Atypical Teratoid/Rhabdoid Tumor (AT/RT) in the Sellar Region

AT/RTs are rare malignant neoplasms that almost always appear in patients less than three years, often below two, or even at or less than one. A small fraction of an already rare tumor occurs in the AT/RT tumor disposition syndrome. The lesion is at least as common in the posterior fossa, e.g. cerebellopontine angle, as in the supratentorial compartment.

AT/RT is thus familiar to pediatric pathologists, neuropathologists and pathologists serving large hospitals attended by pediatric neurosurgeons. It is otherwise largely unknown. The tumor does occur in adults, albeit rarely however, with an unexplained predilection for the sellar/suprasellar region, a site where, thus far, all examples seem to have occurred woman. This age and gender favoritism is unexplained. As illustrated in the present case, such a rare, histologically undifferentiated lesion is almost certainly to be diagnostic problems, and clearly not one of the conventional regional lesions such as pituitary adenoma, craniopharyngioma, pituicytic tumor, meningioma, etc. A metastasis may be suspected.

AT/RTs assume many histological guises: 1) rhabdoid tissue with round cells with large eccentric pale nuclei with prominent nucleoli, 2) non-descript yet distinctive large pale, jumbled cells, 3) spindling populations large and dense enough to mimic medulloblastoma or supratentorial PNET. Mixed patterns are common. Most lesions are focally necrotic.

The lesion’s polyimmunophenotypic profile may come as a surprise since, aside from uncommon focal crude epithelial formations or neuroblastic rosettes, tumors are undifferentiated. Immunoreactivities, generally of individual cells or small clusters, variably for EMA, GFAP, cytokeratins, smooth muscle actin, and synatophysin. Most lesions are not positive for all markers, but many are reactive at least for EMA, cytokeratins, and GFAP. In this context, the diagnosis wants only confirmation by loss of staining (“loss of retention”) of tumor cell nuclei for INI1. Normally stained cells in the background, whether endothelial, fibroblast, or inflammatory, are valuable internal controls. Staining of tumor cell nuclei should be interpreted as lost only in the presence of these non-neoplastic cells whose immunopositivity confirms reactivity of the tissue and appropriate execution of the staining procedure.

Pituitary Blastoma

Only six cases, some of incompletely studied, record this rare lesion that is felt to represent neoplastic transformation of embryonic adenohypophysis. In accord with the early stage thereof, the normally early-appearing ACTH is the principal hormone produced, and is released in sufficient quantity to produce Cushing disease. Other clinical expressions relate to mass effect. All patients have presented in the first year of life. There is an evolving understanding that some cases are part of Dicer1 syndrome (pleuropulmonary blastoma, cystic nephroma, etc.).

The cellular lesions are remarkable for overall high cellularity and, most distinctly, epithelia, including goblet cells in some cases, that suggest differentiation to Rathke cleft. Small blastoma cells and larger, paler secretory cells fill out the spectrum. Necrosis and high mitotic activity have been present in some, but not all cases.

Immunohistochemically, secretory cells are positive for ACTH and synaptophysin whereas epithelia are more likely to react for cytokeratins and EMA. Overlap in staining between secretory and epithelial cells may be present. Positivity for annexin-1 has been used to support the presence of differentiation to folliculostellate cells.

Electron microscopy reveals cells with features suggesting folliculostellate cells.

Outcomes have varied.

Osteolipoma of the Tuber Cinereum
This rare lesion that present variably with visual disturbances, cranial nerve signs and endocrine abnormalities. It is pea- or bean-sized shell of calcification with a lipomatous center. One or more such closely apposed lesions may be present. By CT, they are spherical or cylindrical with a hyperdense bony shell encompassing hypodense adipose tissue.

There are no surprise histopathologically – only distinctive U-, C-, or J-shaped spicules and mature adipose tissue. Hematopoietic elements may be present.

Whether all reported cases are the same entity since the calcification in some are more solid and “lumpy” than alphabetical.

