SCHWANNOMAS

Lixana Vega Vega, MD
Long Island College Hospital
Department of Surgery
59 y/o male presented w/ a palpable left neck mass.

PMH: DM, Hyperlipidemia, left acoustic neuroma 5x3mm and pituitary macroadenoma 17x27mm (2008)

Meds: Pioglitazone, Metformin, Glipizide, Lipitor, Gemfibrozil, Cabergoline (dopamine receptor agonist)

PSH- Laparoscopic cholecystectomy

Soc Hx: denies ETOH or tobacco use
CASE PRESENTATION

- **PE:**
  - Gen: NAD, no respiratory distress
  - Neck: lower left neck mass 8x4 cm, solid, elongated, right tracheal deviation.
  - Chest: equal BS, no heart murmurs
  - Neurological exam: no gross deficits

- **Labs:** unremarkable

- **Neck US**
  - Discrete heterogeneous mass - 4.7 X 2.8 X 2.5 cm

- **Neck CT scan**
  - Heterogeneous mass in left carotid sheath causing anterior displacement and compression of IJ vein
  - 7.4 x 2.9 cm extends from thyroid notch to below level of sternal notch. No lymphadenopathy.
CAROTID SHEATH
NECK EXPLORATION
PATHOLOGY

- Encapsulated, ovoid mass 7 x 3 x 1.5 cm
- Schwannoma
  - Benign peripheral nerve sheath tumor
  - Nerve fibers adherent to tumor capsule
POST OP COURSE

- Uneventful
- POD #1 - Tolerated PO diet.
- POD #2 - No voice changes.
  Discharged home.
SCHWANNOMAS

Neuron

Schwann cells

Myelin
**Neural Crest Neoplasms**

### Neuronal Cells
- Nerve ganglion cells
  - Ganglioneuroblastoma
  - Ganglioneuroma
- Neuroendocrine cells
  - Pheochromocytoma
  - Paraganglioma

### Neural Sheath = Schwann Cells
- Schwannoma
- Neurofibroma
Neurogenic tumor arising from the Schwann cells of the sheath of myelinated nerves.

- Neurilemoma
- Cranial, spinal, or peripheral nerves
  - Cranial nerve VIII- Acoustic neuroma or vestibular schwannomas
    - Annual incidence 1 in 100,000
    - Bilateral → diagnostic of Neurofibromatosis (NF) 2
- Head and neck → 25% to 45% of cases
- Most patients between 25 and 55 years of age
- No gender preference.
- Benign, slow-growing tumors.
- Solitary except when seen in association with neurofibromatosis (NF-2).
Inactivating mutations in the *NF2* gene (tumor suppressor gene) on chromosome 22.
- Schwannomas associated with NF2
- Sporadic schwannomas

Loss of expression of the *NF2* gene product: merlin or schwannomin.

Merlin restricts the cell-surface expression of growth factor receptors (EGFR)

In the absence of merlin, cells hyperproliferate in response to growth factors.
Malignant peripheral nerve sheath tumor (MPNST)
Aggressive spindle cell neoplasms - Sarcomas
Most commonly arising from nerve trunks
Can arise from
- Malignant transformation of benign lesions (rare)
- Degeneration of neurofibroma in NF-1
- Within the field of prior radiation therapy.
Constitutional symptoms and symptoms due to mass effect
Histology- Mitotic figures
The tumor advances locally, invading nearby structures.
Metastases
- Lung, liver, bone, skin
Heterogeneous mass.
- Carotid sheath
  - spread the carotid and jugular vessels apart
- Enhance after IV contrast on both CT and MRI.
- High signal on MRI
  - T2-weighted
  - T1-weighted with gadolinium
GROSS PATHOLOGY

- Well-delineated, globular, firm to rubbery yellow-tan mass.
- As opposed to neurofibromas, schwannomas typically have a capsule and push the parent nerve aside rather than invading it.
**Histology**

- **Antoni A**
  - palisading spindle cells
- **Antoni B**
  - loose myxoid hypocellular areas with scattered spindle cells
- **Immunohistochemistry**
  - S-100 protein positivity
  - Collagen IV positivity
  - Lack of neurofilament-positive entrapped axons
TREATMENT

- Surgery - Complete excision
- Sinonasal tumors
  - Endoscopic surgery
- Vestibular schwannoma
  - Surgical excision (tumors > 3cm)
  - Ablation - stereotactic radiation (Gamma Knife)
  - Serial monitoring with MRI
    - Minimal symptoms
    - Small tumors

- Tumors < 2 cm to > 4 cm.
- Radiosurgery technique - mean dose 14.5 Gy.
- Microsurgery included translabyrinthine, suboccipital, and middle fossa approaches.
- Median follow-up: radiosurgical group 48 months (3-84 months), microsurgical group, vs. 24 months (3-72 months).
Treatment of acoustic neuroma: stereotactic radiosurgery vs. microsurgery.


- **Tumor growth control** - no statistical difference between the two groups
  - 100% in the microsurgery group vs. 91% in the radiosurgery group (p > 0.05).

- **Radiosurgery** → superior in **measurable hearing preservation**
  - 57.5% vs. 14.4% (p = 0.01)
  - There was no difference in **objective hearing preservation**.

- **Microsurgery** → greater rate of **facial and trigeminal neuropathy** in the immediate postoperative period and at long-term follow-up.
  - Facial neuropathy 35% vs. 0%, p < 0.01 in the immediate postsurgical period and 35.3% vs. 6.1%, p = 0.008, at long-term follow-up.
  - Trigeminal neuropathy was significantly higher in the microsurgical group than in the radiosurgical group (17% vs. 0% in the immediate postoperative period, p < 001, and 22% vs. 12.2%, p = 0.009, at long-term follow-up).

- **Microsurgery** associated with a longer **hospital stay**.
  - (2-16 days vs. 1-2 days, p < 0.01)
REFERENCES


Townsend: Sabiston Textbook of Surgery, 18th ed.


THANK YOU!