin lesion reveals stasis dermatitis-like congestion with angiectasia in the upper dermis. Eosinophilic thrombotic material is focally associated (H&E-1).

Skin Bx



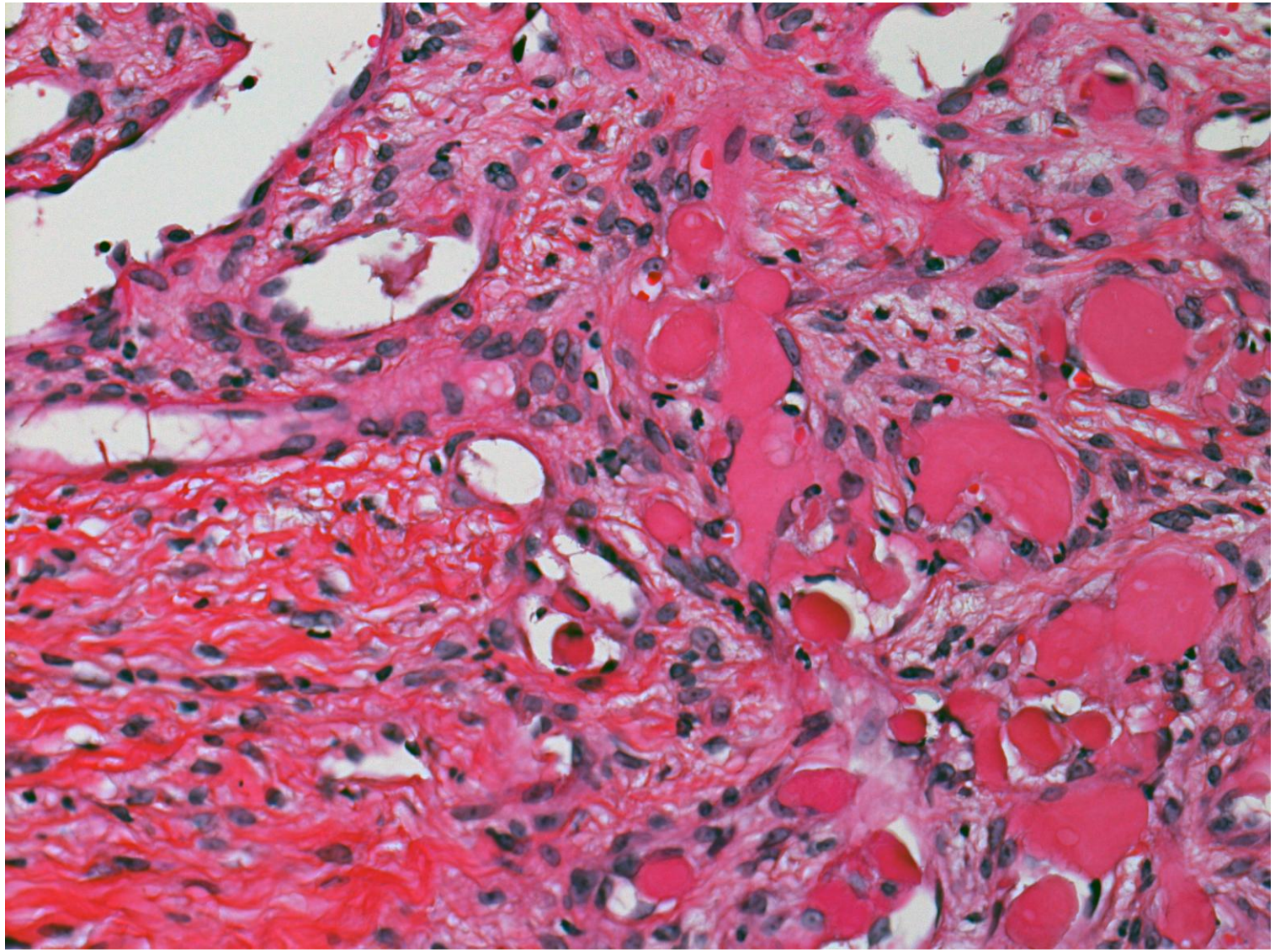
Immunotactoid dermatopathy seen on the lower leg of a 61 y-o male patient. The purpuric skin lesion reveals stasis dermatitis-like congestion with angiectasia in the upper dermis. Eosinophilic thrombotic material is frequently associated. No vasculitis is noted (H&E-2).

Skin Bx



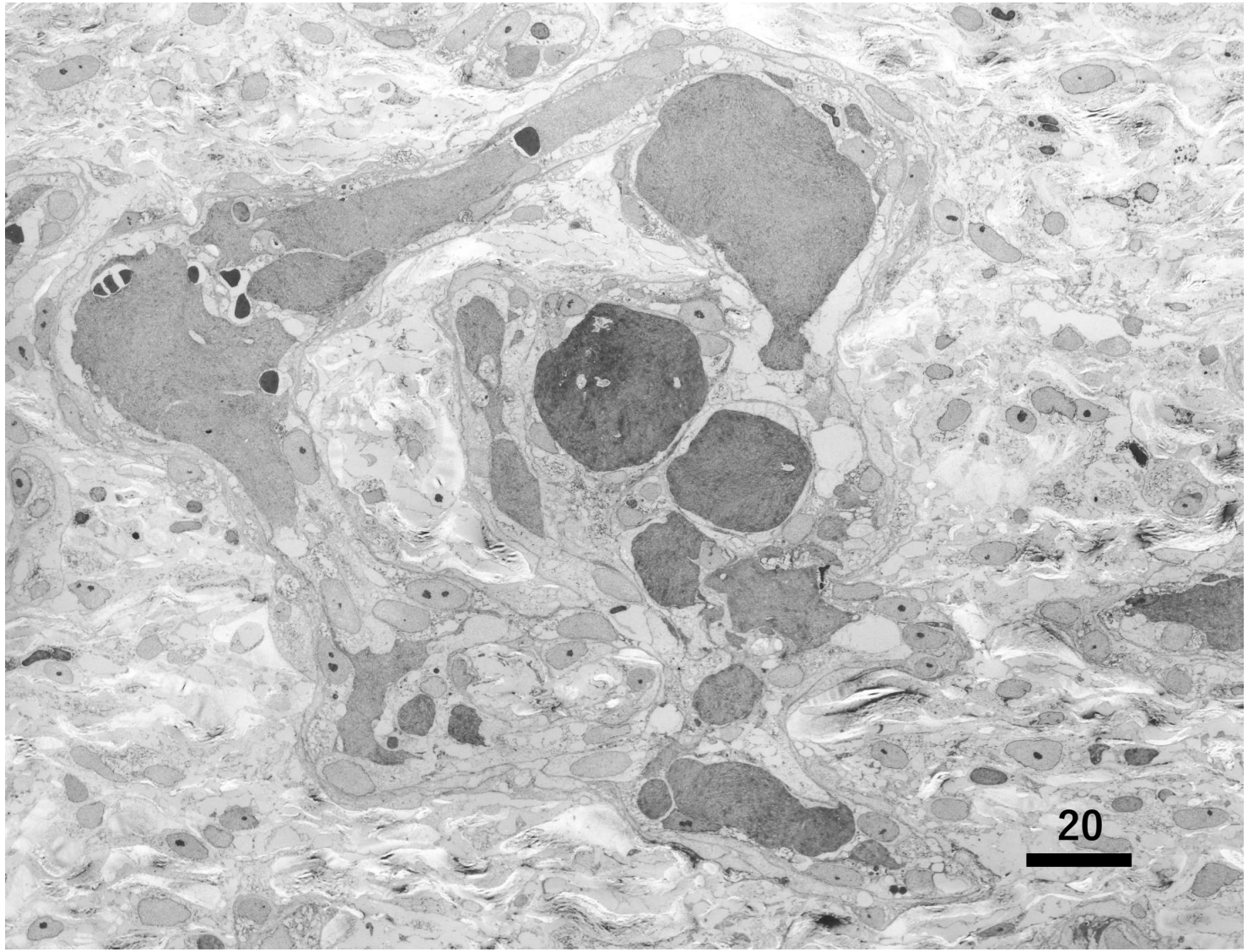
Immunotactoid dermatopathy seen on the lower leg of a 61 y-o male patient. The purpuric skin lesion reveals stasis dermatitis-like congestion with angiectasia in the upper dermis. Eosinophilic thrombotic material is frequently associated. No vasculitis is noted (H&E-3).

Skin Bx



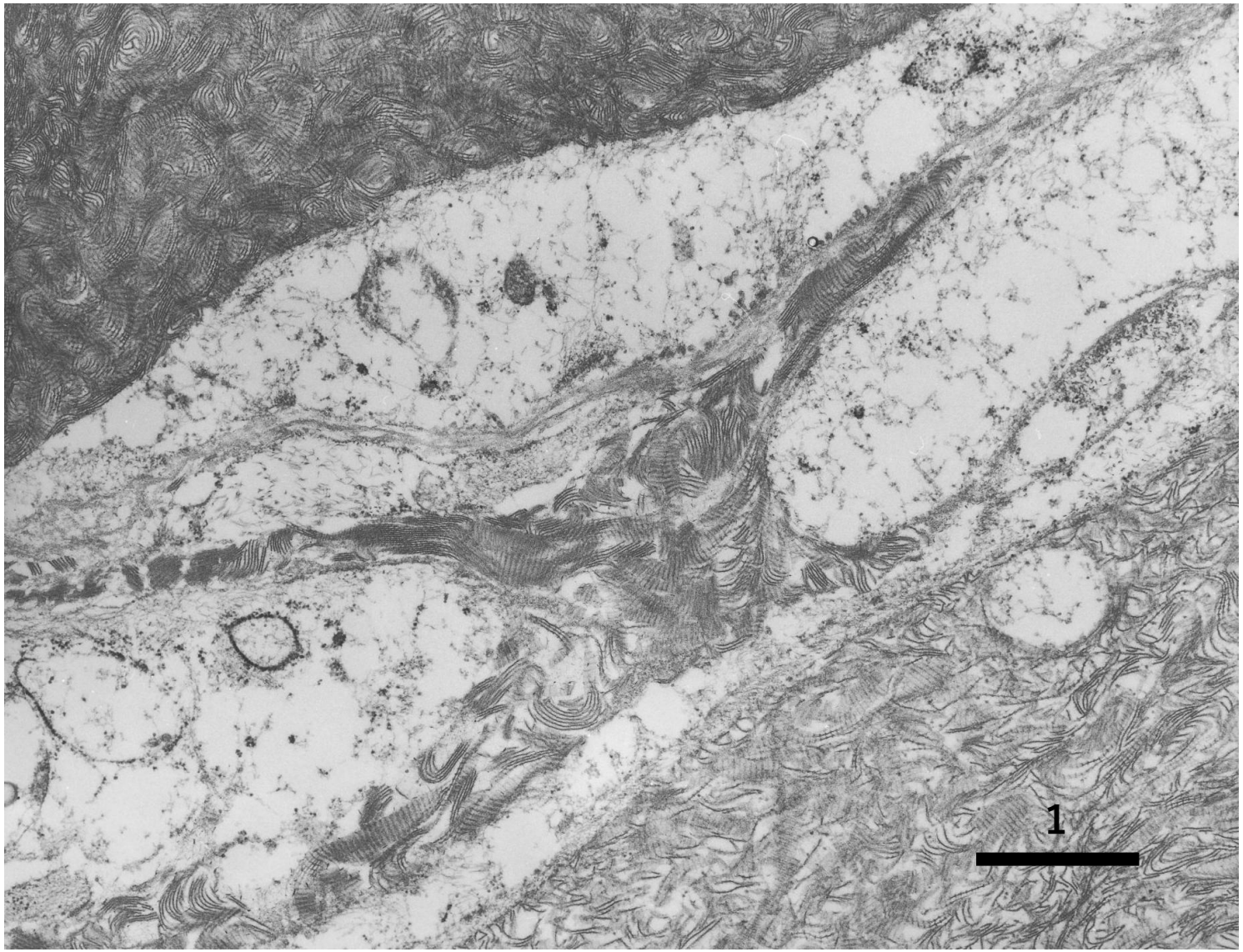
Immunotactoid dermatopathy seen on the lower leg of a 61 y-o male patient. The purpuric skin lesion reveals stasis dermatitis-like congestion with angiectasia in the upper dermis. Eosinophilic thrombotic material is frequently associated. No vasculitis is noted (H&E-4).

Skin Bx



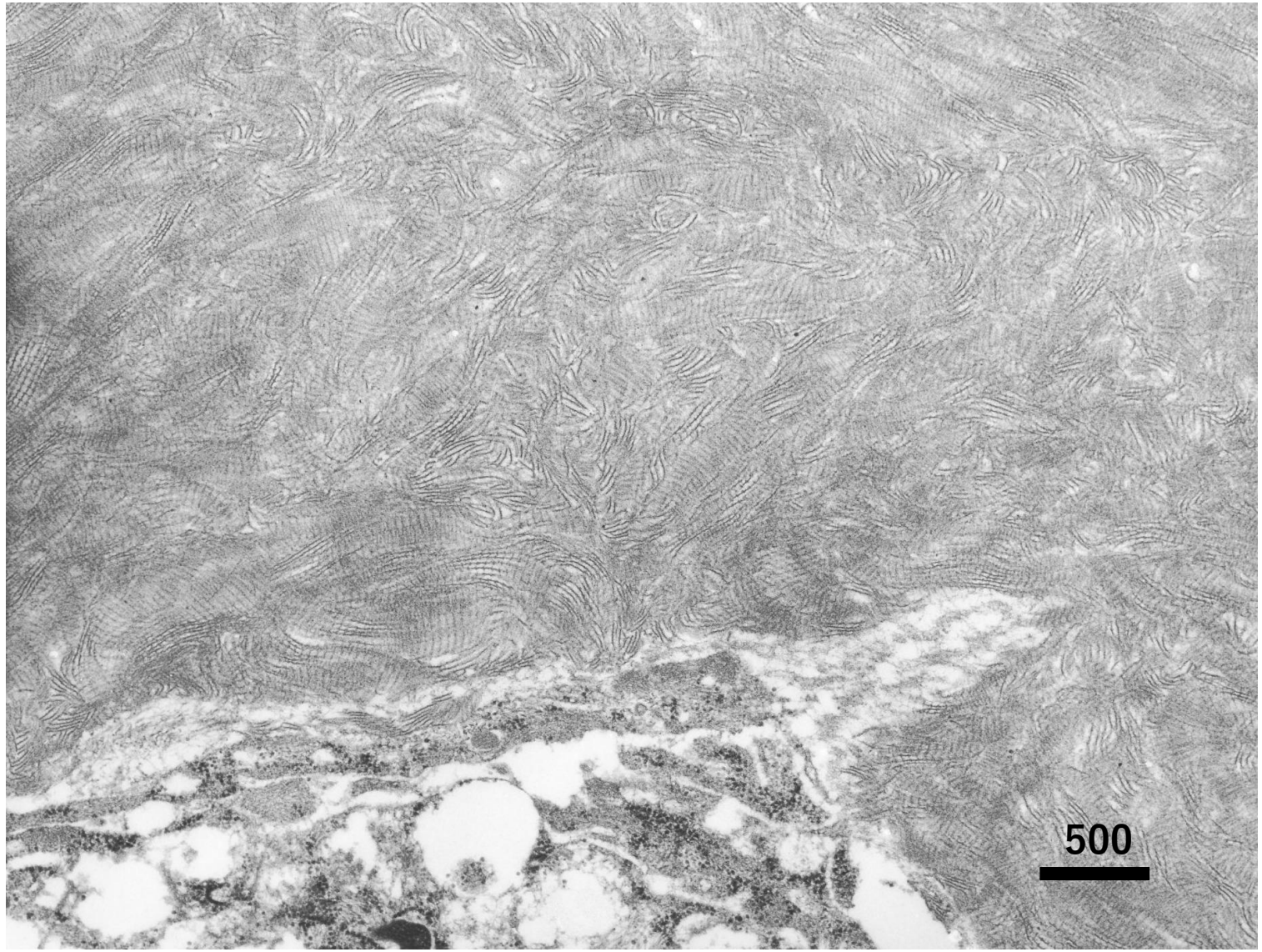
Ultrastructure of immunotactoid dermatopathy seen on the lower leg of a 61 y-o male patient. The purpuric skin lesion reveals deposition of thrombotic material in capillary vessels in the upper dermis. Fibrillar structures are observed in the deposit (TEM-1)

Skin Bx



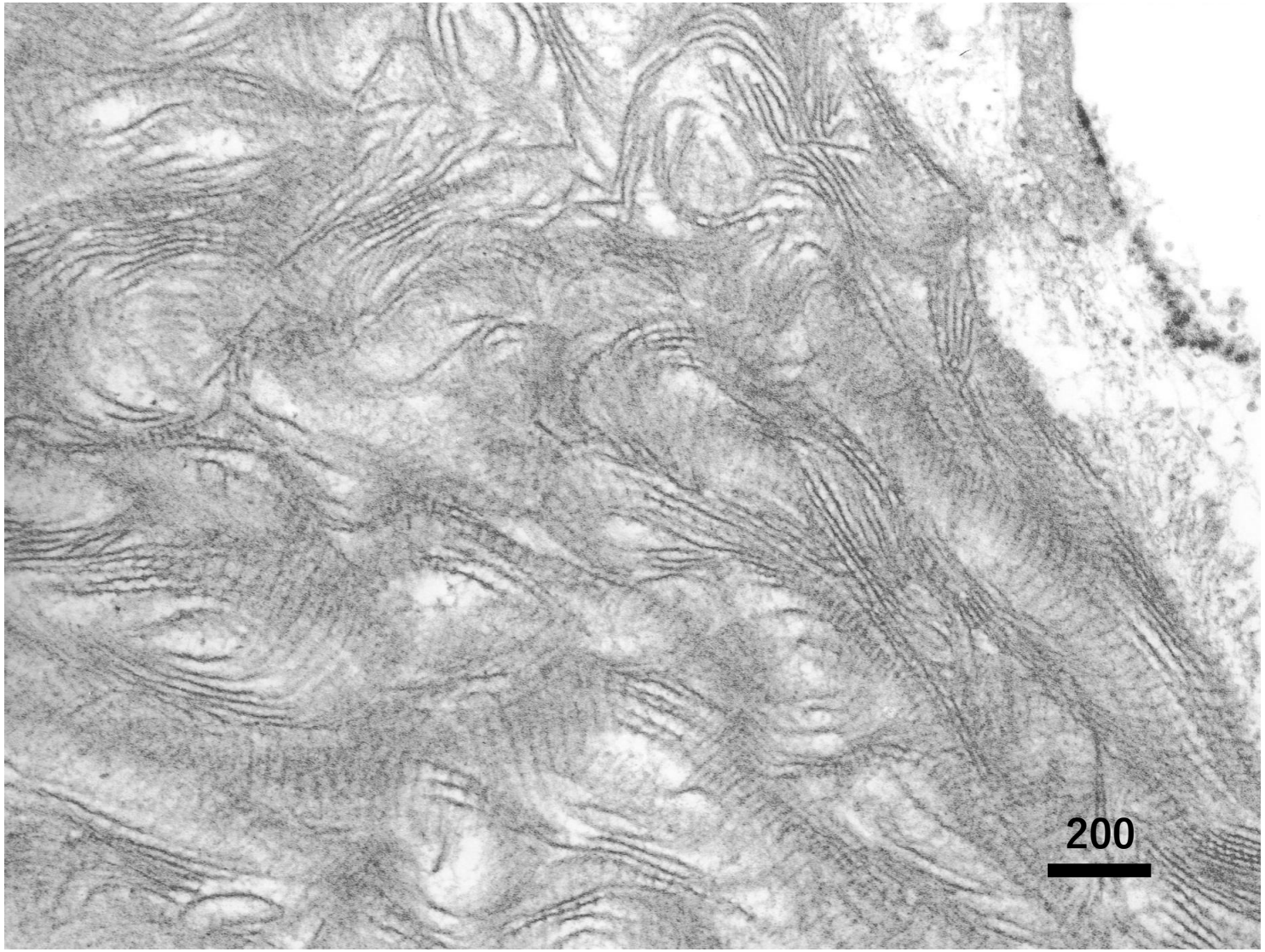
Ultrastructure of immunotactoid dermatopathy seen on the lower leg of a 61 y-o male patient. The purpuric skin lesion reveals deposition of thrombotic material in capillary vessels in the upper dermis. Fibrillar structures, focally intermingled with microtubular substances, are observed in the deposit (TEM-2)

Skin Bx



Ultrastructure of immunotactoid dermatopathy seen on the lower leg of a 61 y-o male patient. The purpuric skin lesion reveals deposition of thrombotic material in capillary vessels in the upper dermis. Fibrillar structures are observed in the deposit (TEM-3)

Skin Bx



Ultrastructure of immunotactoid dermatopathy seen on the lower leg of a 61 y-o male patient. The purpuric skin lesion reveals deposition of thrombotic material in capillary vessels in the upper dermis. Fibrillar structures with cross striations are observed in the deposit (TEM-4)