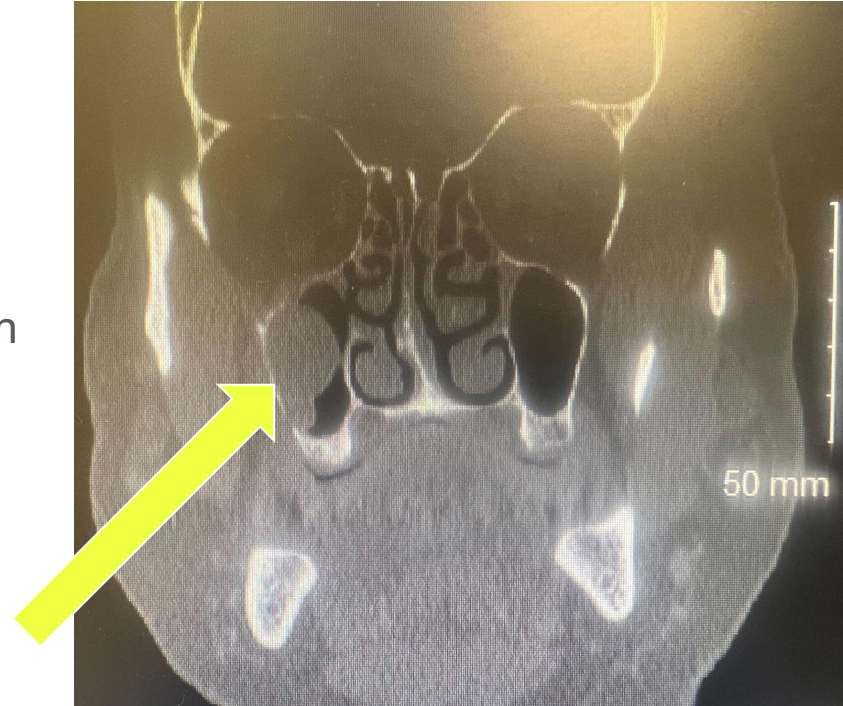


A Case of Adenoid Cystic Carcinoma

Julliette Lucas MS4

Patient Presentation

- DA is an 82-year-old male referred to Dr. Bruening from Green Bay for evaluation and resection of biopsy-confirmed right maxillary sinus adenoid cystic carcinoma (ACC)
- Dentures ill-fitting due to “lump” in roof of mouth, first noticed one year ago, had grown in size
- Denied changes in swallowing, voice, weight changes, hearing and vision changes
- Endorsed tenderness to palpation of right cheek
- PMHx: 45 pack year smoking history (quit 1997), Bilateral PE 2014, DVT, T2DM, OA, cataracts



Epidemiology

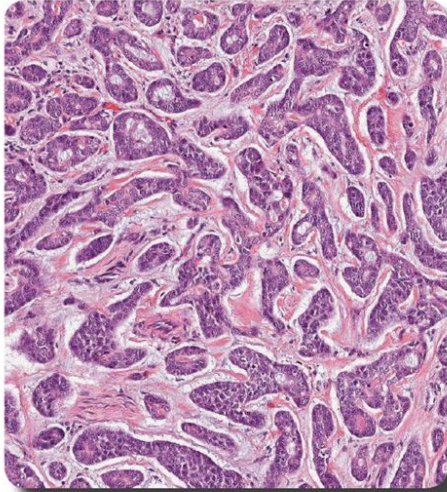
- Age is the strongest independent risk factor (mostly in 50s-60s)^[1]
- 60% of cases in females and 40% in males^[1]
- 4.5 cases per 100,000 (age adjusted)^[1]
- ~1% of head and neck cancers^[1]
- Also seen rarely in glands of reproductive tract, trachea, skin, and breast ^[1]

Etiology

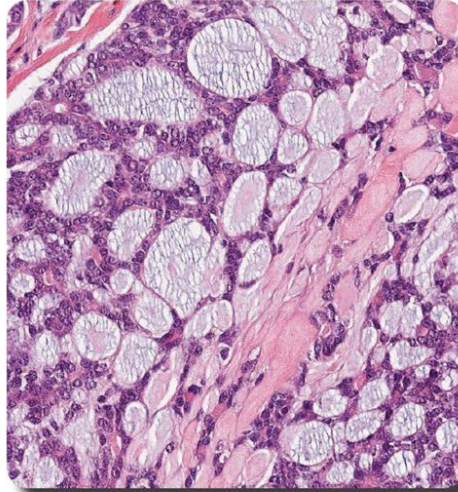
- No link has been found with tobacco, alcohol use, or HPV^[1]
- Associated with deletion of 1p35-36, translocation of 6q and 9p causing MYB:NFIB fusion gene → overexpression of MYB oncogene → tumorigenesis (>85% of tumors)^[2]
- p53, RAS, and PI3K (implicated in SCC) are rarely altered in ACC^[2]

Histopathology

TUBULAR (BEST PROGNOSIS, LOWEST GRADE) – 2 distinct cell populations arranged into ducts within a hyaline stroma. The luminal epithelial population are completely surrounded by myoepithelial cells

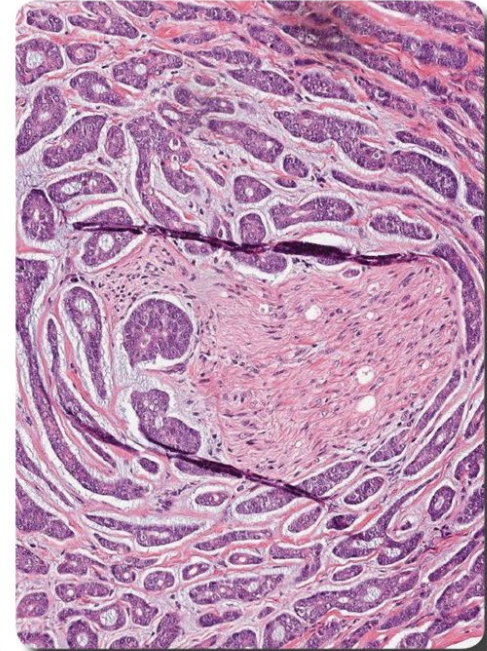


CRIBRIFORM (INTERMEDIATE PROGNOSIS AND GRADE) – this is the 'classic' or 'swiss cheese' pattern. The 'holes' are pseudocysts, lined by replicated basement membrane produced by the neoplastic cells

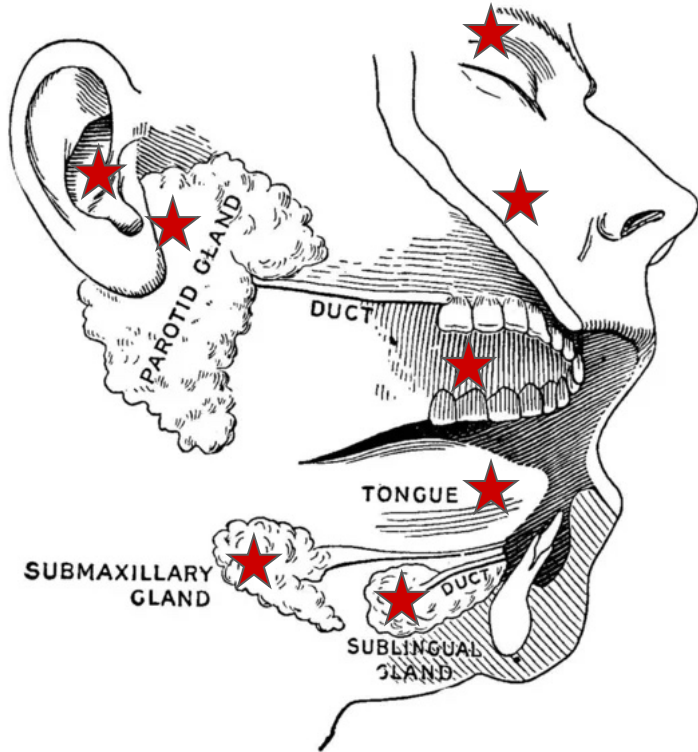


SOLID (WORST PROGNOSIS, HIGH GRADE) (NOT PICTURED) – if 30% of the tumor consists of non-cystic sheets or nests of neoplastic cells, the tumor is considered the solid variant

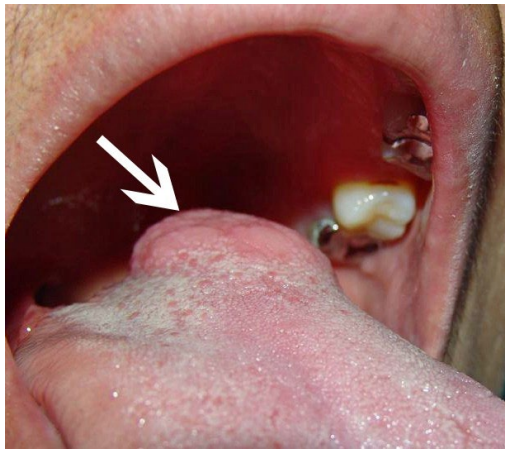
NEUROPHILIC CHARACTER - Peri and/or intra-neural invasion is a classic characteristic of AdCC



Primary Sites of ACC^[1]



- Secretory cells of the minor salivary glands (most common)
- Hard palate (patient described)
- Nasopharynx
- Lacrimal glands
- Tongue
- External auditory canal
- Rarely in glands of breast, reproductive tract, trachea, and skin



Evaluation

- Commonly presents as a hard, slow growing, painless swelling, site specific sx due to perineural invasion^[1]
- Flexible fiberoptic laryngoscopy^[1]
- CT and MRI to determine extent of primary tumor and for staging^[1]
- PET scans to evaluate for metastases^[7]
- Biopsy with immunohistochemistries for smooth muscle actin, S100, vimentin, and for MYB and CD117^[1]

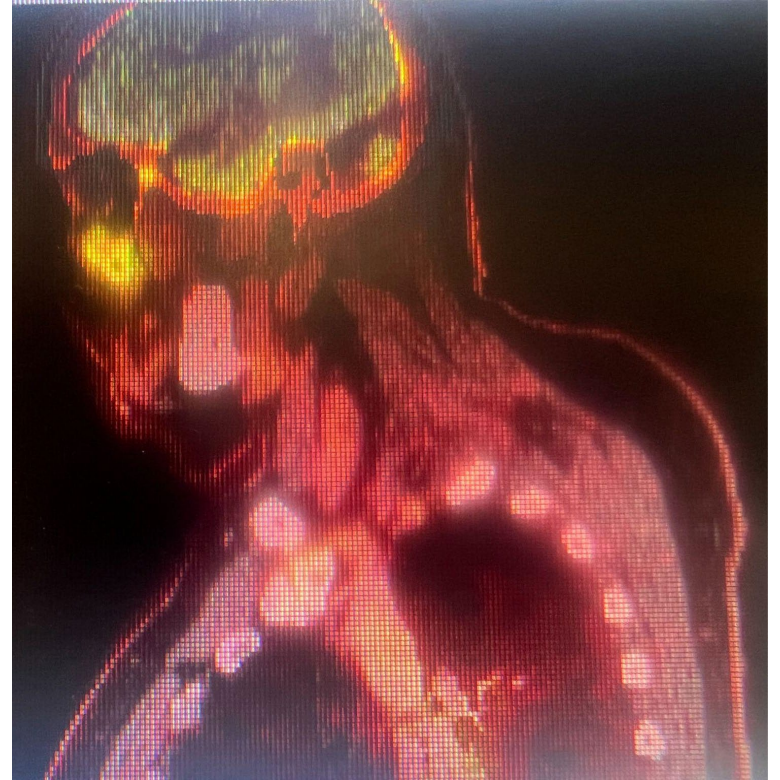
DA's Findings

- Primary lesion in right maxillary sinus 47mm diameter, with premaxillary, retroantral, and inferior extraconal orbital extension and probable extension into lower anterior nasal cavity at the inferior margin of the nasolacrimal duct and destruction of the alveolar ridge, anterior and posterolateral sinus walls, orbital floor, and zygoma
- Presumed right V2 retrograde perineural tumor spread at the level of the posterior orbital floor
- Enlarged abnormal LNs in levels I and II bilaterally (favor variant, reactive, or manifestation of systemic lymphoproliferative process over nodal mets on basis of symmetry, morphology, and distance from primary tumor)



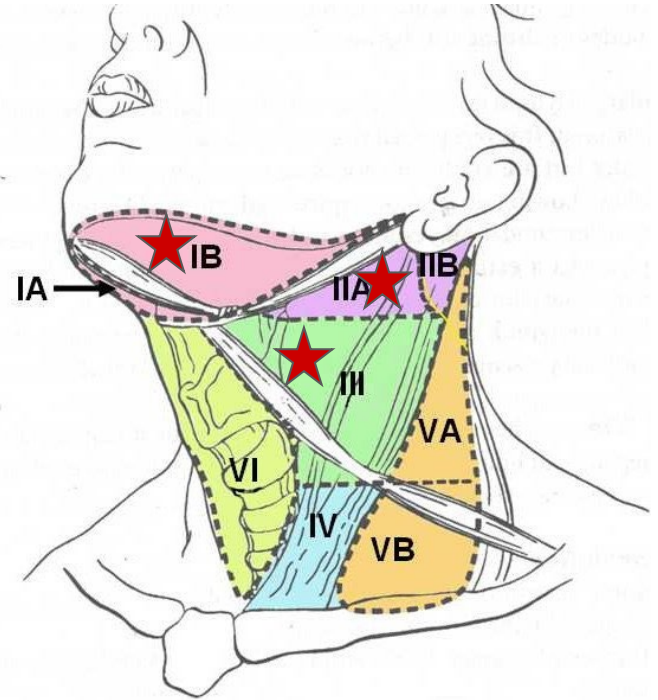
DA's Findings continued

- PET scan showed no evidence of distant mets, hypermetabolic right maxillary sinus mass, with likely degenerative TMJ hypermetabolism
- Bx displayed positive immunohistochemistry for CKAE1/3, CD117, S100, and CK7
- Showed cribriform and tubular pattern
- Flexible laryngoscopy showed no gross abnormalities throughout the sinuses, glottis and subglottis, and nasopharynx

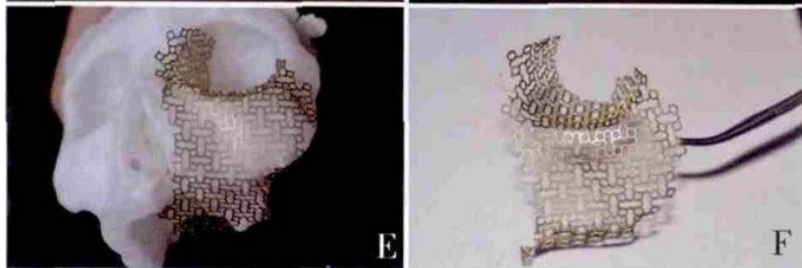
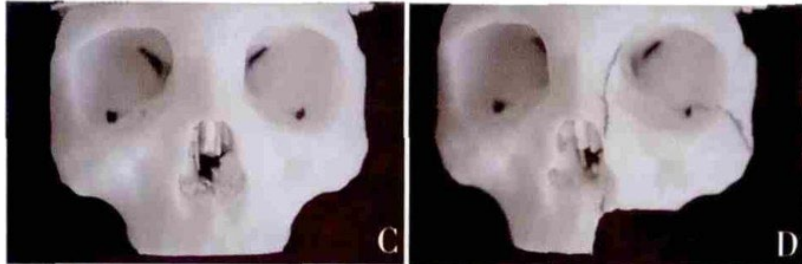
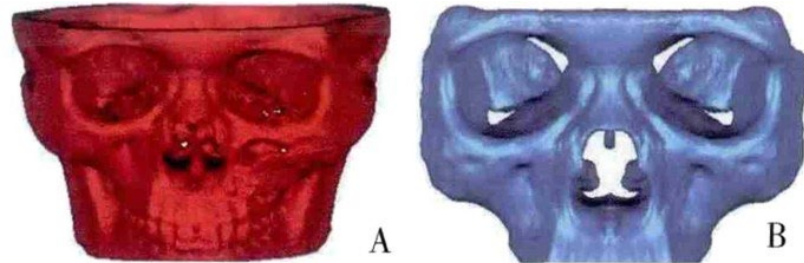


Treatment

- Complete resection of the with a goal of negative margins including:
 - Partial right maxillectomy
 - Right orbitotomy
 - Right orbital reconstruction with implant
 - Left free flap radial forearm to oral cavity
- Modified radical neck dissection
 - Levels Ib, Ila, III
- Initial margins all positive



Titanium Midface Implant^[4]



Recurrence

- >30% of ACCs eventually metastasize via hematogenous spread to lungs, bone, and liver^[1]
- 5-10 year recurrence rate with negative margins is 75%^[1]
- Intermediate or high grade cancer with positive margins? → post-operative radiation with ≥ 60 Gy^[1]
- Even with negative margins and post-operative radiation, recurrence in sites outside radiation field^[1]

Targeted Therapies and Prognosis

- ACC has not been responsive to traditional chemotherapy regimens^[1]
- Some promise with axitinib (a TKI) “With a median follow-up of 25.4 months, the 6-month PFS rate was 73.0% with axitinib and 23.0% with observation”^[3]
- 5-year survival 80%, 10-year survival 61%, 15-year survival 29%^[3]

Questions?

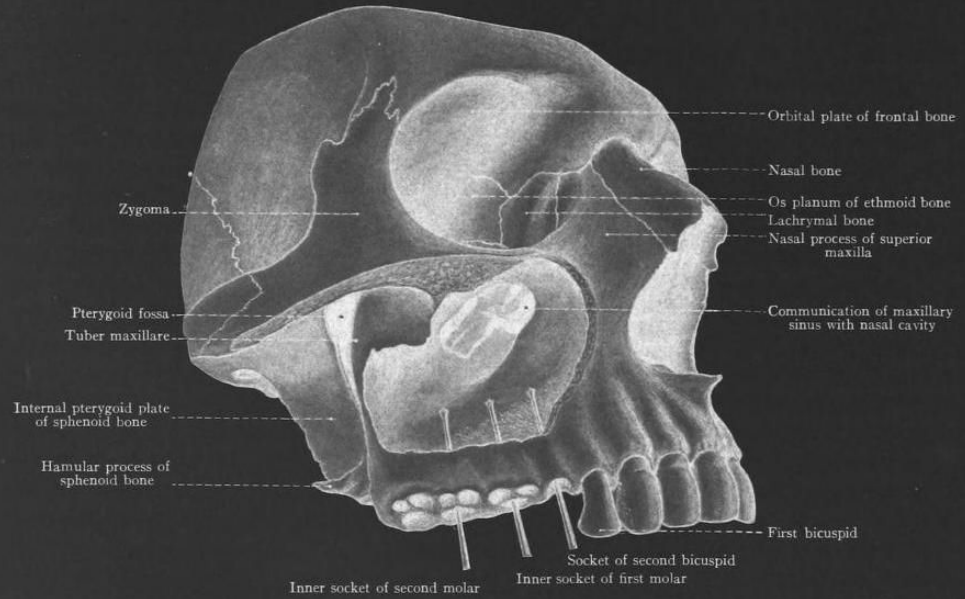


FIG. 18.—A portion of the skull in which the outer wall of the maxillary sinus has been removed in order to show its communication with the nasal cavity and the relations of the dental alveoli to the floor of the sinus. The alveoli have been perforated from below.

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FIG. 18.—A portion of the skull in which the outer wall of the maxillary sinus has been removed in order to show



References

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