


## Novel Case of Sinonasal Epithelioid Schwannoma with Benign Histopathologic Features



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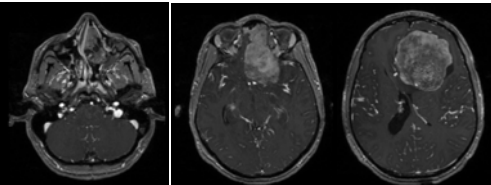
### Outline

- **Case Presentation**
- Schwannoma
- Epithelioid Schwannoma
- My Case
- Conclusions

### Mr. Ivent Smelt

<p><b>HPI:</b> 21 y/o male with HAS x 2-3 months Collapsed in hospital visiting friend Spontaneously recovered called mother Progressive decline in consciousness requiring intubation</p> <p><b>PMHx:</b> Type I DM Migraines</p> <p><b>PSHx:</b> Sinus surgery with post-op rhinorrhea</p> <p><b>All:</b> NKDA</p> <p><b>Meds:</b> Humalog Insulin Pump</p>	<p><b>FamHx:</b> N/C</p> <p><b>SocHx:</b> (+) tobacco Occ EtOH No IVDA</p> <p><b>PE:</b> Intubated/Sedated OE to voice PERRLA Face Symmetric FC x 4, Symmetric</p>
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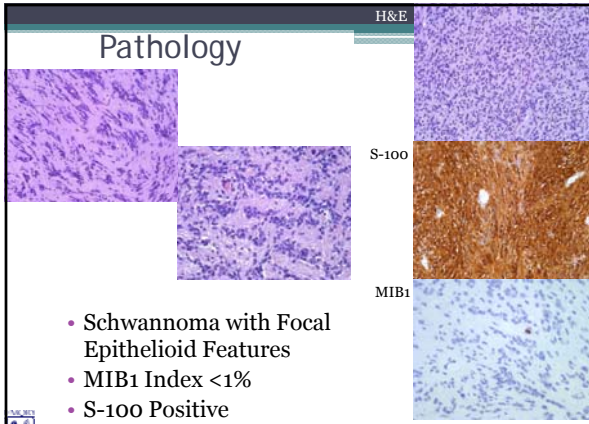
### Mr. Ivent Smelt (Con't)

**Hospital Course:** Allowed to awaken and extubate from presumed seizure with post-ictal confusion.

**Surgery:** HD# 4. Bi-frontal craniotomy with orbital bar removal for trans dural resection of tumor and EVD placement

**Post-op:** Uncomplicated. D/C'd home on POD6

### Pathology



- Schwannoma with Focal Epithelioid Features
- MIB1 Index <1%
- S-100 Positive

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## Peripheral Nerve Sheath Tumors

- Schwannoma
- Neurofibroma
- Traumatic Neuroma
- Perineuroma
- Malignant peripheral nerve sheath tumor
  - Epithelioid Schwannoma

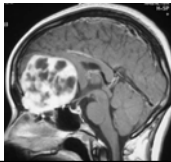
## Schwannoma : Peripheral Nerve Sheath Tumor

- Schwann Cells
  - neural crest cells
- Verocay in 1908
- 6-8% of intracranial tumors
- Age 40-50 unless with NF2
- Sensory Nerves of the PNS

- Location
  - Superior vestibular nerve
  - Trigeminal nerve
  - Vagus Nerve
  - Glossopharyngeal Nerve
  - Rarely intra-axial
    - From perivascular nerves
  - Anterior skull base?

## Sinonasal Tumors And Tumors Of The Anterior Skull Base

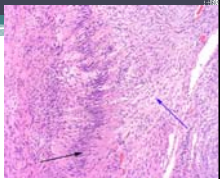
- DDx
  - Squamous cell carcinoma, adenocarcinoma, meningioma, esthesioneuroblastoma, sarcoma
  - Schwannoma?
- Schwann cells not present in optic nerve and olfactory bulb/tract
- Schwannoma
  - 34 cases documented
  - Etiology
    - Developmental
    - Non-developmental
    - Olfactory Ensheathing Cell Tumor

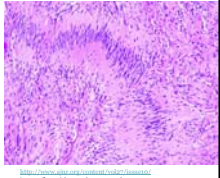


## Schwannoma

- Grossly:
  - Encapsulated
- Microscopically
  - Biphasic pattern
    - Antoni A
      - Verocay bodies
    - Antoni B
  - Frequently cystic, hemorrhagic or contain fat
- Histochemically
  - S100 positive
- Variants
  - Common,
  - Cellular,
  - ancient,
  - plexiform,
  - melanotic,
  - dermal nerve sheath myxoma,
  - granular-cell tumors,
  - Epithelioid

-Antoni A – Black Arrow  
-Antoni B – Blue Arrow








## Outline

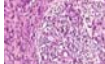
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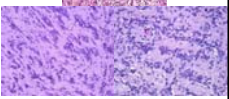
## Epithelioid Schwannoma : Malignant Peripheral Nerve Sheath Tumor

- McCormack et al. in 1954
  - Poorly differentiated spindle cell sarcoma
- 5%-17% of all MPNST
- Arise from benign nerve sheath tumor
  - Below the clavicles
  - Cranial involvement rare
    - 6 cases ranging from benign to malignant
- Aggressive
  - Local infiltration and metastasis
  - Superficial form less virulent
- Classically occurring in patients with type 1 neurofibromatosis (NF-1).

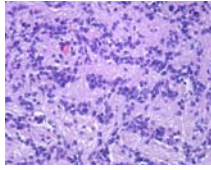






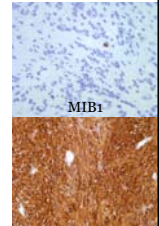
## Epithelioid Schwannoma

- Large epithelial
  - Contrast to spindle cells
- S-100 protein positive
  - 80%
- Lack melanoma-associated antigen and cytokeratin.
  - MPNST express S-100 protein in 50%
- Prognosis is poor with <20% 5-year-survival



## Benign Sinonasal Epithelioid Schwannoma

- Immunohistochemistry employed to determine malignancy
  - Malignancy in PNST
    - Proliferation index in excess of 10%
    - p53 immunoreactivity
  - Benign PNST
    - Proliferation index below 5%
    - Absence of p53 immunoreactivity
  - S-100 protein immunostaining



**This Is The First Case Documented Of A Benign Appearing Epithelioid Schwannoma Of The Anterior Skull Base**



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- Mr. Ivent Smelt

- GTR
- Long-term follow-up pending

- **Conclusion**

- Sinonasal/Anterior Skull Base Schwannomas are extremely rare
- First reported case of Epithelioid Schwannoma of the Anterior Skull base
- Epithelioid Schwannomas are classically malignant unless with low mitotic staining, absence of p53 reactivity and ubiquitous S-100 staining.

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