CERVICAL SYMPATHETIC CHAIN SCHWANNOMA: A CASE REPORT

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INTRODUCTION
DDX: PARAPHARYNGEAL SPACE TUMOR

- Paragangliomas
- Salivary gland tumors
- Carotid body tumors
- Schwannomas
EPIDEMIOLOGY

• Parapharyngeal tumors: 0.5% of head and neck neoplasms.
  • Paragangliomas/salivary tumors: ~50%
  • Schwannomas: 12%

• Vagus nerve schwannomas are the most common form of schwannoma.

• Cervical sympathetic chain schwannoma: 60 cases reported in literature as of 2012

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<th>Table II. Tumoral types of the princi…</th>
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<td>Total</td>
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<tr>
<td>Benign</td>
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<tr>
<td>Trabecular adenoma</td>
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<td>Pleomorphic adenoma</td>
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<td>Basal cell adenoma</td>
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<td>Schwannoma</td>
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<tr>
<td>Neurofibroma</td>
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<td>Paraganglioma</td>
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<td>Lipoma</td>
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<td>Cyst</td>
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<td>Hemangioma</td>
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<td>Lymphangioma</td>
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<td>Chondroma</td>
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<td>Other</td>
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VAGUS NERVE SCHWANNOMA

• Superior and Inferior Laryngeal nerves are often affected

• Complications of Treatment
  • Hoarseness
  • Aspiration
  • Treat with Rehab or corrective surgery
CERVICAL SYMPATHETIC CHAIN SCHWANNOMA

• Tumor tends to grow into the parenchyma of the nerve

• Complications of treatment
  • Horner's syndrome
  • First bite syndrome
VALUE

• Raises awareness of a rare diagnosis

• Details difficulties with diagnostic imaging

• Provides an example of some unique post operative complications and their difficult managements

• Highlights the importance of informed consent
CASE PRESENTATION
HPI

• 36 year old caucasian female presents to an ENT clinic with dysphagia and a painful mass in her right carotid sheath
• Onset: 8 months earlier
• Patient describes slow growth

ROS

• Chronic fatigue
• Ear infection
• Tonsillitis
• Sinus infection
• Bronchitis
• Gall bladder disease
• Heartburn
• Dermatitis
• Headaches
• Anxiety
PAST MEDICAL HISTORY
• Anxiety
• Type 2 Diabetes Mellitus
• Gallstones
• Shingles

MEDICATIONS
• Citalopram 25 mg
• Metformin 1000mg
• NKDA
SURGICAL HISTORY
• Tonsillectomy and adenoidectomy
• Cholecystectomy
• Endoscopic sinus surgery
• Rotator cuff repair
• Tuboligation

FAMILY HISTORY
• Father: cancer, stroke, heart disease

SOCIAL HISTORY
• Never smoked
• Does not drink alcohol
PHYSICAL EXAM

• General Exam: Vitals WNL, no apparent distress
• Eyes: PERRLA, EOMI
• Ears: ear canals clear. Tympanic membranes intact
• Nose: nasal mucosa is noninflamed. Septum midline
• Oral cavity: no inflammation
• Pharynx/Larynx: indirect laryngoscopy reveals epiglottis, hypopharynx, true and false vocal cords appear normal with normal mobility
• Nasopharynx: indirect exam reveals no inflammation or abnormalities
• Neck: the neck reveals a large, firm, mobile neck mass on the right measuring approximately 4 cm. Carotid pulse palpable without bruit
• Cardio/respiratory/abdominal/extremities/neurologic exams benign.
DIAGNOSTIC WORKUP

• A CT scan was ordered and demonstrated an oval heterogeneous mass in the right carotid sheath measuring 35 x 24 x 28 mm.
DIAGNOSTIC WORKUP

• MRI w/wo contrast was then ordered which further demonstrated a homogeneous mass extending from C3 to C5.

• Radiology Interpretation: Likely a schwannoma localized to the vagus nerve.
ASSESSMENT AND PLAN

• Likely carotid sheath schwannoma of the vagus nerve

• recommendation: Surgical excision and nerve rehabilitation

• Right vocal cord paralysis leading to hoarseness, aspiration, and dysphagia were outcomes that were discussed with the patient in depth
SURGICAL PROCEDURE

- Dissection to the mass
- Carotid Artery and Jugular vein retracted
- Vagus nerve visualized and confirmed with nerve monitor
- Visualization of vagus, changes diagnosis
SURGICAL PROCEDURE

- Enucleation attempted by incising multiple layers of capsule

- the continuous nature of ganglion of the sympathetic chain with the tumor was confirmed.

- Enucleation with nerve was required

- Specimen sent to pathology which confirmed schwannoma showing spindle cells with Antoni A and Antoni B areas with Verocay bodies
POST-OP

• 1 week post op: complications
  • right sided ptosis
  • right sided facial neuralgia
  • visual disturbances: difficulty concentrating

• 2 months post op: minimal improvement
  • Neurologist: Facial neuralgia management
    • Topamax, Tramadol, gabapentin
  • Ophthalmologist: Ptosis repair
OUTCOME

• 2 years post-op:
  • No tumor recurrence
  • Ptosis was resolved with surgery
  • Facial neuralgia and visual disturbances persist despite medical management.
  • Patient unable to work due to permanent damage.
DISCUSSION
IMAGING

- Diagnostic imaging techniques include
  - CT and MRI: cornerstones
  - US
- Vagus nerve schwannoma: increased distance between ICA and IJV.
- CSCS: anteriorly displaced ICA and IJV without increased distance between.
- Variance can make pre operative diagnosis difficult
POST OPERATIVE COMPLICATIONS

• Horner’s syndrome
  • Very common, occurring in nearly every case of CSCS
  • Often resolves spontaneously but may require corrective surgery
• This case
  • Visual disturbances did not resolve

• First bite syndrome
  • Pain during first bite that resolves with subsequent mastication
  • Recurs with each meal
  • Often resolves spontaneously
  • Unknown pathophysiology
• This case
  • Facial neuralgia and headaches
  • No resolution
INFORMED CONSENT

• Defined as willing and voluntary acceptance of a medical intervention by a patient after adequate discussion about the …
  • nature of the intervention
  • Risks
  • Benefits
  • Alternative Treatment

• Crucial in cases involving nerves, will likely involve life altering side effects
• Fosters trust between patient and physician which benefits continuing management
CONCLUSION

• This case shows value as it serves as heightens awareness of a rare diagnosis, it’s diagnostic difficulties and it’s potential complications

• CSCS of the carotid sheath is a diagnosis that should not be overlooked in patients despite its rare occurrence

• Further study is recommended to determine the incidence of and potential avoidance of stated complications
QUESTIONS