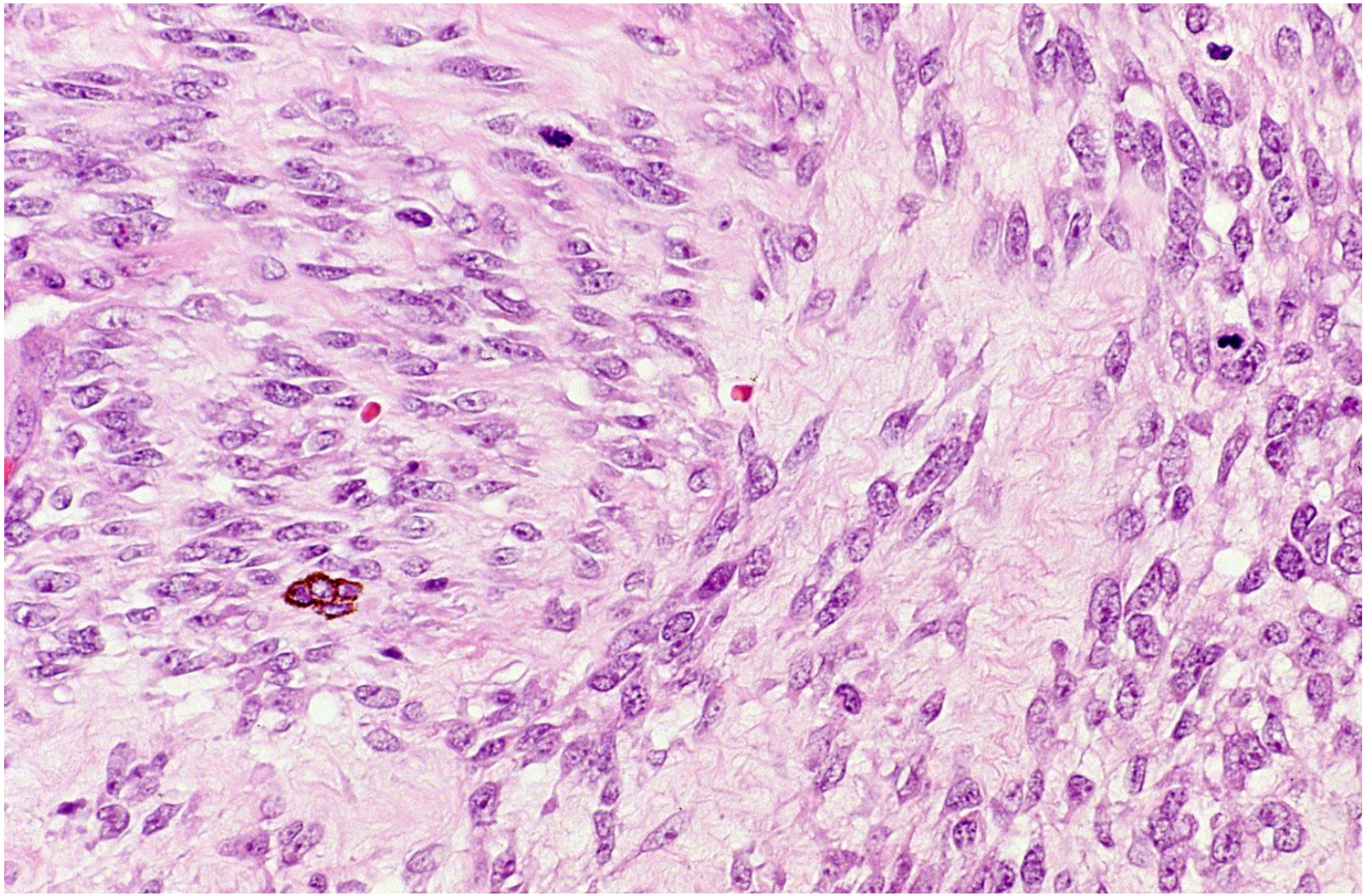
Pigmented perivascular epithelioid cell tumor of the skin

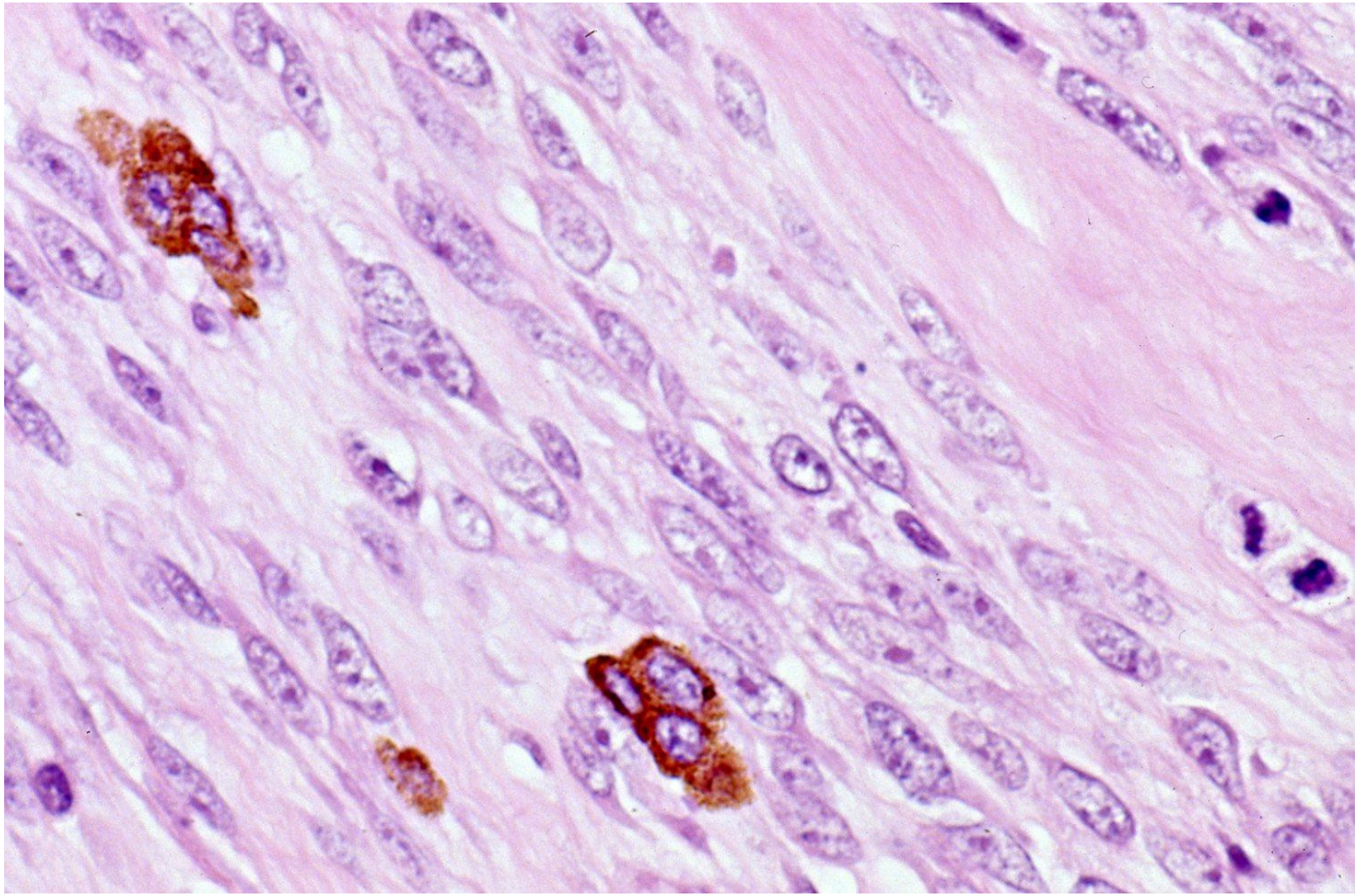
Navale et al reported in 2015 a case of pigmented perivascular epithelioid cell tumor (PEComa) of the skin. A subcutaneous tumor of the flank microscopically resembled renal clear cell carcinoma but with focal melanin pigmentation and strong positivity for HMB-45. The lesion was mistaken for metastatic melanoma. PEComa is a rare mesenchymal tumor defined by the presence of perivascular epithelioid cells expressing both smooth muscle markers (SMA, desmin and caldesmon) and melanocytic markers (HMB45, melan A and MiTF). PEComa is commonly seen in the gynecologic tract: the uterine corpus is the most common site, and may also occur in the cervix, vagina, ovary, broad ligament and vulva. Renal angiomyolipoma and pulmonary lymphangioleiomyomatosis/clear cell sugar tumor are included in the PEComa group. Pigmented PEComa is occasionally seen in the kidney. Cutaneous PEComas are very rare. Here presented is a nostril spindle cell tumor seen in a 37 y-o female patient. The spindle cells with active mitosis expressed smooth muscle markers, and a small number of melanin-containing melanocytes are distributed in the tumor matrix. The possibility of pigmented PEComa of the cutaneous origin is highly likely. Regrettably enough, only two microphotographs are left in hand.

Ref.-1: Navale P, et al. Pigmented perivascular epithelioid cell tumor of the skin: first case report. Am J Dermatopathol 2015; 37(11): 866-869. doi: 10.1097/DAD.0000000000000320

Ref.-2: Chang H, et al. Pigmented perivascular epithelioid cell tumor (PEComa) of the kidney: a case report and review of the literature. Korean J Pathol 2012; 46(5): 499-502. doi: 10.4132/KoreanJPathol.2012.46.5.499



Cutaneous pigmented PEComa of the nostril in a 37 y-o female patient. The spindled tumor cells express smooth muscle markers (SMA and desmin). Melanin-containing melanocytes are scattered among the tumor cells. HMB45 was not evaluated, but the possibility of pigmented PEComa is highly likely (H&E-1).



Cutaneous pigmented PEComa of the nostril in a 37 y-o female patient. The spindled tumor cells express smooth muscle markers (SMA and desmin). Mitoses are scattered. Melanin-containing melanocytes are scattered among the tumor cells. HMB45 was not evaluated, but the possibility of pigmented PEComa is highly likely (H&E-2).