HEAD & NECK
INTERESTING CASE #5

65-year-old female with a mass involving the maxillary sinus

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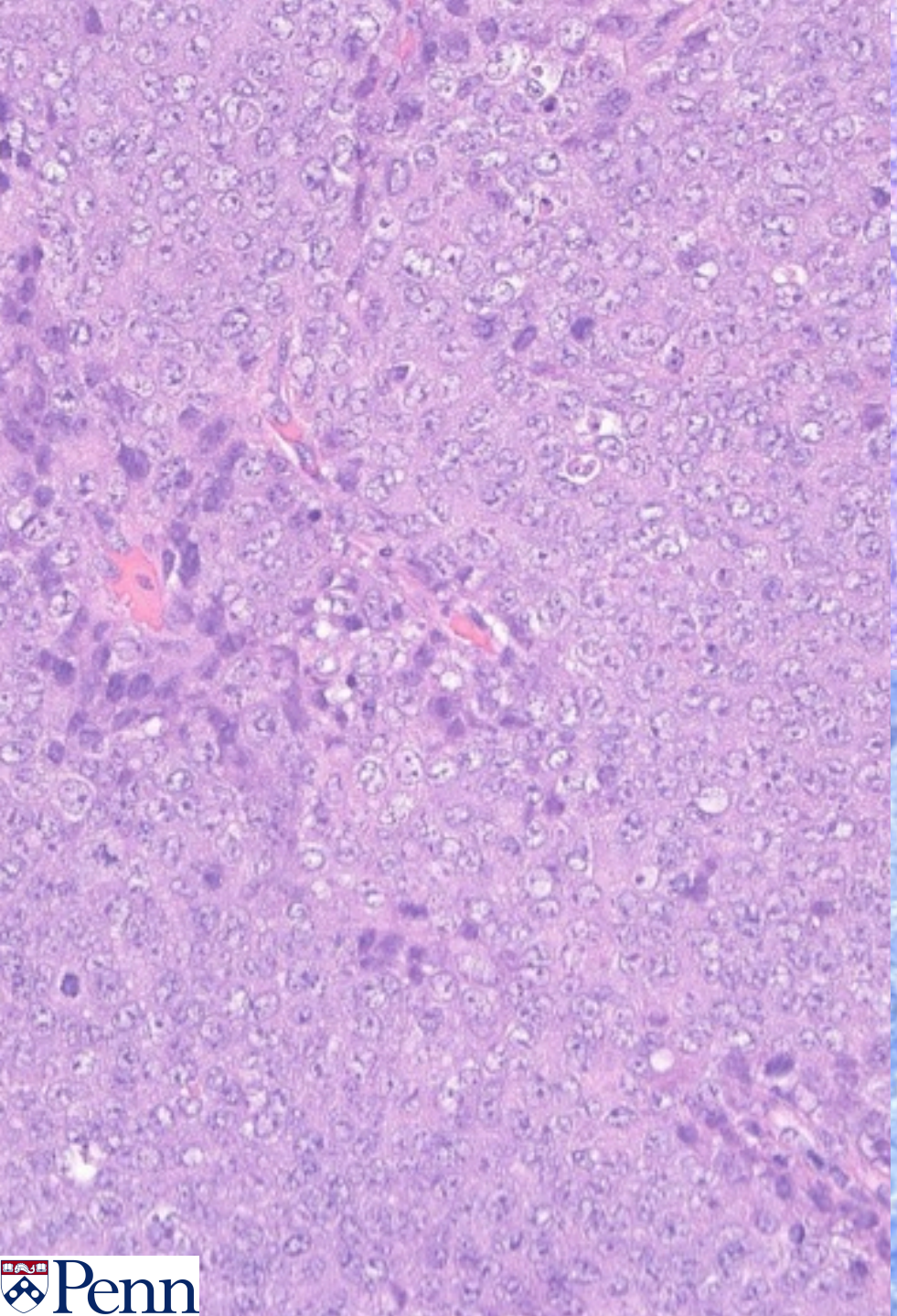
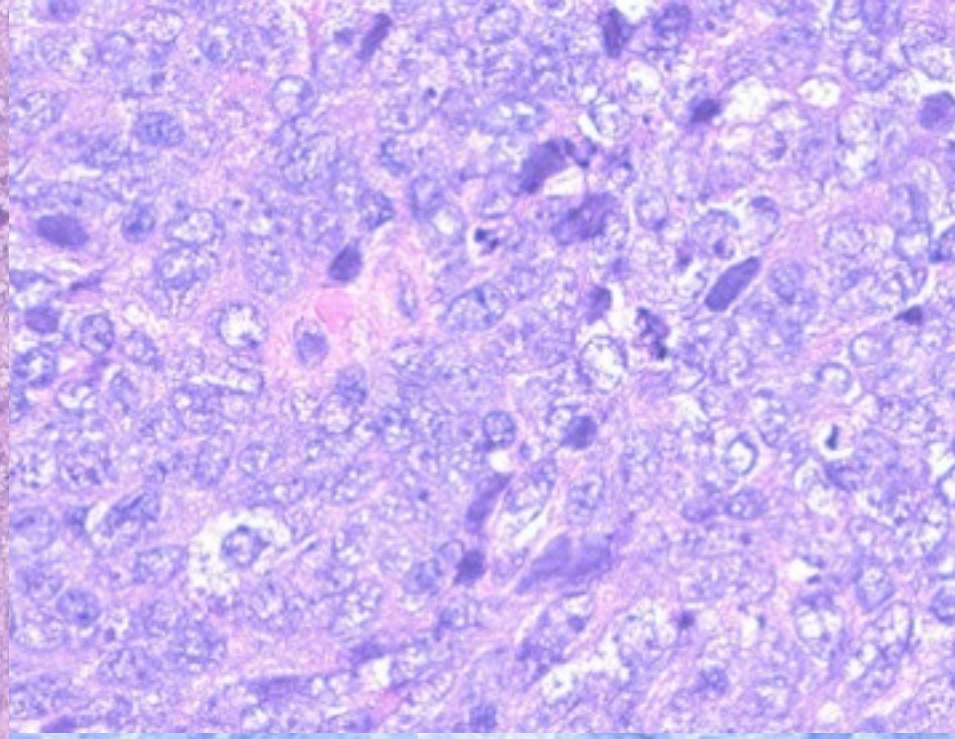
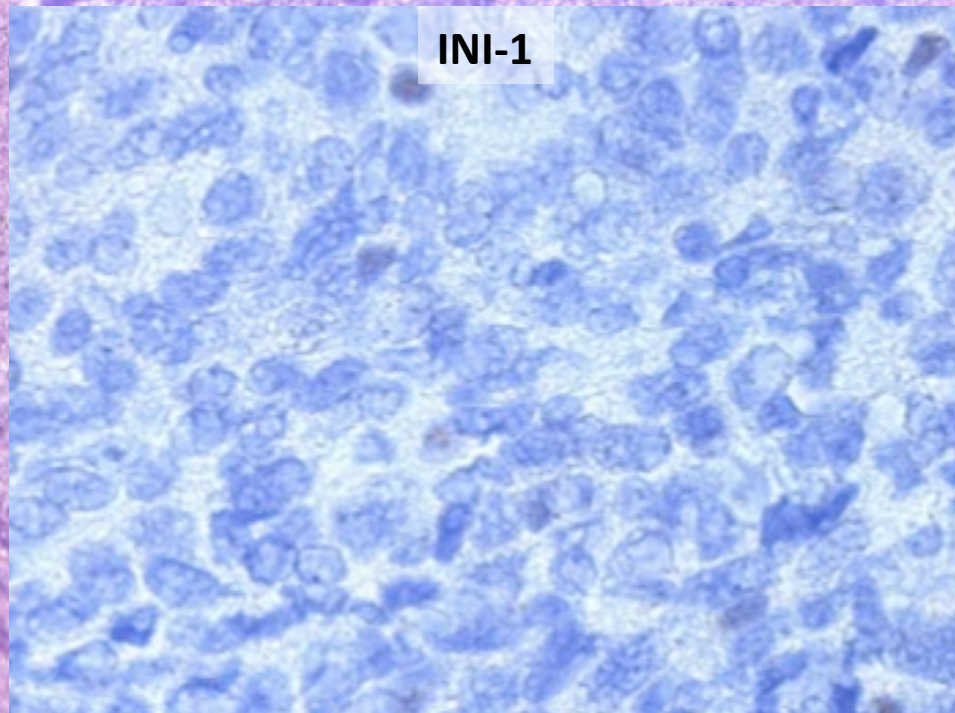
Prepared by Sharon Song, MD







Penn

SMARCB1 (INI-1) deficient sinonasal carcinoma

Clinical: Aggressive tumor with recurrences and regional/distant metastases; >65% mortality

Histology: Composed of basaloid cells with large, round nuclei, variably sized nucleoli and scant cytoplasm. Rhabdoid cells show abundant, eccentric eosinophilic cytoplasm (plasmacytoid). There is no in-situ component, but the tumor may spared in a pagetoid fashion into the overlying surface epithelium.

Ancillary studies: Loss of SMARCB1 (INI-1) by IHC; deletion can be confirmed by FISH. PAN-CK+; variable CK5+; focal or diffuse P63; focal CK7; variable, focal neuroendocrine markers; rare c-KIT; NUT-, EBV-, HPV-

Other SMARCB1 deficient tumors: Atypical teratoid/rhabdoid tumor of CNS, malignant rhabdoid tumors of the kidney and soft tissue, epithelioid sarcoma, renal medullary carcinoma, myoepithelial carcinoma of soft tissue, epithelioid malignant peripheral nerve sheath tumor, extraskeletal myxoid chondrosarcoma, SMARCCB1 deficient sinonasal tumors