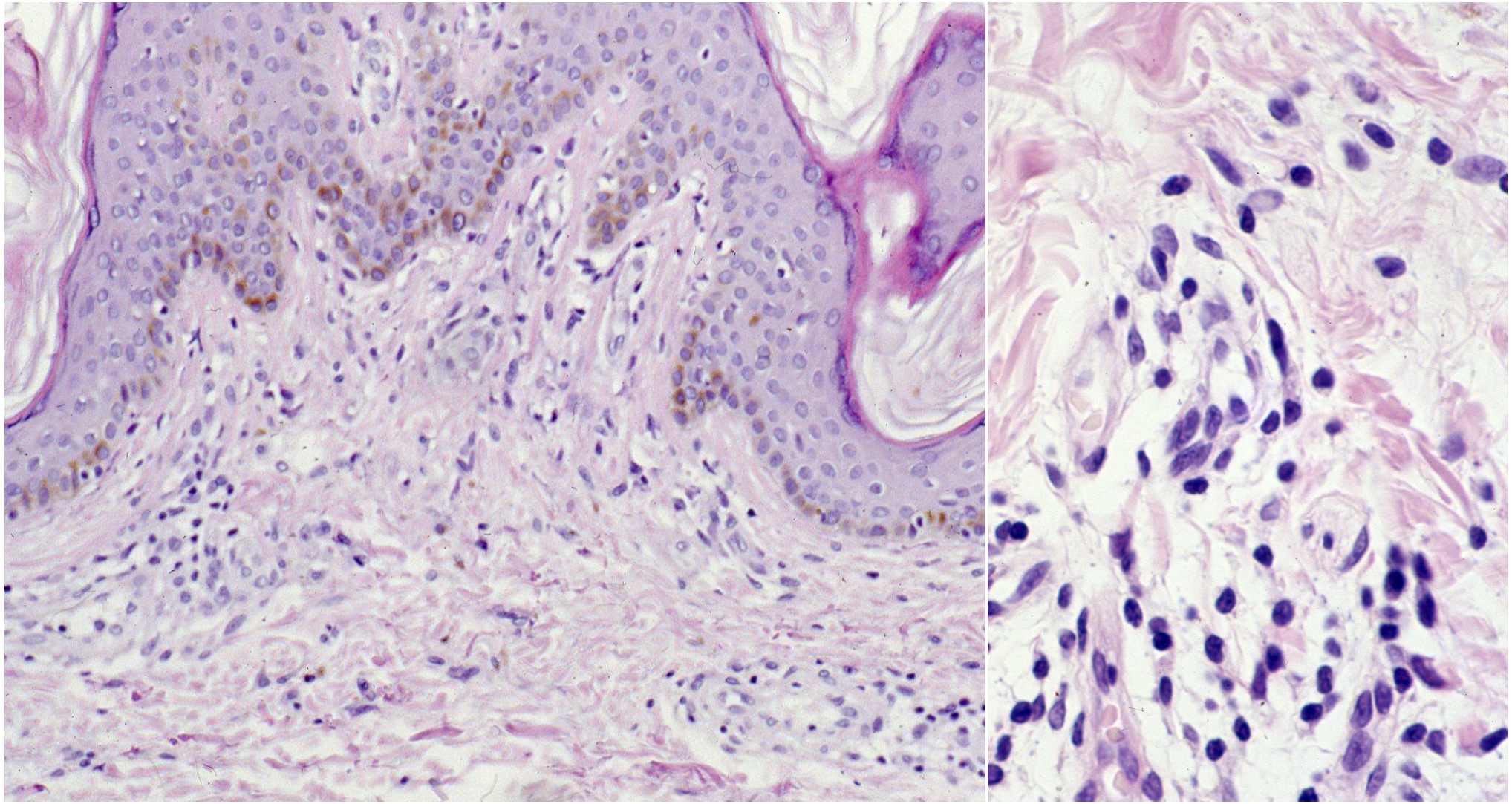
Systemic mastocytosis with urticaria pigmentosa

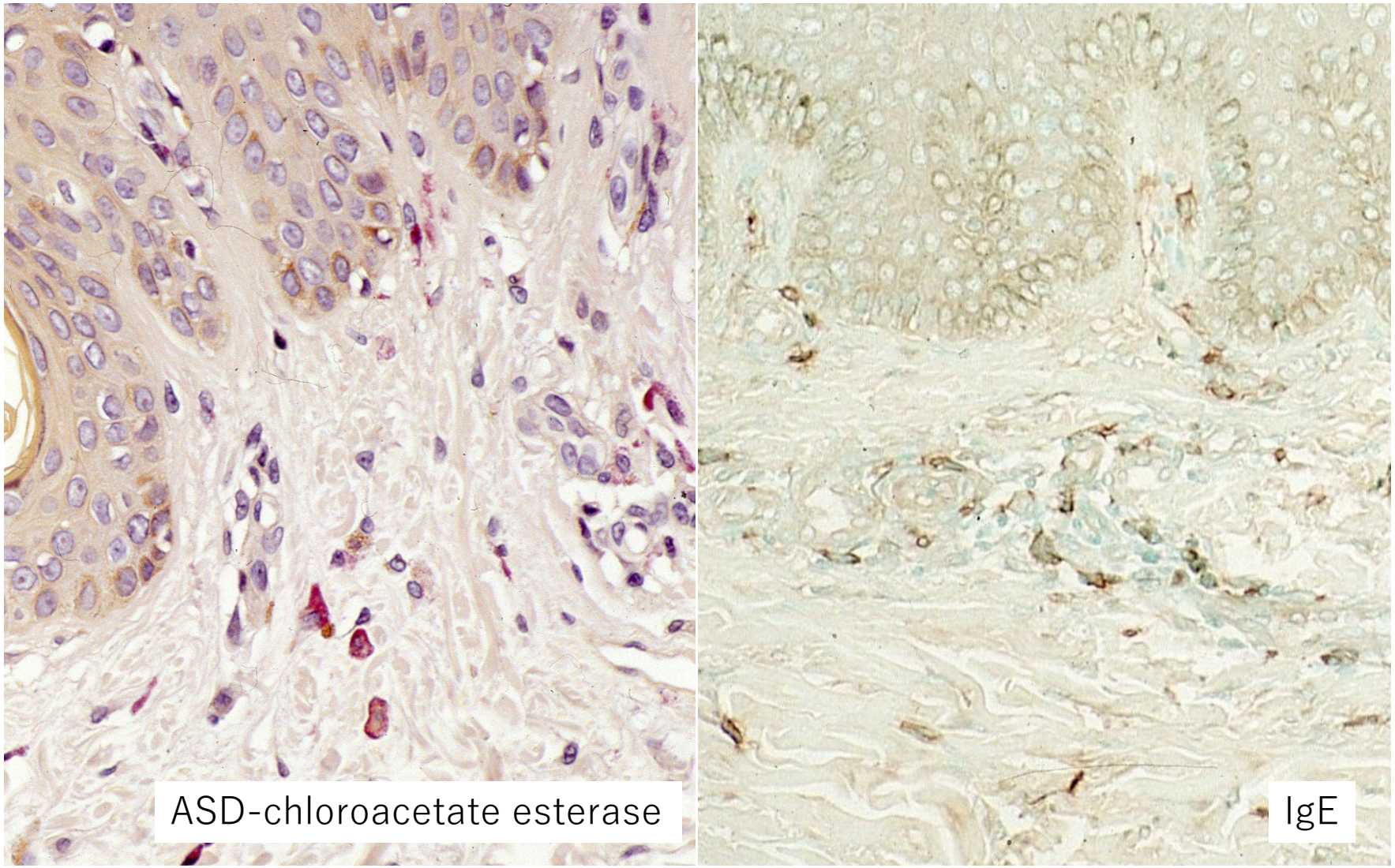
Systemic mastocytosis shows neoplastic growth of atypical mast cells involving the bone marrow, skin, liver, spleen, lymph nodes and others. Skin manifestations are common, including urticaria, flushing and dermatographism. Large osteolytic lesions with or without bone pain may be seen. Histamine-related systemic events (abdominal pain, headache, hypotension and syncope) can be associated. It occurs primarily in adults and is strongly associated with c-KIT mutations with the majority being KIT D816V. Aberrant expression of CD25, CD2 or CD30 and elevation of serum tryptase levels are observed. As cutaneous manifestations, urticaria pigmentosa, diffuse cutaneous mastocytosis or mastocytoma is experienced.

Ref.; Girton M, Aguilera N. Systemic mastocytosis. PathologyOutlines.com website. 2025. https://www.pathologyoutlines.com/topic/bonemarrowsystemicmastocytosis.html

Systemic mastocytosis seen in a 27 y-o male patient. Urticaria pigmentosa with small brown-colored patches are seen on the leg skin at autopsy. The patient acutely died of anaphylactic shock after skin flushing and diarrhea.

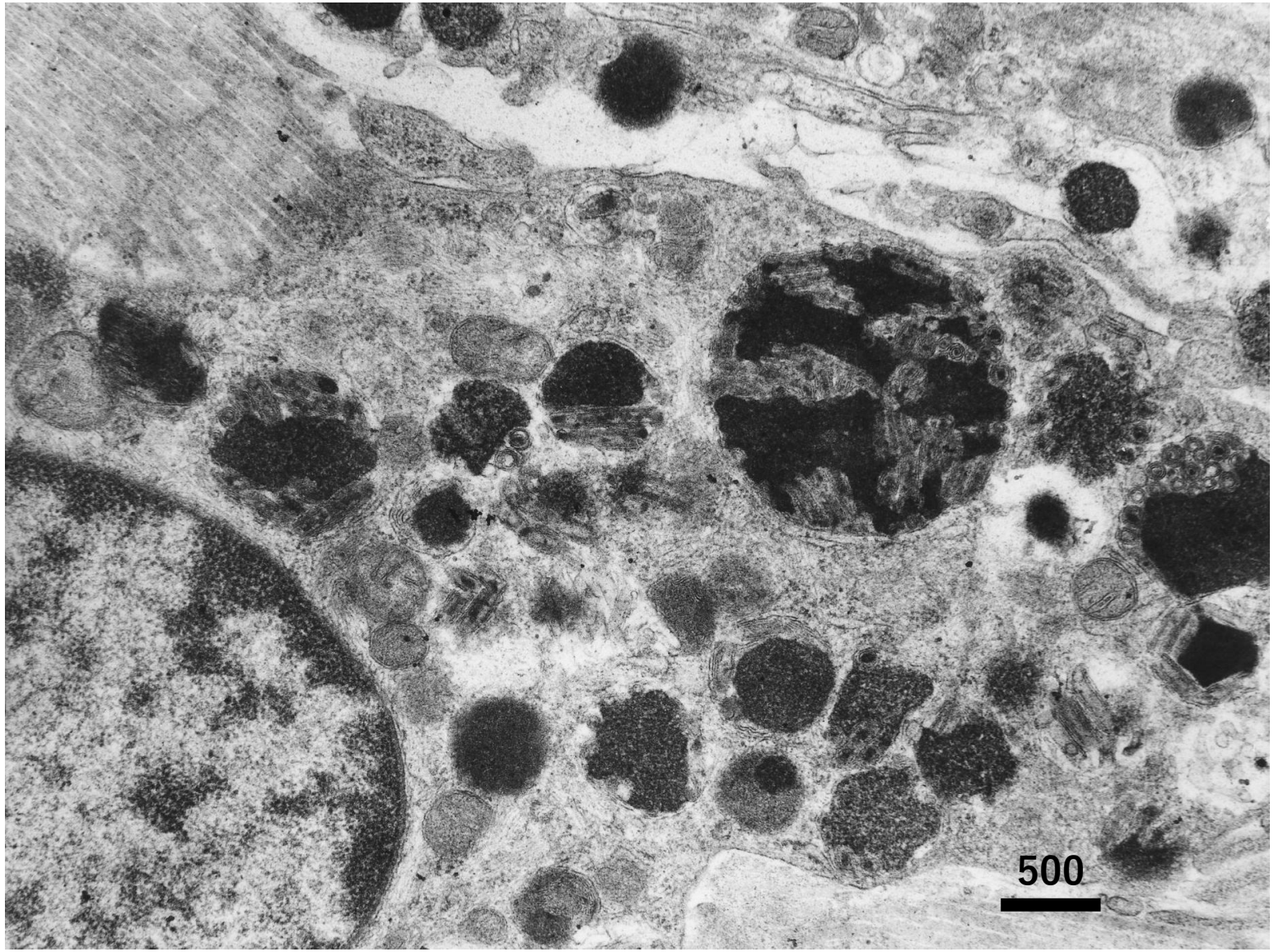


Systemic mastocytosis seen in a 27 y-o male patient. The patient acutely died of anaphylactic shock after skin flushing and diarrhea. The urticaria pigmentosa lesion on the leg skin at autopsy shows mild dermal infiltration of round cells. Mild basophilia is discerned in some mononuclear cells. Basal melanosis is associated (H&E).

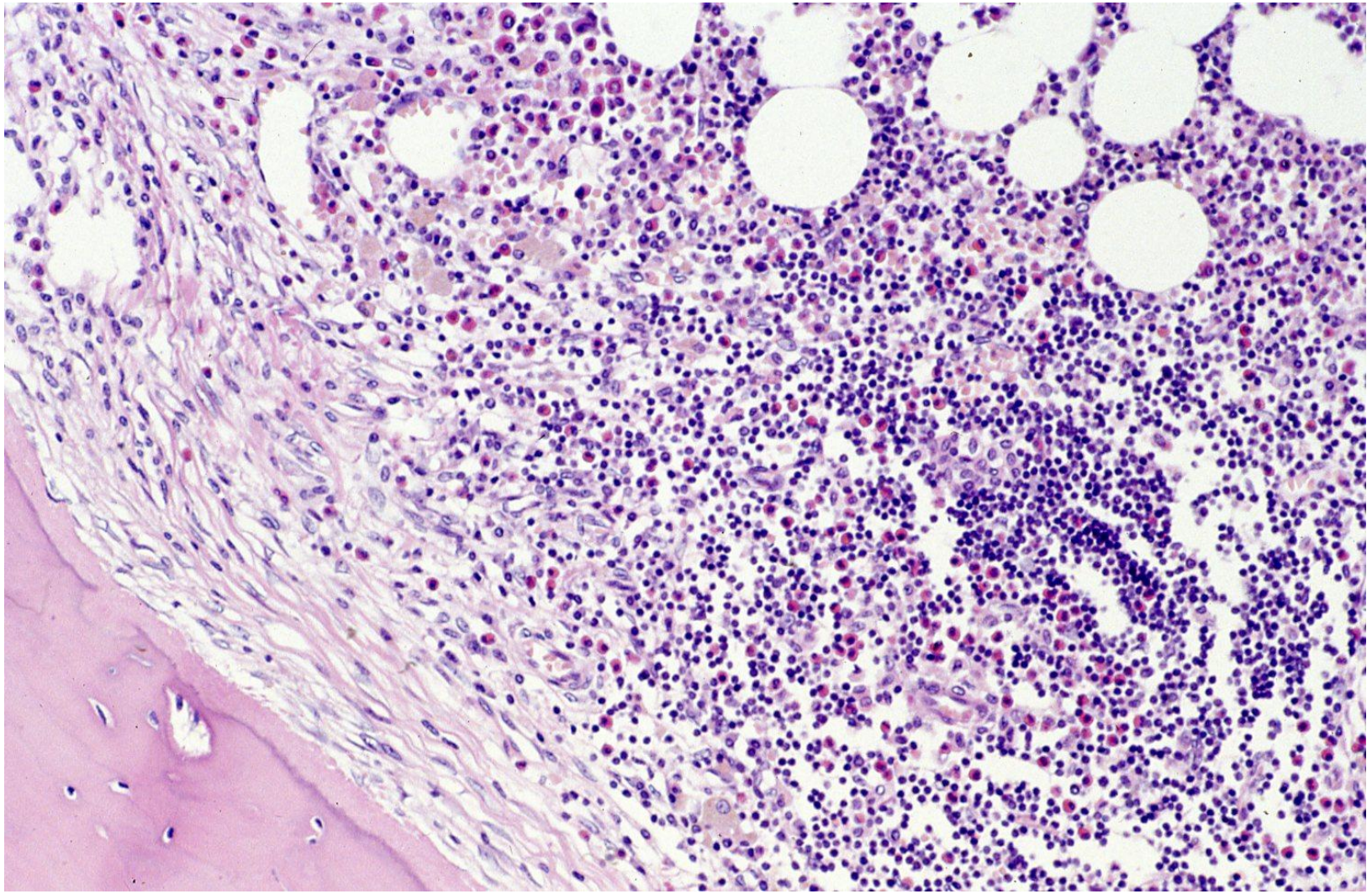


Systemic mastocytosis seen in a 27 y-o male patient. The patient acutely died of anaphylactic shock after skin flushing and diarrhea. The urticaria pigmentosa lesion on the leg skin at autopsy shows mild dermal infiltration of mast cells, demonstrated by ASD-chloroacetate esterase (left) and IgE immunostaining (right) (ASD-CAE activity and IgE immunostaining).

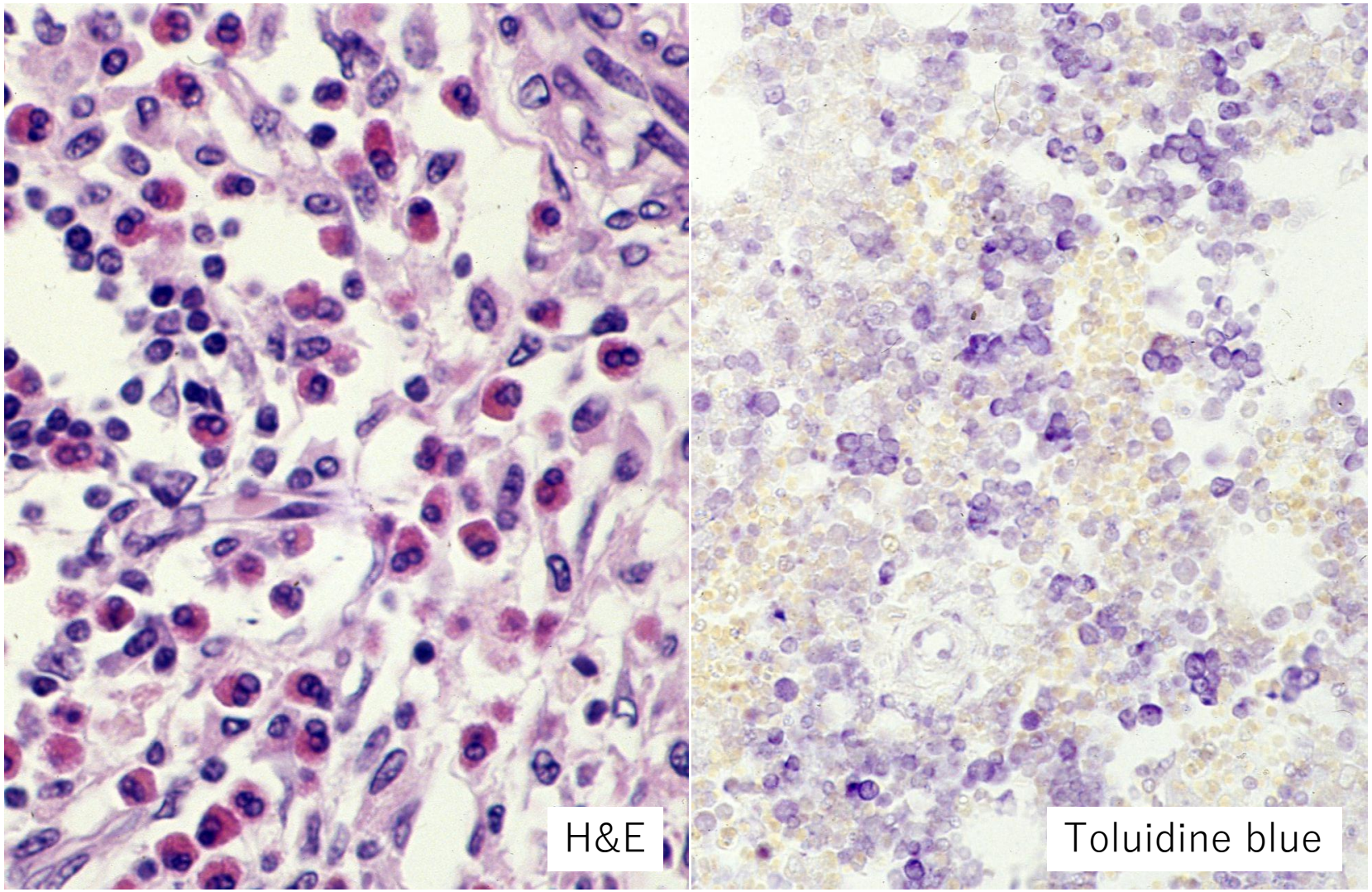
Ultrastructure of systemic mastocytosis seen in a 27 y-o male patient. The patient acutely died of anaphylactic shock after skin flushing and diarrhea. The urticaria pigmentosa lesion shows infiltration of mast cells with large-sized electron-dense granules, some with finger print-like appearance (TEM-1).



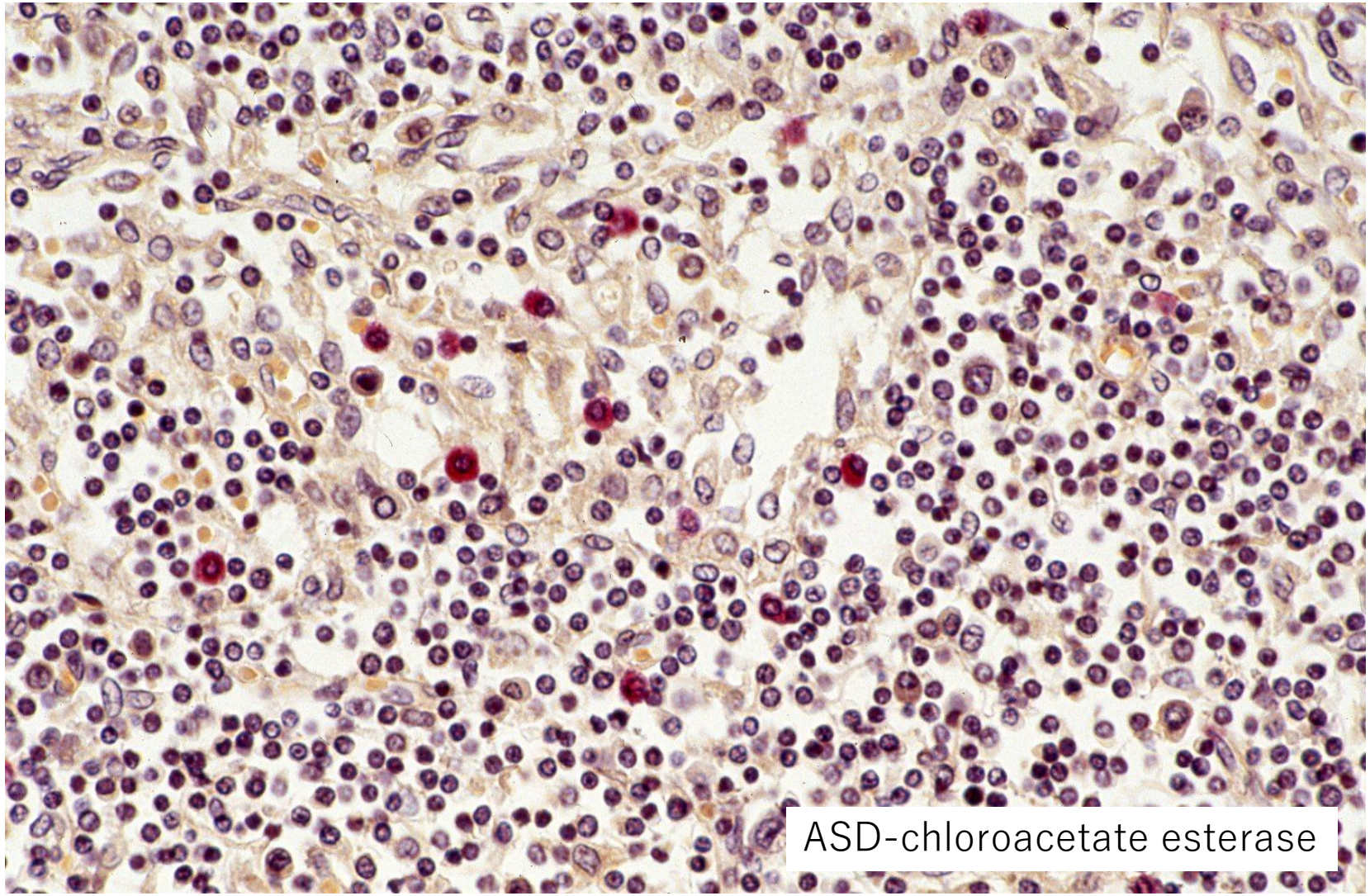
Ultrastructure of systemic mastocytosis seen in a 27 y-o male patient. The patient acutely died of anaphylactic shock after skin flushing and diarrhea. The urticaria pigmentosa lesion shows infiltration of mast cells with large-sized electron-dense granules, some with finger print-like appearance (TEM-2).



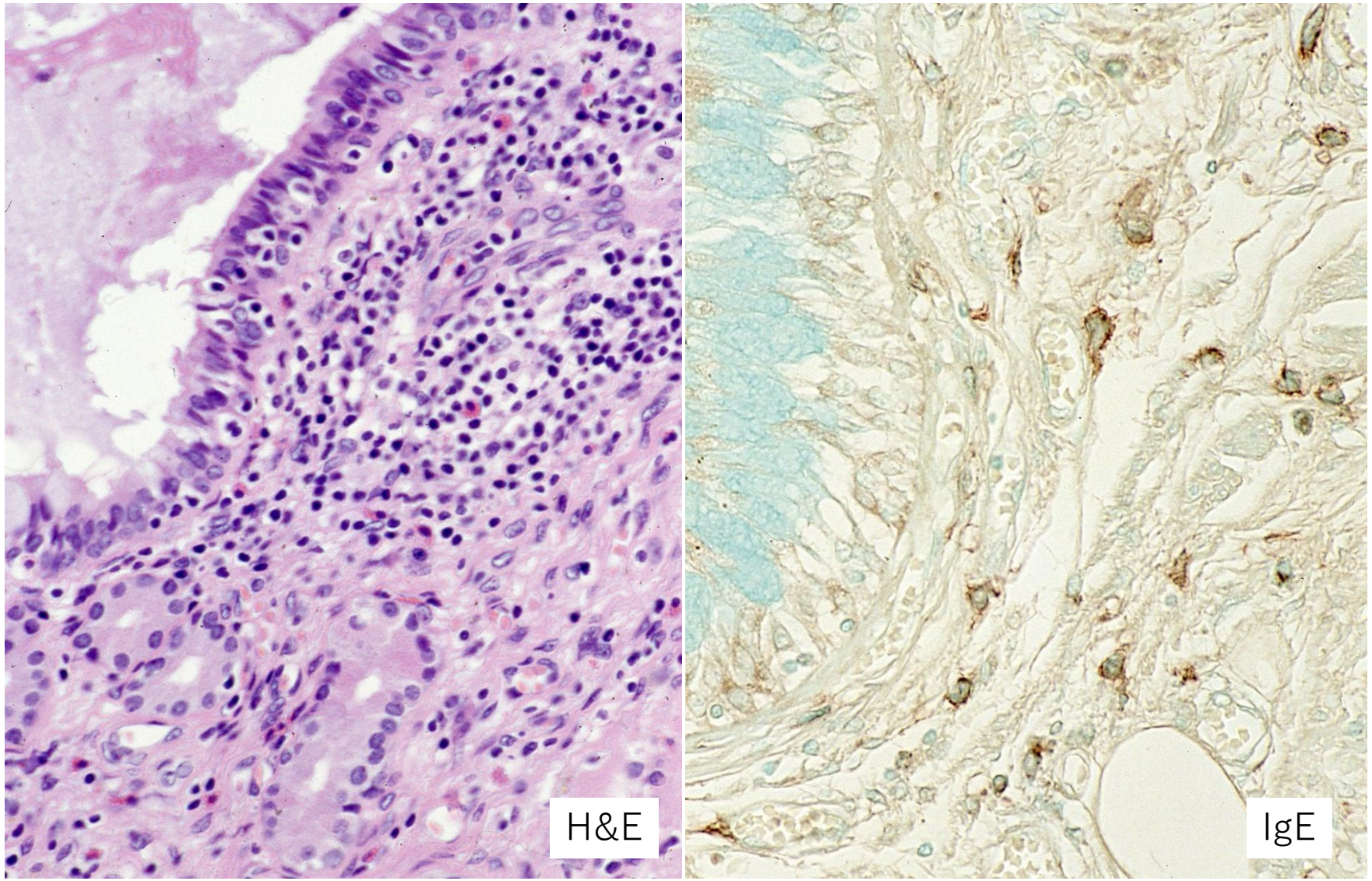
Systemic mastocytosis seen in a 27 y-o male patient. The patient acutely died of anaphylactic shock after skin flushing and diarrhea. The bone marrow is mildly cellular with periosteal fibrosis. Eosinophilic infiltration is diffusely observed (H&E).



Systemic mastocytosis seen in a 27 y-o male patient. The patient acutely died of anaphylactic shock after skin flushing and diarrhea. The bone marrow shows infiltration of mast cells with finely basophilic cytoplasm (left: H&E). Metachromatic granules are seen in the infiltrating mast cells (right; Toluidine blue).



Systemic mastocytosis seen in a 27 y-o male patient. The patient acutely died of anaphylactic shock after skin flushing and diarrhea. The retroperitoneal lymph node at autopsy shows mild parenchymal infiltration of mast cells, revealed by ASD-chloroacetate esterase staining (red-colored granules seen in the cytoplasm).



Systemic mastocytosis seen in a 27 y-o male patient. The patient acutely died of anaphylactic shock after skin flushing and diarhea. The nasal mucosa is infiltrated by mast cells intermingled with eosinophils (H&E: left). The bronchial wall in the lung is also infiltrated by mast cells revealed by IgE immunostaining (right: IgE).