Case 2 – Facial Reanimation
Douglas Henstrom, MD, FACS

69 year old male presents with a complaint of left facial paralysis.

Figure 1 – Facial paralysis. Patient presents with left facial paralysis.

HPI: State that you would:

- Obtain a detailed medical history beginning with a history of the present illness, and in the case of facial paralysis-establish a timeline of injury and any recovery to that point.

What additional historical information would you seek? State that you would ask:

- How did the facial paralysis occur?
- How long has he had the facial paralysis?
- Have there been any changes in the degree of facial paralysis since it started (improved or worsened?)
- Has he done anything to treat the consequences of this paralysis?
- What is he currently doing to protect his eye?
- What is bothering him most about the paralysis?
- Any associated otologic symptoms such as hearing loss, tinnitus, otorrhea or vertigo?

He suffered a tractor accident while working on his farm approximately 3 years ago. He was working on the tractor when a large piston fired and hit him in the side of the head. He was hospitalized for 6 days. It was noted at the time that he had complete facial paralysis on the left side. He has had essentially no change in the intervening 3 years and not sought surgical correction. He continues to use drops and ointment in the eye during the day and covers it at night with a moisture chamber. He is most bothered by his lack of smile and the constant care for his eye. He has no hearing loss or tinnitus.

PMH: *Otherwise healthy*
Allergies: *NKDA*
Medications: *Daily vitamins*
FH: *Non-contributory*

SH: *Non-smoker, occasional social alcohol (1-2 drinks/week), back working on farm*
ROS: *Negative*

What would you look for on physical exam? State that you would:

- Perform a full head and neck examination with special attention to all cranial nerves.

PE:

Vitals: *Normal.*
GA: *Normal in appearance other than HB VI/VI left sided facial paralysis. EAC and TM's normal and intact with clear ME spaces, Weber is midline, Rinne AC > BC, SRT 10/10.*
Eyes: *PERRLA, Left lower lid ectropion of 4mm, lagophthalmos of left upper lid. with Bell's phenomenon, and left brow ptosis without movement. OC/OP/NP-normal without mass or lesion, normal occlusion, normal tongue movement.*
Nasal: *Collapse of left external nasal valve; septum straight and midline*
Neck: *Soft, no masses or lymphadenopathy*
Neuro: *CN's 2-12 intact bilaterally, other than FN exam*
Left facial nerve exam: flaccid paralysis.
Brow is ptotic with no movement Incomplete eye closure with both gentle and forceful effort. Nasolabial fold is effaced. Oral commissure is inferiorly displaced and with attempts at smile there is no movement. Lower lip is weak with respect to the other side. No signs of synkinesis

What is your differential diagnosis?

- Blunt head trauma resulting in likely temporal bone fracture.
- Other causes-neurologic, infectious, neoplastic, toxic, metabolic, iatrogenic.

Next steps in management: What tests would you order?

- Audiologic evaluation if patient also has complaints of hearing loss.
- Electrodiagnostic studies are most useful in the acute period (this injury occurred 3 years ago).
- Evaluate the cause and possible topographical location of facial nerve injury in order to understand both prognosis of any recovery, as well as optimal choice for reanimation.
- To evaluate the extent of injury to the facial nerve, and the functionality of the facial muscles.
- Imaging:
  - In this patient, review of the CT temporal bone that was obtained after his injury (3 years ago) can be used to confirm the diagnosis.
  - In general when patients present with facial paralysis one can consider CT Scan in cases of suspected intratemporal pathologies (cholesteatoma, facial nerve hemangioma or schwannoma, fracture). A MRI with gadolinium detects inflammatory causes (Bell's palsy, Ramsey Hunt) and can differentiate those from other CPA or direct facial nerve masses. It can also map the full course of the extratemporal facial nerve.

What are the most commonly used electrodiagnostic tests, and what do they show?

- ENOG:
  - Stimulates facial nerve transcutaneously at the stylomastoid foramen.
  - Not useful until at least 3 days after injury to allow for complete Wallerian degeneration.
  - Provides objective comparison of latencies and compound action potential between muscles on paralyzed and normal side, with a supramaximal stimulation.
  - Most useful to stratify patients into observation vs. surgical decompression following nerve injury (most specifically intratemporal pathologies). A degeneration of 90% or more is typical threshold for consideration of facial nerve decompression surgery.
- EMG:
  - Needle electrodes are inserted into facial muscles (commonly orbicularis oris and orbicularis oculi) and the muscle response/activity to voluntary contraction is measured.
  - Will determine the existence of functional motor units.
  - Normal resting muscle exhibits no spontaneous electrical activity, however presence of voluntary action potentials indicates at least some continuity of nerve.
  - Most commonly used in conjunction with ENOG to verify degree of nerve damage
  - When denervated, a muscle will show spontaneous fibrillation potentials and positive sharp waves. May take 2-3 weeks for fibrillation potentials to show on EMG.
  - Only test that can follow the degree and pace of facial nerve recovery.
  - Polyphasic action potentials indicate recovery/reinnervation.
- Nerve Excitability Test:
  - Not useful during first 3 days following nerve injury.
  - Performed with Hilger nerve stimulator.
  - Measures the difference in amperage (objective measure) required to produce a barely visible twitch on paretic side compared to normal side (subjective comparison).
  - A difference greater than 3.5mA is indicative of poorer prognosis.
- Maximum Stimulation Test:
  - Not useful during first 3 days following nerve injury.
  - Performed with Hilger nerve stimulator.
  - Provides sufficient electricity to depolarize all axons and greatest amplitude of facial movement is observed.
  - Facial muscles on paralyzed side are subjectively described in comparison to healthy side as: equal, slightly decreased, markedly decreased, or absent when compared with healthy side.
  - Generally only done on patients with HB VI/VI paralysis because test is uncomfortable.

Test results:

- Given the history of facial nerve paralysis after head trauma further imaging at 3 years for evaluation is not necessary. However, the temporal bone imaging that was obtained after injury (3 years ago) should be evaluated.
- The temporal bone CT shows a transverse temporal bone fracture affecting the labyrinthine segment of the facial nerve (Figure 2).

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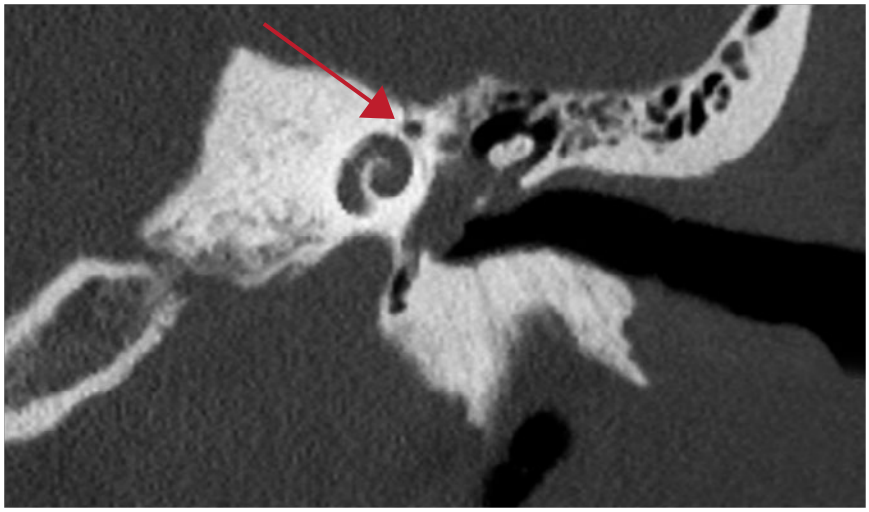
Figure 2 – Coronal CT temporal bone shows fracture involving the labyrinthine segment (red arrow) of the facial nerve.



Diagnosis:

- The patient had suffered an immediate complete facial nerve paralysis from a temporal bone fracture as a result of his farming accident. CT demonstrated fracture at the region of the labyrinthine segment.
  - Because this occurred approximately 3 years ago, there is no role for decompression of the nerve, and the facial muscles will have atrophied and would not be useful for reinnervation.
  - He will likely receive the most benefit from proper management of the periocular complex (eyelid weight and lower lid tightening) and an attempt to reanimate the mouth with a temporalis muscle or tendon transfer. He would be a possible candidate for free tissue transfer using the gracilis muscle, but due to his age and personal choices, he elects for a simpler operation.

Management: What are treatment options for nerve reinnervation?

- Primary Neurorrhaphy:
  - Optimal timing is immediately following injury/disruption of nerve.
  - Provides the best chance of recovery (meaningful facial movements) and least amount of side effects (synkinesis).
    - Generally performed via epineural repair anywhere nerve has been disrupted.
- Interposition grafting:
  - Provides good resting tone and some spontaneous facial movement.
  - Used to provide a tension-free end-to-end connection between disrupted nerve sections.
  - Use a cable graft from donor nerve (great auricular and sural nerves most common) between nerve endings.
- Cross-face grafting:
  - Typically preserved for staged free muscle transposition.
  - Can be utilized to re-innervate specific segmental nerve loss. i.e.- buccal/zygomatic branches on the non-paralyzed side are cut and connected to the corresponding branches on the paralyzed side.
  - Sural nerve provides the best, longest donor nerve for length required.
- Other nerve transfers:
  - Includes direct XII-VII, XII-VII utilizing a jump graft, and increasingly more common V-VII nerve coaptation.

  - Used when proximal VII nerve is unavailable but the distal nerve is anatomically intact and viable.
  - Advantages: Relatively straight forward surgery, quicker return to some function (3-6 months), mimetic motion can closely resemble normal action with practice, very little if any loss of normal functionality from donor nerve.
  - Disadvantages: Possible synkinesis, potential donor nerve compromised function.
  - Must be performed before atrophy and fibrosis of native facial musculature (1 year).

What are secondary treatment options for facial (smile) reanimation?

- Static Procedures:
  - May be used in isolation or as an adjunct to other reanimation procedures to bring more facial symmetry by providing static support.
  - Materials commonly used: fascia lata, Alloderm®, and Gore-Tex®.
- Dynamic Procedures: Indicated either when the distal nerve is unavailable or facial musculature is atrophic and cannot be reinnervated.
- Regional Muscle Advancement:
  - Temporalis (and to a lesser extent Masseter) muscle are utilized to reanimate the smile function.
  - Major goals are to create some facial movement, improve oral competence and symmetry at rest.
  - Drawbacks: A bulge over the zygoma, temporal hollowing, lack of meaningful movement created, asymmetry of the mouth.
  - Recent advancements have shown the Temporalis Tendon Transfer technique to be very useful and avoids some of the complications of other techniques.
- Microvascular Muscle Transfer:
  - Most commonly the gracilis muscle (other muscles include pectoralis minor, latissimus dorsi).
  - Goals: Restore normal resting tone and symmetry, improve amount of movement with smile, and improve oral competence.
  - May be done as a 1 or 2 stage procedure. Most commonly a 1 stage procedure connects the transferred muscle to a local nerve-Masseter nerve. In a 2 stage procedure the muscle is connected to a previously placed cross face nerve graft, which is connected to a branch of the contralateral facial nerve.

What are other useful adjunctive treatment options for other areas of the face affected by the paralysis?

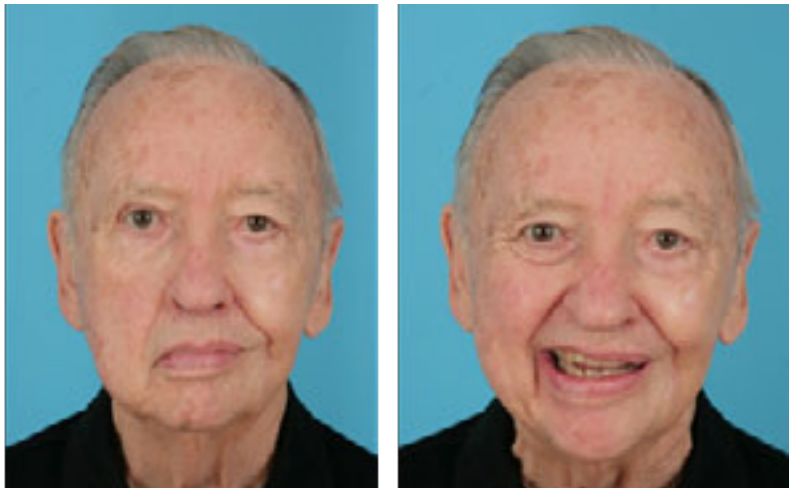
- Browlift
- Facelift
- Functional Rhinoplasty (or static sling to nasal base) to correct nasal valve collapse
- Highly Selective Neurectomy
- Selective myomectomy
- Botox
- Platysmectomy for synkinesis

Surgical techniques:

- Upper Eyelid Loading (Platinum or Gold weight):
  - After marking supratarsal crease, inject a small amount of local anesthesia to the fold.
  - Incise skin-centering incision over medial limbus of iris in neutral position.
  - Elevate skin and muscle (Orbicularis oculi) over the tarsal plate creating a pocket big enough for implant and down to lash line.

  - Place implant in pocket and fixate to tarsal plate with 6-0 permanent suture through all holes on appropriately sized weight.
  - Close muscle and skin.
- Lower Lid (Lateral Tarsal Strip):
  - After marking line for canthotomy-inject a small amount of local anesthesia to the area-including lateral aspect of inferior and superior lids.
  - Perform a lateral canthotomy and inferior cantholysis of lateral canthal tendon.
  - Separate anterior lamellae (skin and muscle) from posterior lamellae (tarsus and conjunctiva).
  - Denude overlying conjunctiva from posterior lamella and trim excess lateral canthal tendon.
  - Suspend lateral canthal tendon to medial aspect of orbital rim periosteum in appropriate position.
- Orthodromic Temporalis Tendon Transfer:
  - Preauricular incision is made. Variations in length are described from a small 6cm incision to a regular parotid incision. Incision is made long enough to allow for complete facial flap elevation and easy access to the coronoid process of the mandible.
  - Branches of facial nerve distal to the anterior parotid gland are identified and protected.
  - Masseter muscle is divided so as to expose the coronoid process, which is where the temporalis tendon inserts.
  - Using right angle retractors and a clamp on the coronoid, a coronoidectomy is performed at the neck using a reciprocating saw.
  - The coronoid is then freed from the surrounding tissue and the bone is removed from the tendon, preserving as much tendon as possible.
  - The tendon is then put on stretch. If extra length is needed to get the tendon to the modiolus of the orbicularis oris muscle, finger dissection on the deep surface of the temporalis, and inferior pull on the tendon will generally suffice.
  - If there is further need for length/stretch modifications for optimizing the movement at the commissure, a fascia lata graft may be harvested from the leg and fixated to the tendon and advanced to the modiolus for a proper length/stretch and subsequent movement.
  - Either the tendon or the fascial extension is advanced to the modiolus and/or nasolabial crease area and fixated with permanent sutures.
  - Pre-operative determination of an appropriate traction point and traction vector is very important for a natural smile.
  - Anchoring sutures placed too superficially in the muscle will evert the lip, and if placed too deeply will lead to lip puckering and inversion.
  - Slight overcorrection of tightness is adequate, showing some of the lateral incisor tooth, however not as much overcorrection is needed compared to the historic temporalis muscle transfer as there will not be as much relaxation of the tendon post-operatively.
  - When completed, the masseter muscle edges are re-approximated with Vicryl sutures.
  - A JP drain in placed in the wound bed after hemostasis is achieved and the wound has been irrigated.
  - A mild compressive dressing is placed after skin closure (Figure 3).
  - There are many modifications of this procedure, including a transoral transection of the coronoid process and advancement and fixation to the modiolus/nasolabial fold through an incision in that area. While approaches may differ, the goal of fixating the temporalis tendon in the correct area, with the correct direction and tension of pull is the same.

Figure 3 (A, B) – Facial paralysis. Patient after platinum weight, lateral tarsal strip and left orthodromic temporalis tendon transfer. (A) at rest (B) smiling.



A. **B.**

What are the potential complications/pitfalls?

- Eyelid Loading:
  - Extrusion (rates lower with platinum weight).
  - May not close fully when patient supine.
  - Visible bump on upper eyelid from weight profile.
- Lower lid (lateral tarsal strip):
  - Lid Malposition.
  - Entropion.
- Temporalis Muscle Transfer:
  - Muscle bulge over the zygoma.
  - Temporal hollowing from muscle excision.
  - Over or under correction of oral commissure.
  - Worsening of oral competence.
- Temporalis Tendon Transfer:
  - Over or undercorrection of the oral commissure.
  - Worsening of oral competence.

Follow-up:

- 1 week post-operative follow-up to ensure proper healing and eye function/protection.
- Periodic follow-up to ensure completion of healing and the desired result is achieved.
- Ensure ongoing proper eye care, including continuing eye drops and/or lubrication if needed.
- Long term follow-up to ensure ongoing eye health and re-evaluation of any re-animation procedure to ensure ongoing benefit and any need for possible revision surgery.

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Case 5 – Child with Respiratory Distress and Hoarseness

Amal Isaiah, MD, DPhil

You receive a call from the ER to evaluate a 7 year old child with increased work of breathing and a hoarse voice after being struck by a bottle rocket firework in the neck.

HPI: State that you would:

- Upon arrival in the unit, you ask for:
  - Onset: Immediately following trauma to the neck (see 1).
  - Duration: No relief since the incident.
  - Progression: Progressively worse over time, and now has biphasic stridor.
  - Severity: He is in mild to moderate distress with a very hoarse voice. He intermittently complains of neck pain and trouble breathing, but does not have cyanosis or drooling. He has had some improvement in distress since being placed on 4L of oxygen. His pulse oximetry reads 99% on the supplemental oxygen as described.

What additional historical information would you seek?

- History of intubation, other surgical history.
- History of voice or breathing problems prior to the current incident.

A description of examiner's statement summarizing the child's condition:

- *A quick physical examination revealed a child in mild to moderate distress with tachypnea, dysphonia, alar flaring, subcostal and intercostal retractions, as well as biphasic stridor. Examination of the neck revealed crepitus over the midline and lateral neck. A focal abrasion was seen over the thyroid cartilage. Pulse oximetry revealed 99% on 4L nasal cannula.*

What is your differential diagnosis?

- Laryngotracheal trauma with possible fracture of the laryngotracheal cartilaginous complex
- Post-traumatic hematoma
- Aerodigestive tract foreign bodies
- Bilateral vocal cord paralysis
- Esophageal injury and perforation
- Laryngocele

Brief discussion:

- Blunt laryngeal trauma is a rare but serious cause of airway injury. Common presenting symptoms include dyspnea, dysphagia, crepitus, anterior neck pain or ecchymosis, and dysphonia. A high clinical suspicion is needed to avoid the consequences of a missed airway injury. In a stable child, initial workup includes flexible laryngoscopy, CT imaging and possibly a direct laryngoscopy and bronchoscopy (DLB) under general anesthesia.
- Children with mild mucosal injuries without airway compromise or cartilage fractures can be managed conservatively with humidified air, a proton pump inhibitor, steroids and possible prophylactic antibiotics. Children are generally observed for at least 24 hours. Initial management of the child with an unstable airway is controversial. The first step is establishing a safe airway. Some advocate for tracheostomy rather than endotracheal intubation.

- Tracheostomy may prevent additional trauma and avoids having to perform an emergent tracheostomy for a failed endotracheal intubation. However, good outcomes can also be achieved with endotracheal intubation under direct visualization.
- After establishing an airway, an immediate or a delayed repair can be considered. Data is limited on this subject in children. In a review of 112 adults with external laryngeal trauma, immediate surgical repair improved airway and voice outcomes compared to a delayed repair. In contrast, delaying surgery may allow for a reduction in edema and more time for surgical planning and stabilization of a precarious patient.
- The optimal management of vocal cord paralysis following blunt laryngeal trauma remains controversial. Up to 60% of children with post-traumatic vocal cord paralysis will regain movement without intervention. The mean time to resolution ranges from 4-9 months with some children showing spontaneous resolution up to 3 years after the original injury. Therefore, in a child with a unilateral vocal fold paralysis, observation for 1-3 years prior to intervention is prudent.

Next steps in management:

Appropriate steps:

- Stabilize the airway: This should be the first step. Use upright positioning, suctioning and passive oxygen flow to relieve distress at least temporarily.
- Flexible endoscopic evaluation of the airway: Bedside endoscopy should be considered in instances where the child is stable.
- If the risk of immediate airway obstruction is high: Consider endotracheal intubation in the emergency room. If endotracheal intubation is unsuccessful consider a tracheostomy.
- If the risk of immediate airway obstruction is low: The next step would be to secure the airway in the OR using rigid instrumentation. Request OR staff to provide endoscopic instruments and bronchoscopes. It is imperative to avoid bag mask ventilation as much as possible due to the risk of worsening subcutaneous emphysema.
- Intubation following endoscopic evaluation of the airway. Obtain photodocumentation.
- Obtain a CT scan following endotracheal intubation, with specific attention to the cartilaginous laryngotracheal framework.
- Based on information obtained from direct visualization and a CT scan, open repair with or without tracheostomy should be considered. Classification of laryngotracheal injuries is best described by a standardized grading system. *Schaefer and Brown* classified laryngotracheal injuries using a combination of clinical, imaging and endoscopic patterns. Class I typically manifests with minimal airway compromise. Significant edema and minor mucosal lacerations are features of Class II injuries, with imaging demonstrating a non-displaced fracture. Class III injuries are characterized by massive edema with exposed cartilage and vocal cord immobility. Addition of more than two fracture lines converts class III to IV. Finally, laryngotracheal separation is the central feature of class V injuries.

Inappropriate steps:

- Investigations such as bedside endoscopy as well as imaging such as CT scan are inappropriate where the immediate need is to address an airway emergency. However, both of these procedures may be useful if the child is stable, and where there is no imminent risk of airway obstruction.

Following endotracheal intubation in the OR, antibiotics were started and a high-resolution CT of the neck was performed.The scan showed pneumomediastinum and a possible anterior thyroid cartilage fracture.The child was taken back to the OR and the fracture repaired with 4-0 PDS sutures and a drain placed. He was extubated on day 5 following demonstration of a cuff leak.

Consultations:

- None.

Postoperative management:

- Admission to the intensive care unit and initiation of feeds by nasogastric tube.
- Administration of steroids to manage airway edema.
- Drain removal (postoperative day 1).
- Initiate and continue broad-spectrum antibiotics for 48-72 hours and carefully monitor for development of mediastinitis.
- Serial examination of the neck to ensure resolution of subcutaneous emphysema.
- Consider extubation once crepitus has resolved and there are no signs of mediastinitis.
- Once extubated, recommend voice rest, humidification and taper steroids.
- Perform bedside endoscopy to assess vocal cord mobility.

Complications and management:

- Acute:
  - Mediastinitis (ICU admission, antibiotics, washout as mandated by general surgery).
  - Laryngotracheal separation (repair under ECMO).
  - Dehiscence of repair site (revision).
- Chronic:
  - Subglottic, glottic or tracheal stenosis (observation for mild cases, dilation and laryngotracheoplasty with tracheostomy for moderate or severe cases).
  - Hoarseness due to vocal cord immobility (unilateral: serial examination, augmentation versus medialization; bilateral: tracheostomy versus cordectomy).

Follow-up plan:

- Absence of fevers, normal swallow established, no airway symptoms.
- Follow-up usually at 1-2 weeks and subsequently depending on the procedures performed (e.g. tracheostomy) and associated injuries (e.g. vocal cord immobility).

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Case 1 – Nasopharynx Cancer with Staging

Jeffrey C. Liu, MD, FACS

A 50 year old male of Chinese descent presents with a 6 month history of neck swelling and nose bleeds.

HPI: State that you would:

- Obtain a detailed medical history beginning with a history of the present illness.

The patient reports that about 6 months ago he noticed swellings in the left and right sides of the neck. They did not bother him very much. About 3 months ago he started having intermittent bleeding from both nostrils. It happens most often when he blows his nose hard. There has been no sustained bleeding, and he has not had to seek care previously. He is seeking medical care because the swellings seem more significant.

He incidentally notes mild hearing loss, more on the right, over the last 1-2 months. It is not significant but he preferentially uses the left ear for the telephone. No tinnitus, no dizziness. No otalgia.

He has lost about a 10 lbs. of weight over this time.

PMH:	*Hypertension*
Medications:	*Lasix for hypertension*
Allergies:	*NKDA*
Family/ Social history:	*He has been a long time smoker, with about 1.5 packs per day for 45 years. He is an occasional social drinker. His family is originally from southeast China, where he grew up. He moved to the United States about 25 years ago.*
ROS:	*No other findings on review of systems.*

What would you look for on physical exam? State that you would:

- Perform a thorough head and neck exam including vital signs.

PE:

Vitals:	*Temperature: 37.0 C, Pulse: 80, RR: 18, BP: 137/85*
GA:	*Thin Chinese male in no apparent distress.*
Ear:	*Right tympanic membrane shows a small effusion with no signs of infection; left ear is normal.*
Nasal:	*Exam shows normal anterior structures. No blood is seen.*
Oral exam:	*Shows normal dentition and oral findings.*
Neck:	*Exam shows palpable bilateral right greater than left adenopathy. The dominant fullness is a 3cm mass at right level II. There is lymphadenopathy bilaterally at level V/posterior triangle. The lymphadenopathy does not extend to the supraclavicular fossa.*
	Cranial Nerve exam is intact.

Relevant bedside procedures:

- A bedside fiberoptic exam is performed:
  - Examination shows a friable mass arising from the right fossa of Rosenmüller. The mass obscures the right choanae and blocks visualization of the right Eustachian tube opening and torus tubarius. The mass extends to the roof of the nasopharynx and inferiorly to the level of the soft palate. Via the left nares the endoscope is passed beneath the mass with good visualization of the larynx. The laryngeal anatomy and vocal cord function are normal.
- An audiogram:
  - Demonstrates a moderate conductive hearing loss on the right with a Type B tympanogram, consistent with a middle ear effusion. The left ear is normal.

What is your differential diagnosis?

- Nasopharynx squamous cell carcinoma
- Minor salivary gland cancer
- Lymphoma
- HIV associated adenopathy
- Adenoiditis

Further investigations or procedures to confirm diagnosis:

- CT scan of the neck with contrast.
- Nasopharynx biopsy in the operating room.

Test results:

- CT scan shows a mass obscuring the right fossa of Rosenmüller with extension to the parapharyngeal space and erosion of the clivus (not seen). A right mastoid effusion is noted (see Figure 1).

Figure 1 – Left: CT scan with contrast. Right: MRI Sinus, T2 image. Collectively, these images reveal a right sided nasopharynx mass with invasion of the pterygoid plates and parapharyngeal space, extending to the skull base. Right mastoid effusion.

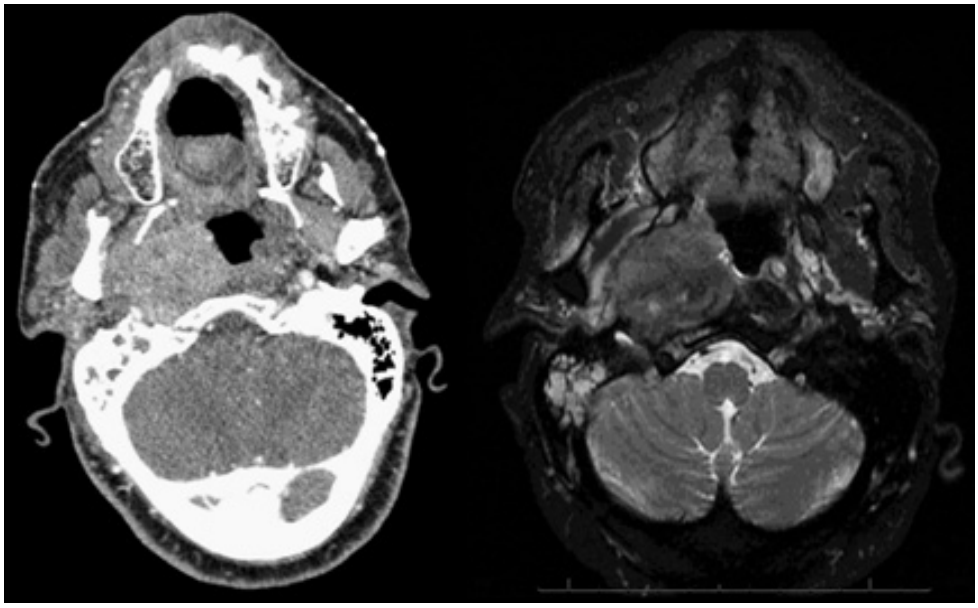


Figure 2 – PET/CT scan. In this image just superior to the hyoid, bilateral level IIA lymph nodes are noted.

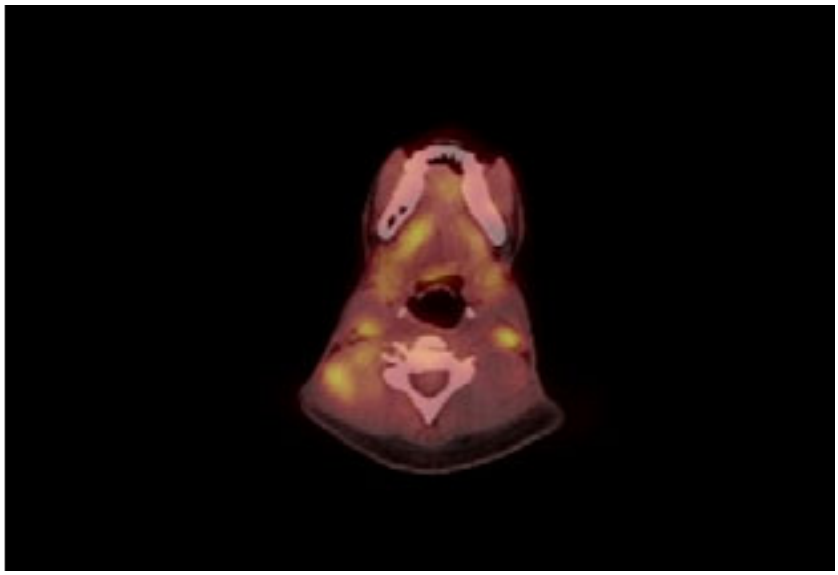
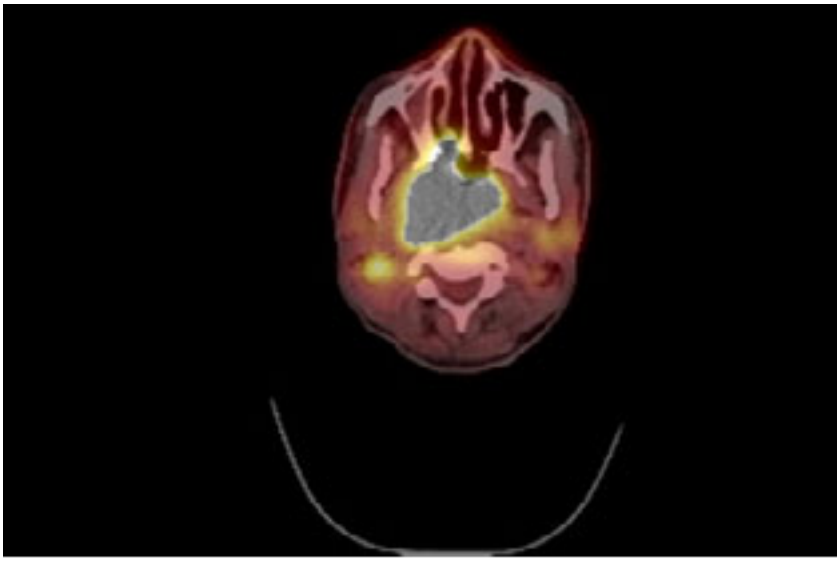


Figure 3 – PET/CT scan. Image showing FDG avid right sided nasopharyngeal tumor. A right level V FDG avid lymph node is also noted.



Confirm diagnosis, include staging information. How would you confirm the diagnosis and stage this tumor?

Based on a biopsy that confirms nasopharyngeal carcinoma and radiological investigations:

- The patient has a T3N2M0 squamous cell carcinoma of the nasopharynx. It is T3 because it erodes into the clivus but does not extend intracranially. The nodal status is N2 because he has bilateral neck disease. This T3N2M0 SCC of the nasopharynx is a Stage III cancer. Note that staging of nasopharynx cancer is distinctly different than other subsites of the head and neck. Both regional disease staging and TNM final staging use distinct criteria.
- His symptoms reflect his disease. The “swelling” in the neck is his lymphadenopathy. The intermittent epistaxis is from primary tumor bleeding. His hearing loss is from a right middle ear effusion secondary to blockage of the Eustachian tube by tumor.

Nasopharynx cancer is classified by the World Health Organization as: 1) Squamous cell carcinoma, typically found in the older adult population; 2) Non-keratinizing carcinoma; 3) Undifferentiated carcinoma.

Tstage:
TX: Primary tumor cannot be assessed.
T0: No evidence of primary tumor.
Tis: Carcinoma in situ.
T1: Tumor confined to the nasopharynx, or tumor extends to oropharynx and/or nasal cavity without parapharyngeal extension.
T2: Tumor with parapharyngeal space extension, and/or adjacent soft tissue involvement (medial pterygoid, prevertebral muscles).
T3: Tumor involves bony structures of skull base and/or paranasal sinuses, cervical vertebra, pterygoid structures.
T4: Tumor with intracranial extension and/or involvement of cranial nerves, hypopharynx, or orbit, parotid gland, or extensive soft tissue infiltration beyond the lateral surface of the lateral pterygoid muscle.

NStage:
NX: Regional nodes cannot be assessed.
N0: No regional lymph node metastasis.
N1: Unilateral metastasis in cervical lymph nodes ≤6cm in greatest dimension, and/or unilateral or bilateral retropharyngeal lymph nodes ≤6cm in greatest dimension (midline nodes are considered ipsilateral nodes) above the caudal border of the cricoid cartilage.
N2: Bilateral metastasis in cervical lymph nodes ≤6cm in greatest dimension, above the caudal border of the cricoid cartilage.
N3: Metastasis in a lymph node >6cm and/or below the caudal border of the cricoid cartilage.

Distant metastasis (M):
M0: No distant metastasis.
M1: Distant metastasis.

Staging:

Stage	T	N	M
0	Tis	N0	M0
I	T1	N0	M0
II	T1	N1	M0
	T2	N0/N1	M0
III	T1	N2	M0
	T2	N2	M0
	T3	N0/N1/N2	M0
IVA	T4	N0/N1/N2	M0
	T Any	N3	M0
IVB	T Any	N Any	M1

AJCC Cancer Staging Form Supplement to the AJCC Cancer Staging Manual, 8th ed., pp. 51-3 June 2018. https://cancerstaging.org/references-tools/deskreferences/Documents/AJCC%20Cancer%20Staging%20Form%20Supplement.pdf. Used with the permission of the American College of Surgeons. Amin, M.B., Edge, S.B., Greene, F.L., et al. (Eds.) AJCC Cancer Staging Manual. 8th Ed. Springer New York, 2017.

Treatment options: Discuss with patient:

- The most common appropriate treatment for a Stage III nasopharynx cancer is concomitant chemoradiation therapy followed by adjuvant chemotherapy.[1] Chemotherapy is usually with Cisplatin. After chemoradiation, additional chemotherapy is given, sometimes termed adjuvant, outback, piggyback or consolidation therapy. Adjuvant chemotherapy is usually either Cisplatin/5FU or carboplatin/5FU. Radiation therapy is usually 66-70 Gy to the primary site and bilateral cervical at-risk lymph nodes.
- Induction chemotherapy followed by concomitant therapy or concomitant chemotherapy alone are also appropriate. This is recommended for all Stage II and higher naso-pharynx cancers treatment.
- Surgery is not part of the standard treatment of nasopharynx cancer in the United States, although it is employed more commonly in Asia.
- The addition of chemotherapy to radiation therapy for advanced nasopharynx cancer was the subject of a major randomized study in 1998.[2] The study was terminated early due to the significant survival benefit from the addition of chemotherapy to radiation therapy. The Intergroup 0099 study evaluated radiation alone vs. concomitant chemoradiation with adjuvant chemotherapy for Stage III/IV nasopharynx cancer. 3 year actuarial survival rates were 24% vs. 69% for RT vs. CRT. This study definitively demonstrated the benefit of chemotherapy for locoregionally advanced nasopharynx cancer. The continued use of piggyback chemotherapy remains controversial. Many patients following concomitant chemoradiation therapy have difficulty completing adjuvant chemotherapy. A recent study suggests that it may not have significant survival impact.[3]
- There are 2 major causes of nasopharynx cancer. One is from tobacco and alcohol, similar to other head and neck cancer sites. These tumors tend to be WHO class I or II. The Epstein Barr Virus (EBV) causes another type of tumor that is endemic to certain geographic regions including Southeast Asia, Northern and Eastern Africa, and Arctic North America in the Eskimo population.[4] Current efforts are underway to better treat EBV positive nasopharynx cancer with an ongoing clinical trial.[5]

Consultations:

- The patient undergoes evaluation by a head and neck surgeon, medical oncologist and radiation oncologist. A nutritionist and speech language pathologist also see him for pre-treatment teaching and evaluation. He will also see a dentist prior to beginning radiation therapy.

Definitive treatment plan:

- The patient is planned to undergo concomitant chemoradiation therapy with cisplatin and 3 cycles of carboplatin/5FU chemotherapy afterwards.

Follow-up plan:

- Following treatment, he will be seen at regular intervals over the next 5 years, usually starting with every 3-4 months for the first few years. There is no standard follow-up regimen or imaging strategy recommendation.[1] He will undergo a post-treatment PET/CT at 12 weeks after treatment. Suspicious residual lymphadenopathy may require additional workup and possible salvage neck dissection.
- He may undergo routine primary site imaging, such as MRI, as determined by the treatment team.

Management of complications: What are the possible complications and their treatment?

- Hearing loss. This can be sensorineural loss from cochlear damage from chemotherapy/radiation therapy, or conductive due to chronic middle ear fluid from Eustachian tube dysfunction after radiation. In this latter scenario, myringotomy and tube placement can improve hearing and function.

- Neck pain secondary to chemoradiation therapy. Management with pain medications.
- Xerostomia secondary to radiation therapy. Management with aggressive dental care and oral hydration.

References:

1. NCCN - National Comprehensive Cancer Network. Available from: https://www.nccn.org/professionals/physician_gls/pdf/head-and-neck.pdf
2. Al-Sarraf M, LeBlanc M, Giri PG, et al., Chemoradiotherapy versus radiotherapy in patients with advanced nasopharyngeal cancer: phase III randomized Intergroup study 0099. J Clin Oncol, 1998; 16(4): 1310-7.
3. Chen L, Hu CS, Chen XZ, et al. Concurrent chemoradiotherapy plus adjuvant chemotherapy versus concurrent chemoradiotherapy alone in patients with locoregionally advanced naso¬pharyngeal carcinoma: a phase 3 multicentre randomised controlled trial. Lancet Oncol. 2012; 13(2): 163-71.
4. Busson P, Keryer C, Ooka T, et al. EBV-associated nasopharyngeal carcinomas: from epidemiology to virus-targeting strategies. Trends Microbiol. 2004; 12(8): 356-60.
5. NRG HN 002. Available from: https://www.nrgoncology.org/Clinical-Trials/NRG-HN002.