The 2022 World Health Organization classification of tumors of the pituitary gland distinguishes the anterior lobe of the pituitary gland (Adenohypophysis) from the posterior lobe (neurohypophysis) and hypothalamic tumors.

Anterior lobe tumors include (i) well-differentiated adenohypophyseal tumors that are now classified as pituitary neuroendocrine tumors (PitNETs; formerly known as pituitary neuroendocrine tumors), (ii) pituitary blastoma, and (iii) the two types of craniopharyngioma.

Tumor of the neonatal pituitary exhibits differentiation to Rathke epithelium and adenohypophysial cells of folliculostellate and secretory type, a reflection of arrested pituitary development and unchecked proliferation (Scheithauer et al. in Acta Neuropathol 116(6):657-666, 2008). Herein, we report the pathologic features of three additional cases, all ACTH-producing. One involved a 9-month-old male presenting with progressive right ophthalmoplegia, MRI findings of a large suprasellar mass with cavernous sinus invasion, and elevated plasma ACTH levels. The second was nonfunctioning and occurred in a 13-month-old female with right third nerve palsy. The third had been previously published as a “pituitary neuroendocrine tumor” in a 2-year-old female (Min et al. in Pathol Int 57(9):600-605, 2007). The subtotally resected tumors were subject to histochemical, immunohistochemical and, in two cases, ultrastructural study. Histologically, the complex tumors consisted of glands of varying from rosettes to glandular structures resembling Rathke epithelium, small undifferentiated-appearing cells (blastema), and large secretory cells. Mucin-producing goblet cells were noted in case 3. Cell proliferation was high in two cases and low in case 3. Immunoreactivity of the secretory cells included synaptophysin, chromogranin, various keratins and, to a lesser extent, ACTH and beta endorphin. MGMT immunolabeling was 40-60%. Mitotic activity was moderate to high in cases 1 and 2 and was low in case 3. The same was true for MIB-1 labeling. Germ cell markers were lacking in all cases. One tumor ultrastructurally consisted of three cell populations including (a) small, polyhedral, primitive-appearing cells (blastema) with scant cytoplasm, abundant glycogen and few organelles, (b) folliculostellate cells and © large corticotroph cells containing rough endoplasmic reticulum, golgi membranes, spherical, 150-400 nm secretory granules and occasional perinuclear, intermediate filament bundles. A second example (case 3) lacked a blastema and glandular component. The clinical and morphologic features of our three cases were those of pituitary blastoma. The finding of cellular elements of adenohypophysial development is consistent with a diagnosis of pituitary blastoma and aligns it with blastomas of other organs. It also suggests an underlying specific genetic abnormality. Marked variations in cellular proliferative activity suggest blastomas occur in low- and higher-grade form. Variable MGMT reactivity suggests an incomplete response to temozolomide therapy. Literature regarding similar morphologically complex, infantile, Cushing disease-associated lesions is briefly reviewed [1)].

1)
Scheithauer BW, Horvath E, Abel TW, Robital Y, Park SH, Osamura RY, Deal C, Lloyd RV, Kovacs K. Pituitary blastoma: a unique embryonal tumor. Pituitary. 2012 Sep;15(3):365-73. doi: 10.1007/s11102-011-0328-x. PubMed PMID: 21805093.