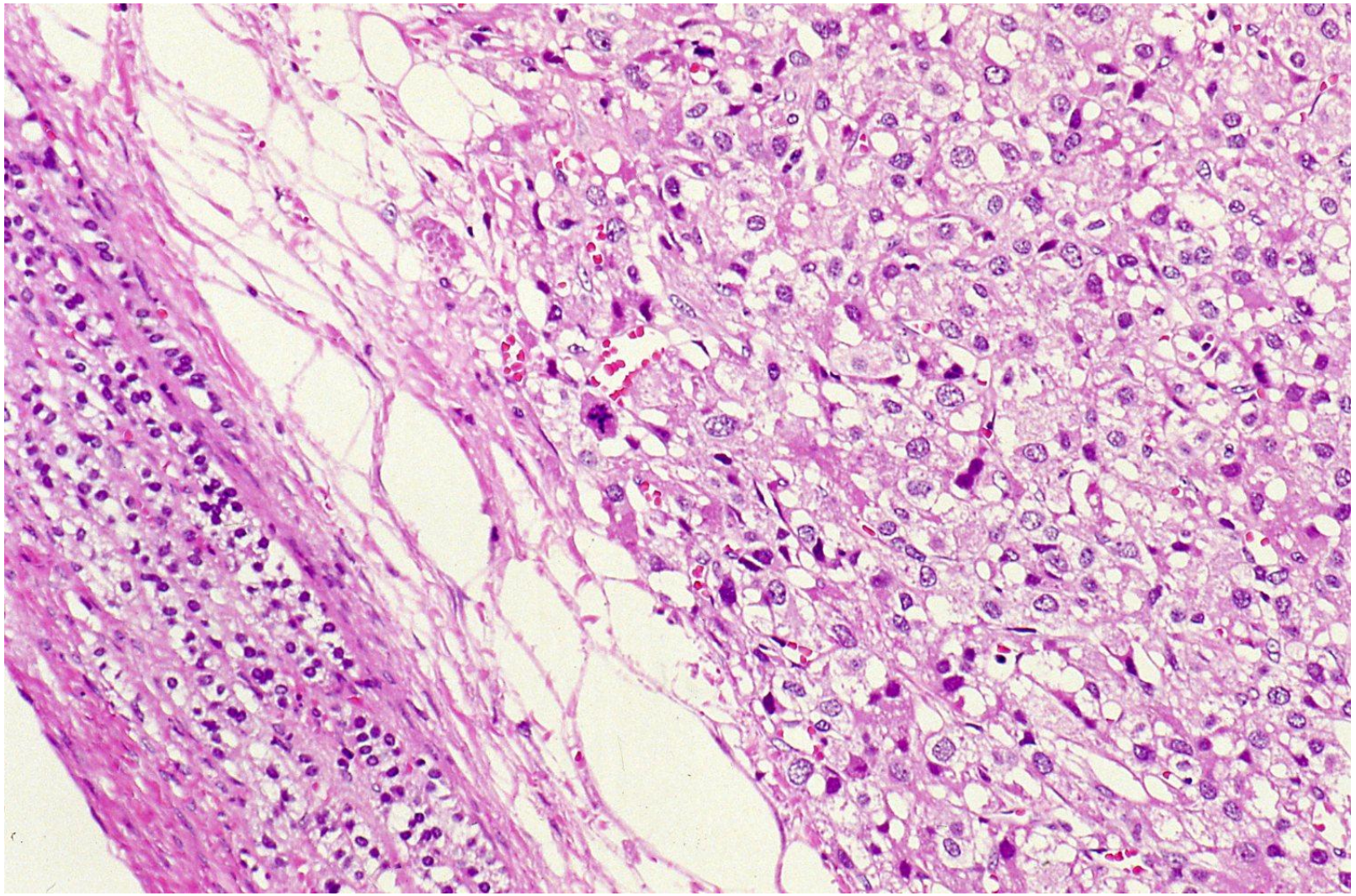
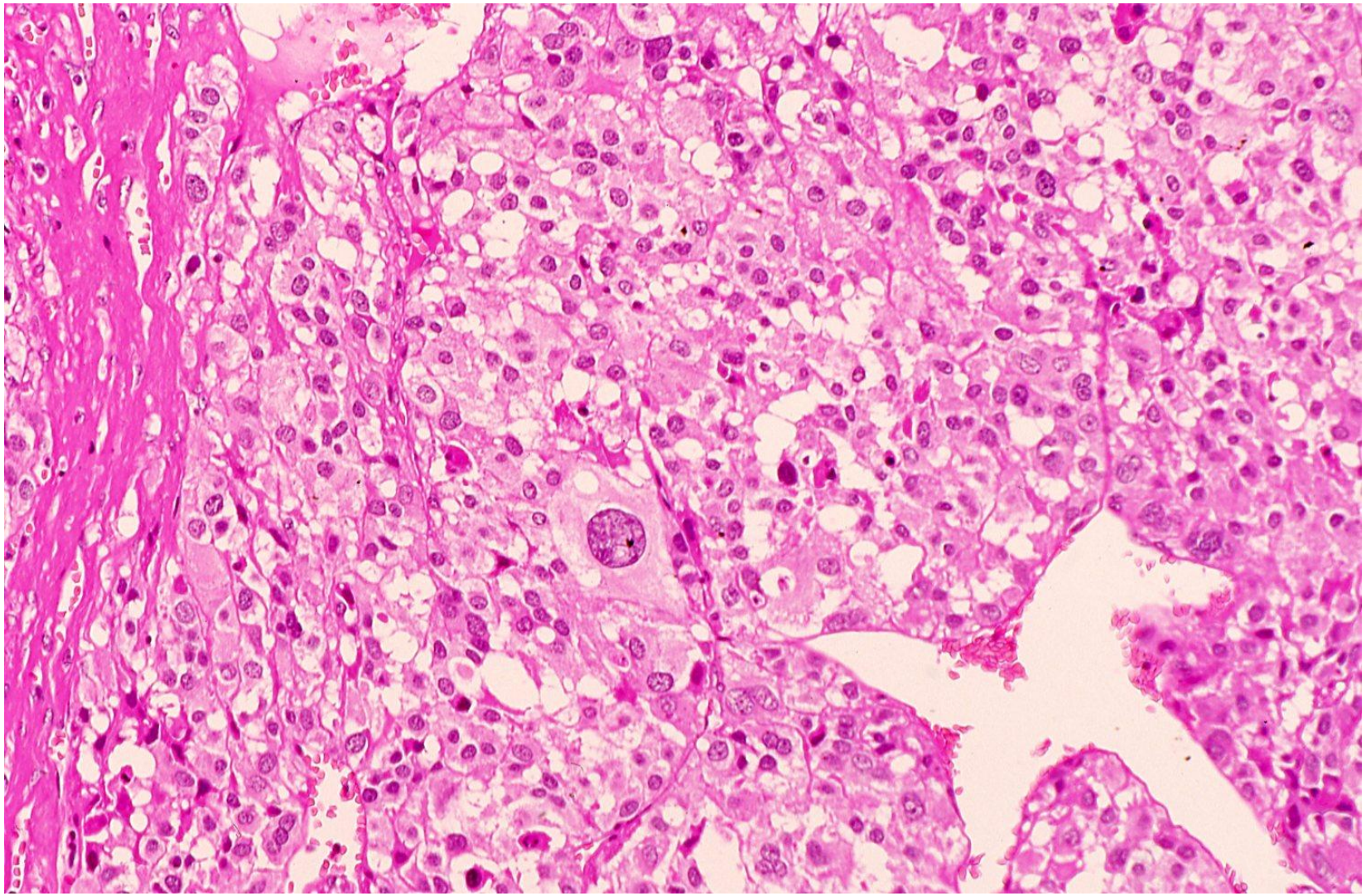
Functioning adrenocortical carcinoma of the infant

Adrenocortical carcinoma (ACC) in infancy present as a unique entity hormonal activity secreting excessive adrenocortical hormones with typical clinical syndromes. The clinical behavior and biological principles of infantile ACC can be different from tumors of adults and young patients (<4 years). Age <4 years at diagnosis has been described as a favorable factor. Grossly, ACC tends to be large, lobulated, yellow masses with heterogeneous cut surfaces and fibrous capsule. Infantile ACC derives from the fetal zone of the adrenal gland, explaining the early age of onset and hormonal secretion, causing virilization due to androgen production. The prognostic pathological indicators include large tumor size, vascular invasion, atypical mitosis, aneuploidies, tumor necrosis, high mitotic index and high Ki-67 labeling index, Molecular prognostic markers include GATA4, SF1, DAX1, IGF1R, IGF2, Aurora kinase A/B, YAP1, placental ALP, p21, beta-catenin and p53. Infantile ACC seen in a 1 y-o girl is presented. The genitals somewhat resembled those of infant boys (virilization).

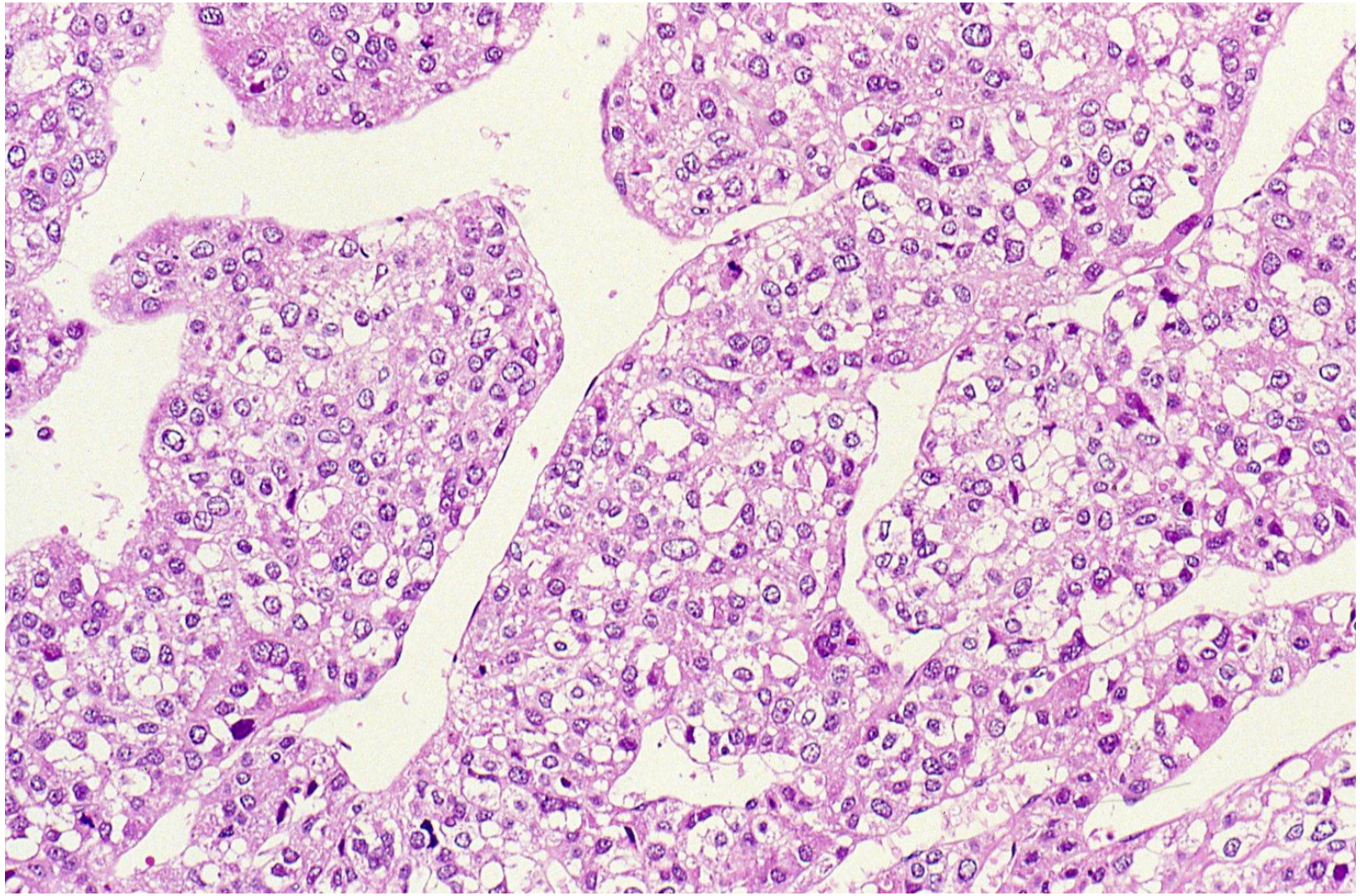
Ref.: Riedmeier M, et al. Adrenocortical carcinoma in childhood: a systematic review. Cancers (Basel) 2021; 13(21): 5266. doi: 10.3390/cancers13215266



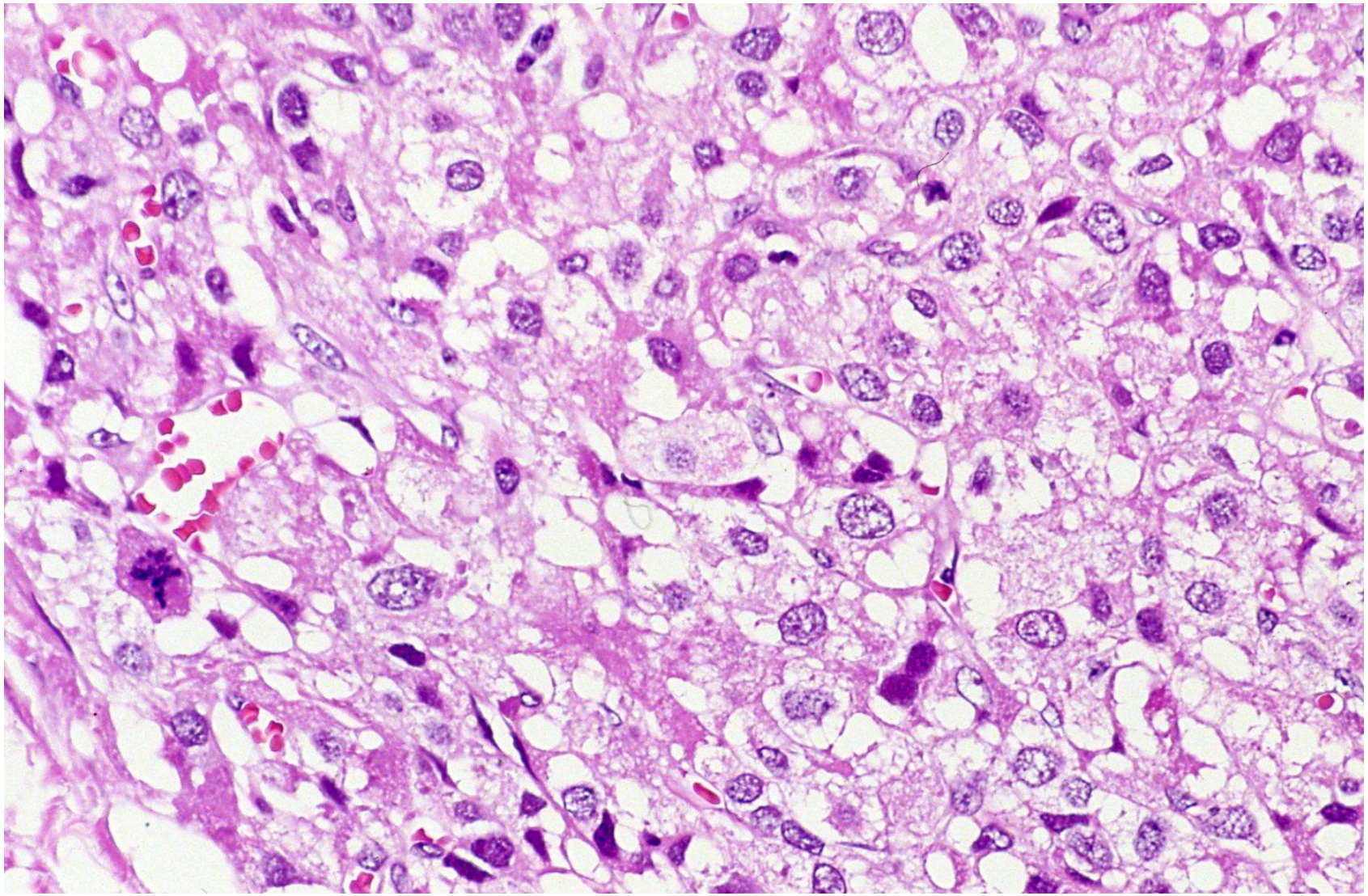
Adrenocortical carcinoma seen in a l y-o girl. The surgical specimen shows solid growth of atypical clear cells with anisonucleosis. Mitoses are scattered. Non-neoplastic adrenal cortex is compressed (H&E-1).



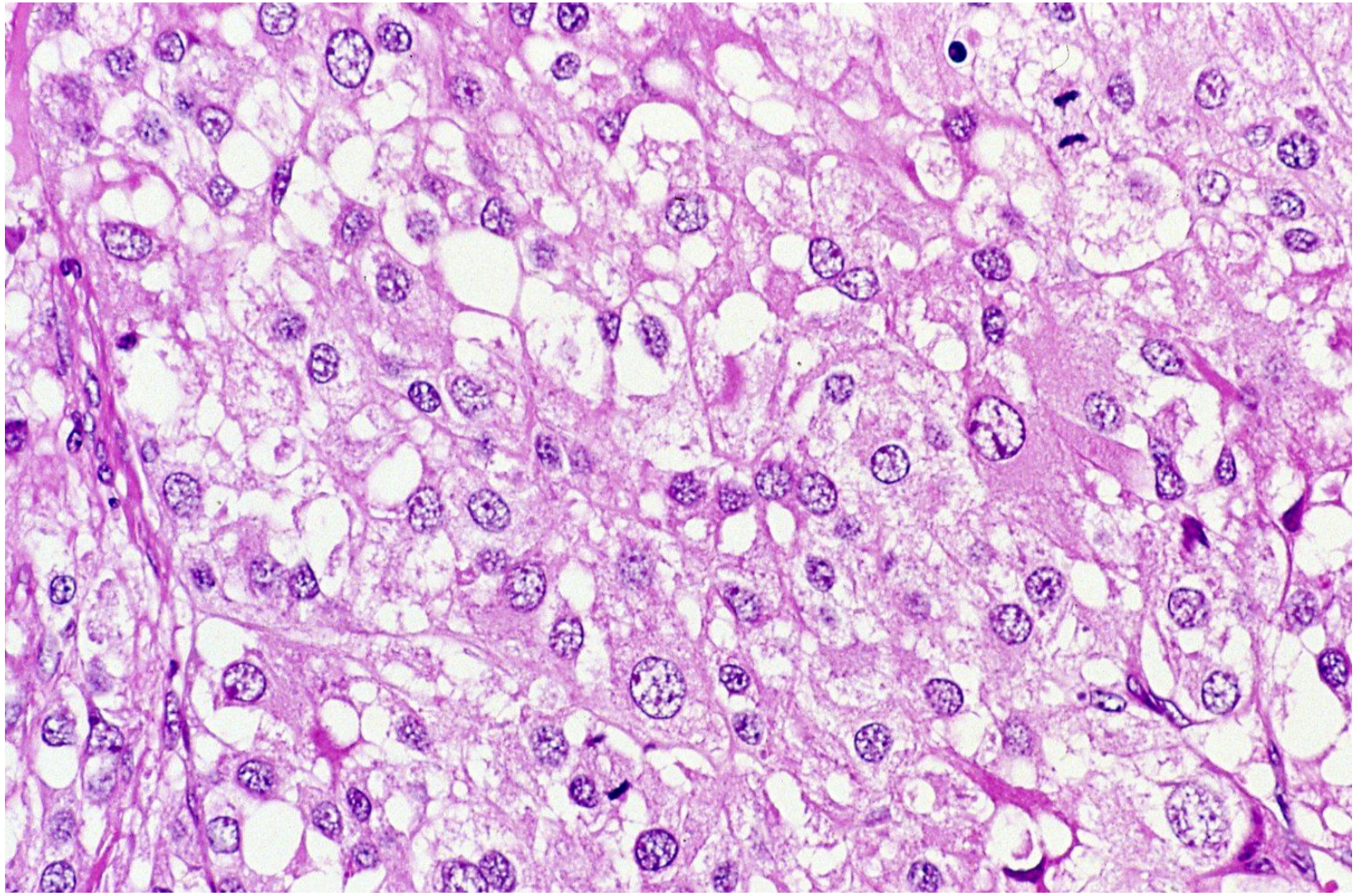
Adrenocortical carcinoma seen in a I y-o girl. The surgical specimen shows solid growth of atypical clear cells with anisonucleosis. A bizarre nucleus is noted. Sinusoidal vessels are distributed (H&E-2).



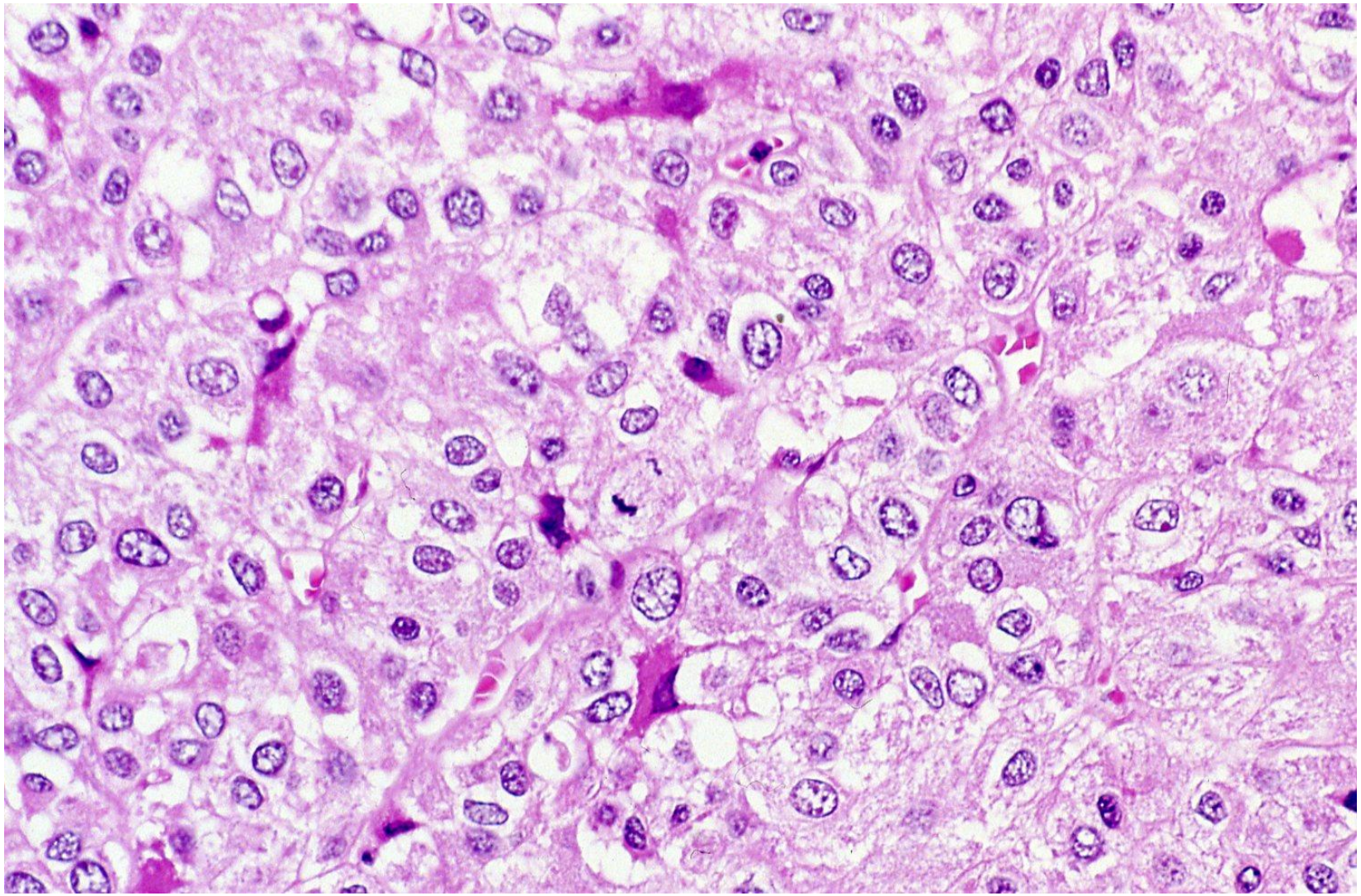
Adrenocortical carcinoma seen in a l y-o girl. The surgical specimen shows solid growth of atypical clear cells with mild anisonucleosis. Sinusoidal vessels are well developed (H&E-3).



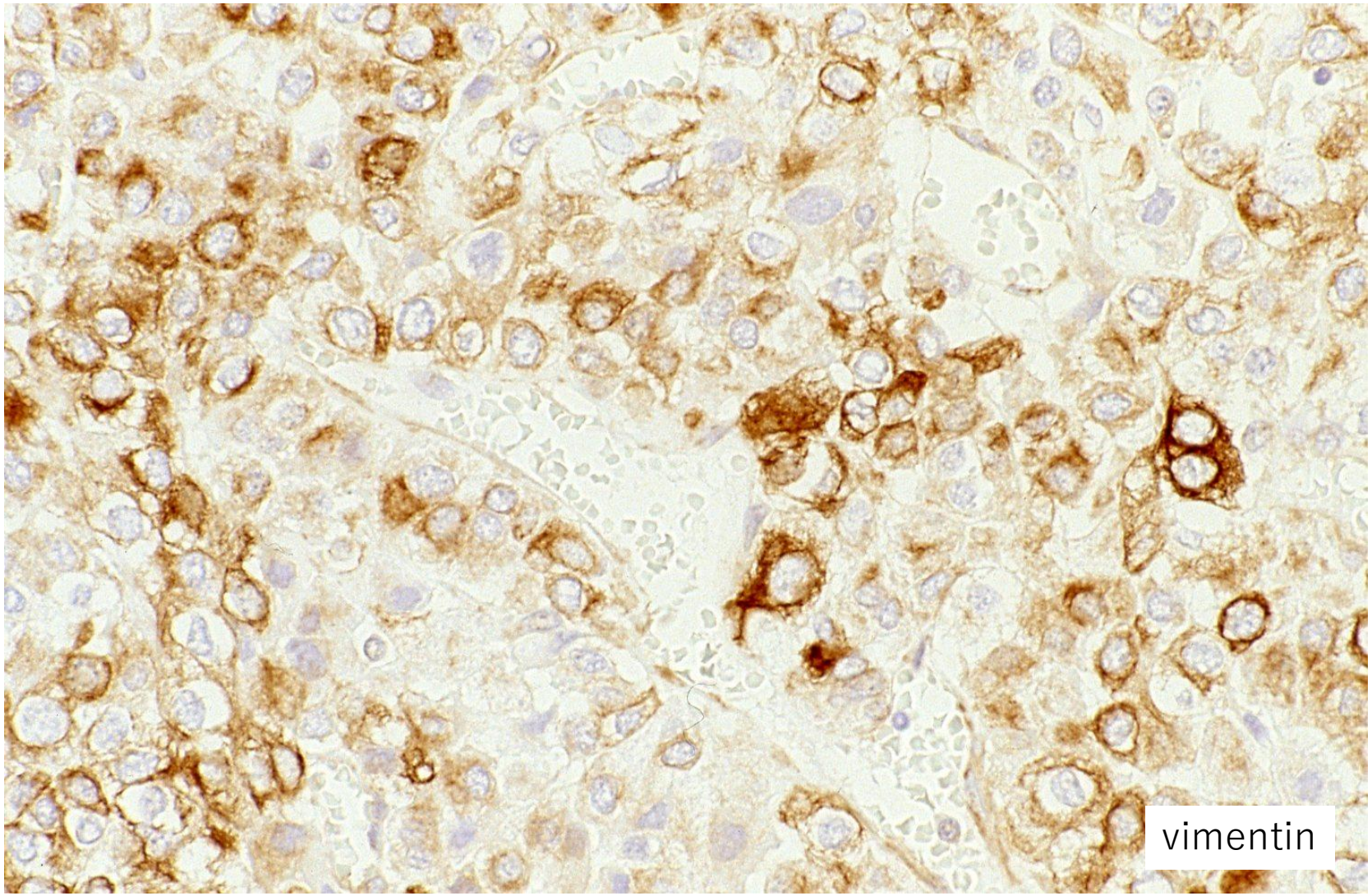
Adrenocortical carcinoma seen in a l y-o girl. The surgical specimen shows solid growth of atypical clear cells with anisonucleosis. Mitoses are scattered. The vascularity is high (H&E-4).



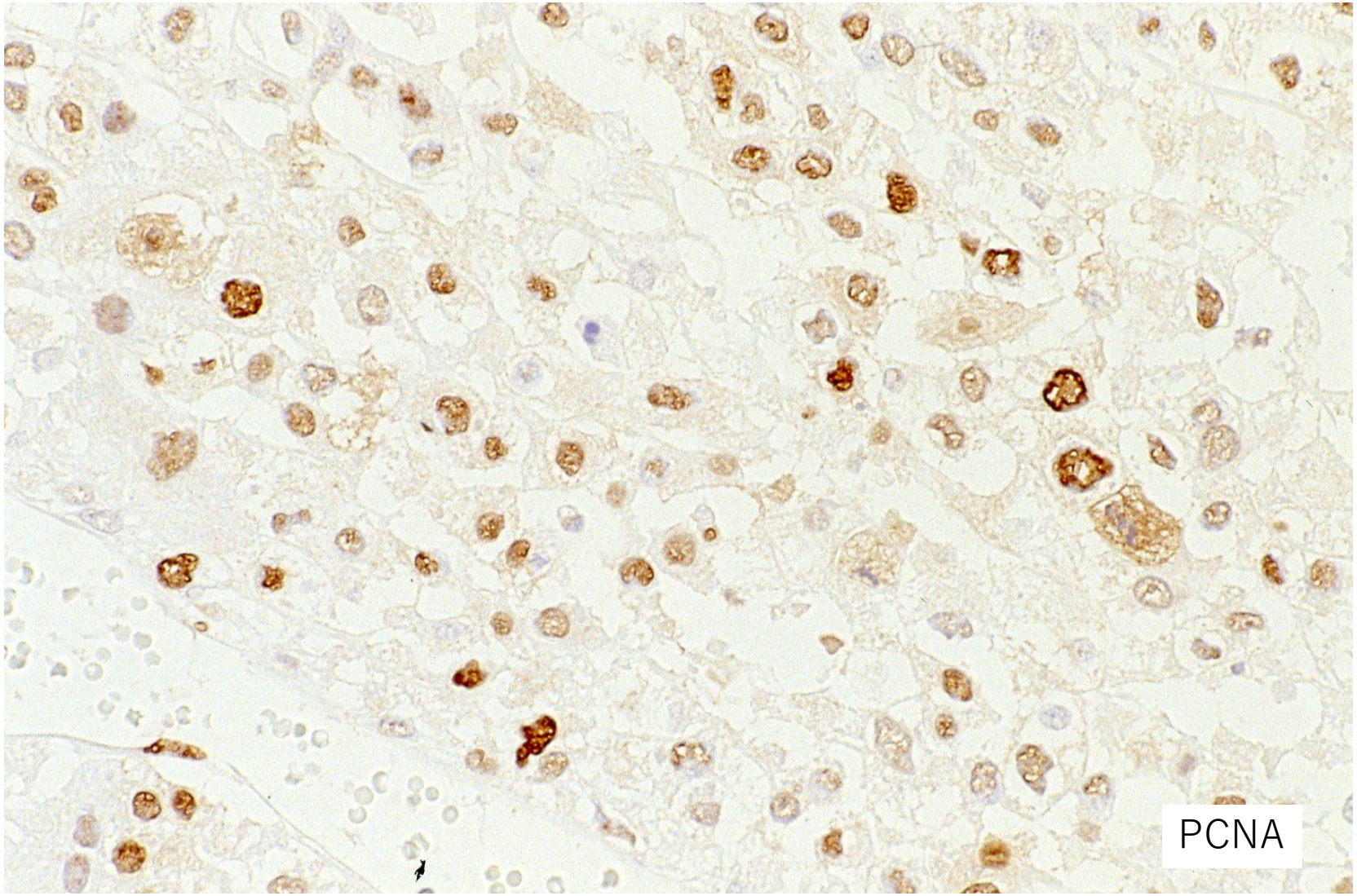
Adrenocortical carcinoma seen in a l y-o girl. The surgical specimen shows solid growth of atypical clear cells with mild anisonucleosis. Eosinophilic cytoplasm is also noted. The vascularity is high (H&E-5).



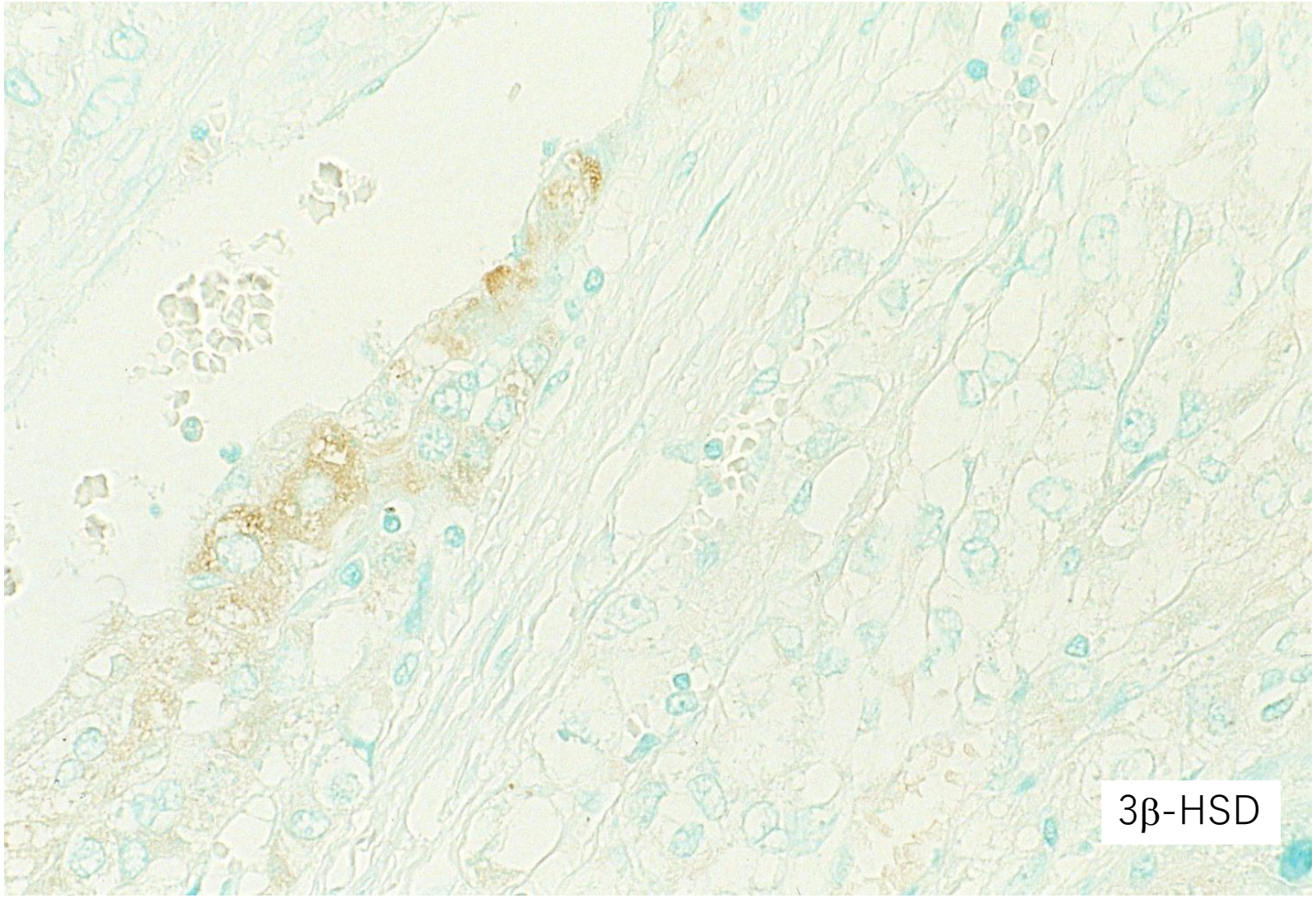
Adrenocortical carcinoma seen in a l y-o girl. The surgical specimen shows solid growth of atypical clear cells with mild anisonucleosis. The vascularity is high and mitoses are scattered (H&E-6).



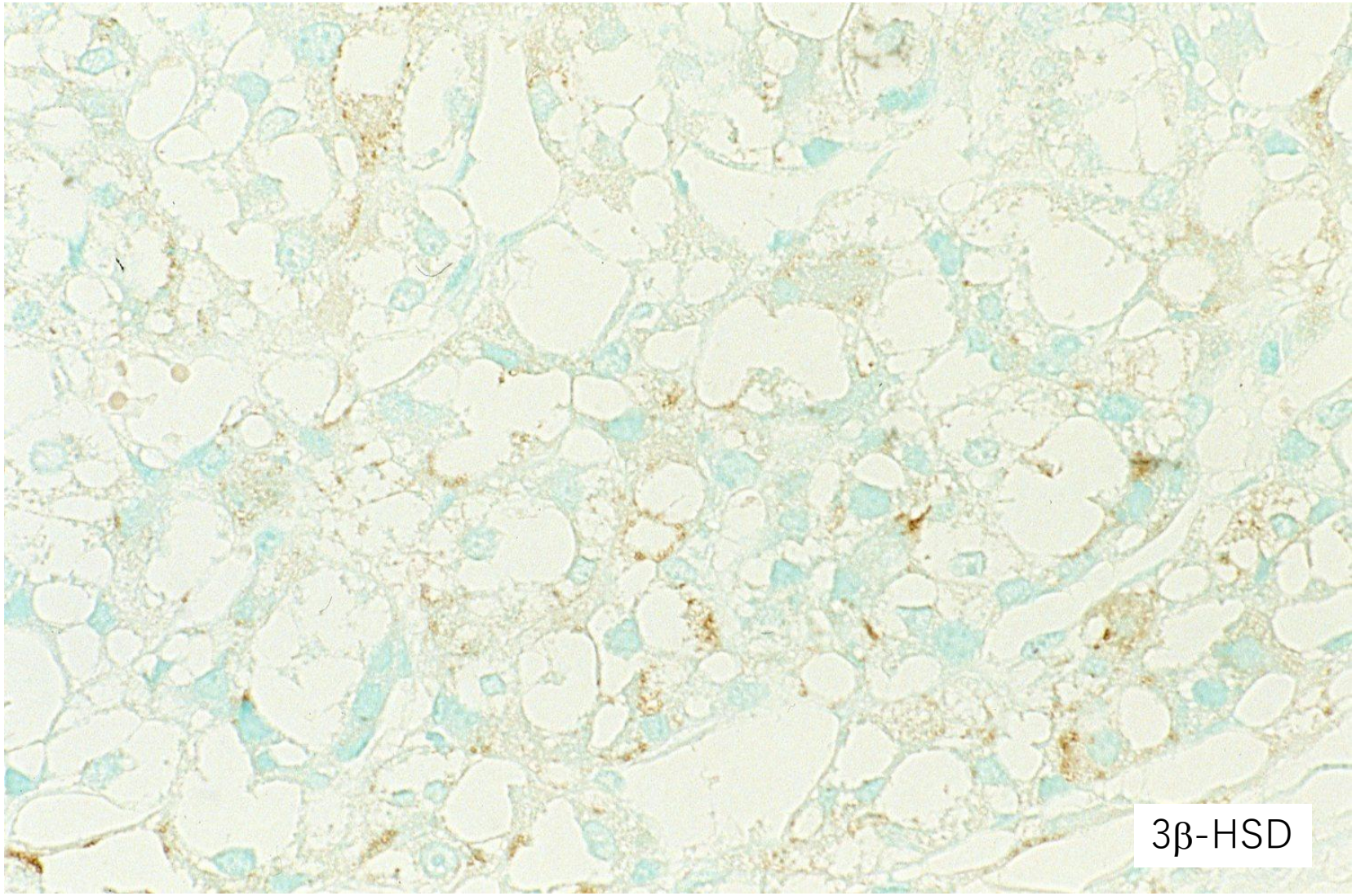
Adrenocortical carcinoma seen in a I y-o girl. The adrenocortical tumor cells in the surgical specimen are immunoreactive for vimentin (immunostaining for vimentin).



Adrenocortical carcinoma seen in a l y-o girl. The adrenocortical tumor cells in the surgical specimen are highly proliferative. More than half tumor cells are labeled for proliferating cell nuclear antigen (PCNA) (immunostaining for PCNA).



Adrenocortical carcinoma seen in a l y-o girl. The adrenocortical tumor cells in the surgical specimen are focally immunoreactive for 3β-hydroxysteroid dehydrogenase (3β-HSD), strongly suggesting a functioning tumor. 3β-HSD is the key enzyme in the adrenal pathway of corticosteroid synthesis (immunostaining for 3β-HSD).



Adrenocortical carcinoma seen in a l y-o girl. The adrenocortical tumor cells in the surgical specimen are immunoreactive for 3β-hydroxysteroid dehydrogenase (3β-HSD), strongly suggesting a functioning tumor. 3β-HSD is the key enzyme in the adrenal pathway of corticosteroid synthesis (immunostaining for 3β-HSD).