Chapter 11
Supportive Care for Patients with Sinonasal and Skull Base Tumors

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Introduction

Skull base and sinonasal tumors represent a heterogeneous group of histologies of varying malignant potential arising in one or multiple sites, each with distinct clinical implications. The skull base is loosely defined as the junction between the cranium and the face, eyes, ears, and sinonasal cavity. Tumors in the skull base and sinonasal region have potential to invade the cerebral cortex, brainstem, cranial nerves, cervical spine, nasopharynx, the orbit, the inner ear, and pituitary sella. Given their close proximity to these vital structures, the tumor and treatment can adversely affect physical function and quality of life.

Historically, clinical and research priorities focused on improving survival. Surgical advances and multidisciplinary care have improved survival dramatically in the past half century. At MD Anderson Cancer Center, for instance, unpublished 5-year survival for sinonasal and skull base malignancies doubled from 35% in the

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1950s to 70% among patients diagnosed in the last decade. In this same time, the aim of surgical resection of these tumors has increasingly focused on reducing functional impact particularly by preserving adjacent structures through endoscopic endonasal technique [1]. However, tumors in this site remain a significant operative challenge given vital neurovascular structures within the vicinity of the skull base and sinonasal cavity.

Functional implications of skull base and sinonasal malignancies are challenging to summarize given the heterogeneous presentations of the disease that results in a wide array of symptoms and functional changes. It is well established that both physical morbidities and patient-reported outcomes vary primarily based on tumor location, surgical approach, and use of radiotherapy (RT) [2–4]. Physical morbidities reported include endocrine (e.g., skin alteration, weight gain, abnormal hair growth, lactation, amenorrhea, hand/foot growth), nasal (e.g., anosmia, chronic nasal drainage, crusting and blockage), neurologic (e.g., cognitive, cranial neuropathy), lymphatic, and visual (e.g., diplopia, optic neuropathy) pathologies [2, 5]. Patient-reported symptoms mirror these physical domains with implications on emotional, financial, social, spiritual, and family wellbeing [3, 4]. This chapter focuses on areas for which established supportive care may improve outcomes—that is, swallowing, communication, and cognitive function, as well as lymphedema. Early intervention with a multidisciplinary team including supportive care specialties is best practice to maximize functional outcomes in this complex population.

Skull Base–Specific Patient-Reported Outcome (PRO) Instruments

Prospective and cross-sectional studies report quality of life decrements after treatment for skull base and sinonasal tumors that improve by 6–12 months, on average, but fail to fully recover in all patients [4–7]. After anterior skull base resection, for instance, almost 60% of patients have persistent physical symptoms on average 40 months postoperatively [8]. Thus, ongoing assessment of the patient experience is vital to enhance the quality of care and triage patients to appropriate supportive care. Significant progress has been made to characterize both clinician-graded and patient-reported outcomes in recent decades. Two psychometrically validated instruments are available to capture QOL with disease-specific domains for skull base tumors:

1. *Anterior Skull Base (ASB) Questionnaire* [4]: 35-item with 6 domains (performance, physical function, vitality, pain, specific symptoms, and influence on emotions) developed originally for open surgical populations.
2. *Skull Base Inventory (SBI)* [3, 9]: 41 items (26 disease specific items) with 10 domains (cognitive, emotional, family, financial, social, spiritual, endocrine, nasal, neurologic, visual) developed for patient with anterior and central skull base pathology, and validated in a surgical cohort.

A third skull base–specific PRO instrument is under development focusing specifically on symptom burden—the *MD Anderson Symptom Inventory Skull Base Module (MDASI-SB)*.

Swallowing

Neuroanatomy and Physiology

Swallowing is the act of transferring food from the mouth into the stomach that involves both volitional and reflexive actions. Swallowing is historically described in four phases: oral preparatory, oral, pharyngeal, and esophageal. The oral preparatory phase involves mastication and manipulation of a food into a cohesive bolus. During the oral phase, the tongue propels the bolus posteriorly into the oropharynx. The swallow trigger initiates a rapid sequence of events during the pharyngeal phase. The soft palate elevates to seal the nasal cavity to prevent nasal regurgitation. The base of the tongue retracts and contacts the bulging posterior pharyngeal wall. The pharyngeal constrictor muscles sequentially contract medially and laterally to propel the bolus downward through the pharynx. To prevent airway invasion or aspiration, the hyoid and larynx move anterior and superiorly, the epiglottis is deflected downward to cover the airway entrance while the vocal cords adduct to create a protective seal in the laryngeal vestibule. Transitioning into the esophageal phase, the cricopharyngeal muscle relaxes and the upper esophageal sphincter opens allowing the bolus to enter the cervical esophagus with peristaltic transfer of the bolus to the gut.

The four phases of swallowing are regulated by a central pattern generator within the medulla of the brainstem and involve sensorimotor innervation to the oropharynx, larynx, and esophagus provided by multiple cranial nerves including trigeminal (V), facial (VII), glossopharyngeal (IX), vagus (X), and hypoglossal (XII). Swallowing impairment, or dysphagia, is one of the most dangerous complications related to surgery for patients with skull base tumors [10]. Dysphagia can lead to longer hospital stays, altered diets, necessitate enteral feeding tubes or tracheostomy, and result in aspiration pneumonia and although infrequent, even death [10–12].

Dysphagia Outcomes

With surgical advances shifting practice historically from open resections to endoscopic technique, there has been substantial improvement in speech and swallowing outcomes for patients with anterior skull base tumors, including sinonasal tumors. Previously, for instance, structures such as the maxilla and mandible would be altered during open resections with subsequent impact to oral preparatory and oral phases of swallowing. Now, with endoscopic surgical approach providing a minimally invasive option, the architecture of normal craniofacial structures is maintained to better preserve both appearance and also function with significantly lower risk for cranial nerve injuries, while still demonstrating similar survival outcomes [13].

Despite these advances, surgical resection of middle and posterior fossa tumors continues to threaten swallowing function not only due to risk of cranial nerve injury but also due to the proximity to the brainstem. In published case series, one-third of patients undergoing resection of posterior fossa tumors demonstrate dysphagia postoperatively [12, 14]. A smaller number require enteral feeding tube, which is associated with older age (58 years old vs. 46 years old), aspiration, and lateral surgical approach [12]. Swallowing was impacted in all phases of swallowing. Modified barium swallow (MBS) studies of patients after surgery of cerebellopontine angle (CPA) tumors, for instance, demonstrated oral (51%), pharyngeal (12%), and oropharyngeal (37%) swallowing difficulties [14]. Anticipated swallowing outcomes as they relate to specific cranial nerve injury and typical skull base tumors are detailed below.

Trigeminal Nerve (V)

Trigeminal schwannomas, although uncommon, originate within the ganglion of the trigeminal nerve arising in the middle fossa and may extend into the posterior fossa. Trigeminal nerve (CN V) injury can cause deficits with oral containment and mastication due to impaired jaw movement and reduced facial, mouth, and jaw sensation. V3 division of the trigeminal nerve also provides motor innervation to floor of mouth muscles (mylohyoid, anterior digastric) that are critical to hyoid excursion in the pharyngeal phase of swallowing.

Facial Nerve (VII)

Relatively one-third of patients with vestibular schwannoma, also referred to as an acoustic neuroma, demonstrate immediate or delayed facial palsy (3 days) after surgery [1]. Fortunately, the majority of these patients recover at least partial nerve function within 1 year [1, 11]. Facial nerve injury impacts facial movement and sensation resulting in residue in lateral sulci and anterior spillage from poor labial seal. On MBS, 64% of patients with facial nerve palsy demonstrated anterior spillage [14]. These deficits may adversely impact swallowing safety or efficiency and result in a modified diet level [11].

Glossopharyngeal Nerve (IX)

Glossopharyngeal nerve (IX) injury may influence both safety and efficiency of the pharyngeal phase of swallowing. Damage to the afferent limb of IX can cause a delayed or absent swallowing trigger, and motor injury (stylopharyngeus) impairs

pharyngeal shortening that is critical to laryngeal lift and bolus clearance. Resultant swallowing problems may include aspiration before and during the swallow, pharyngeal residue, and nasal regurgitation. Damage to the glossopharyngeal nerve often co-occurs with vagal nerve injury given they both exit through the jugular foramen. Nearly half of patients with unilateral vagal palsy after CPA surgery also present with pharyngeal palsy [15].

Vagus Nerve (X)

Postoperative unilateral vagal nerve palsy (X) was demonstrated in 10% of patients undergoing CPA surgery [15]. Fortunately, many patients (42–76%) demonstrated varying degrees of recovery of vagal function from 2 weeks, 3 months, and 1 year postoperatively [11, 15]. Damage to the pharyngeal branch can result in pharyngeal residue and swallowing inefficiency due to reduced pharyngeal contraction. Superior laryngeal nerve injury impairs sensation to airway entry and is associated with silent aspiration wherein a patient does not cough or show any signs of awareness of food or liquid entering the airway. Recurrent laryngeal nerve injury (RLN) may result in vocal fold paresis and impact glottic closure. Patients with RLN injury demonstrate a weak cough and poor vocal fold closure, which can also reduce airway protection during the swallow. Over two-thirds of this population with vagal nerve injury demonstrate aspiration [11].

Hypoglossal Nerve (CNXII)

Hypoglossal nerve injury is a risk among patients with jugular foramen tumors within the posterior fossa [16], and is also expected with hypoglossal schwannomas. Hypoglossal nerve (CN XII) controls the motor movements of the tongue muscles. Injury may cause deficits in lingual range of motion and strength, thus influencing bolus formation and transportation through the oral cavity and pharynx and result in oral and pharyngeal residue.

Evaluation

Evaluation of swallowing includes clinical and instrumental assessments in addition to patient reported outcome measures [17].

Clinical Swallowing Evaluation

A clinical swallowing evaluation, also referred to as a bedside swallowing assessment, is often the first level of assessment. A trained speech pathologist conducts a thorough chart review, clinical interview, oral motor examination, and motor speech profile to identify risk factors for oropharyngeal dysphagia. The clinician then assesses adequate labial closure, mastication, oral clearance with a variety of liquid and solid consistencies. Pharyngeal swallow function is inferred by palpating laryngeal elevation, counting the number of swallows per bolus, and observing clinical indicators of aspiration including coughing, throat clears, or wet vocal quality. Patient's cognitive status is considered in relation to their level of alertness, attention, judgment, and impulsivity and how these factors and their feeding behaviors may impact swallowing safety. A variety of compensatory strategies or diet modifications may be trialed to alleviate signs and symptoms of dysphagia and/or aspiration. Further testing is often recommended to detail physiology of the oropharyngeal swallow using instrumentation such as the MBS study or fiberoptic endoscopic evaluation of swallowing (FEES).

Modified Barium Swallow Study

A modified barium swallow (MBS) study also referred to as a videoflouroscopic swallow study (VFSS) is a dynamic x-ray of the oropharyngeal swallow completed by a speech pathologist in conjunction with a radiologist. During MBS, a patient is tested swallowing a standard protocol of barium contrast agents to assess the safety and efficiency of bolus flow during all four phases of swallowing. The MBS provides data safety and efficiency of bolus clearance as well as details of anatomy and physiology of disordered swallow to guide restorative and compensatory treatment recommendations. Figures 11.1 and 11.2 show examples of a normal and disordered swallow as tested on MBS.

Fiberoptic Endoscopic Evaluation of Swallowing

Fiberoptic endoscopic evaluation of swallowing (FEES) is completed at bedside or in the clinic using a transnasal fiberoptic endoscope while the patient swallows a standard series of bolus trials. FEES can directly visualize aspiration before and after a swallow and detect pharyngeal residue. However, FEES can only infer oral

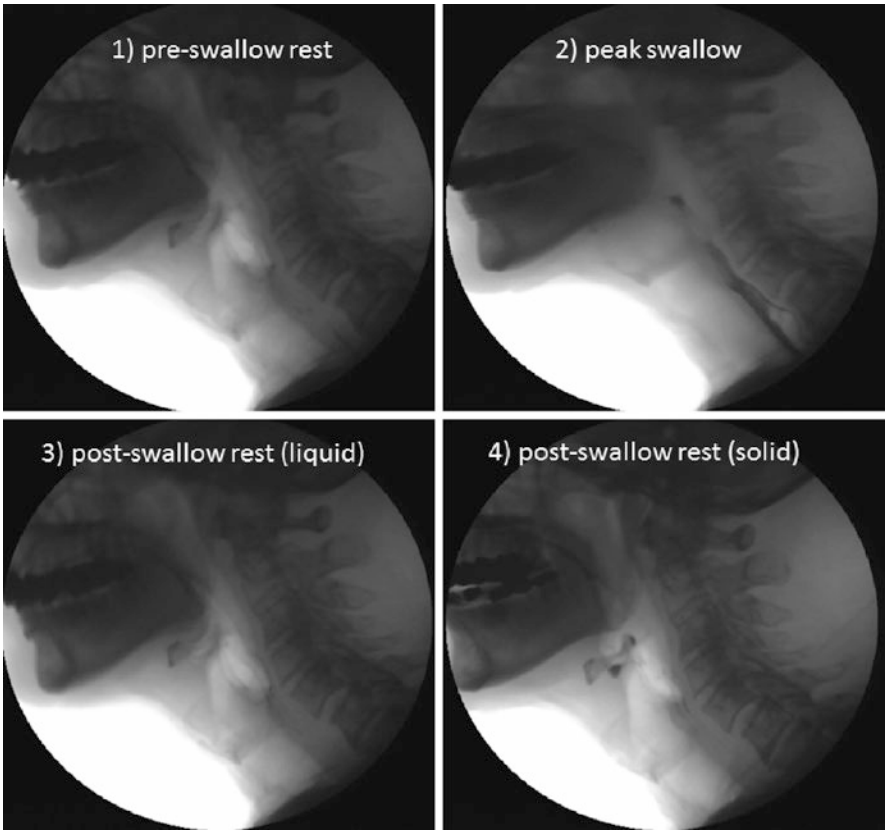

Fig. 11.1 Example images from a normal modified barium swallow (MBS) study. Image series demonstrate normal pharyngeal swallow function with near complete pharyngeal constriction and complete laryngeal vestibule closure at peak swallow (frame 2) then clear airway and negligible residue post-swallow (frames 3 and 4)

and esophageal phases as well as events that occur during the swallow due to white out of the image at peak swallow when the pharynx contracts around the endoscope. Unlike the MBS, FEES allows direct visualization of the aerodigestive tract structures including the ability to assess vocal fold mobility, laterality of pharyngeal constriction, and velopharyngeal function which can be beneficial for this patient population given risk for vagal injury. FEES can also assess compensatory strategies and guide treatment recommendations, and FEES is a useful method to provide biofeedback during therapy when training patients to more safely or efficiently clear foods or liquids through the pharynx. Figures 11.3 and 11.4 show examples of a normal and disordered swallow as tested on FEES.

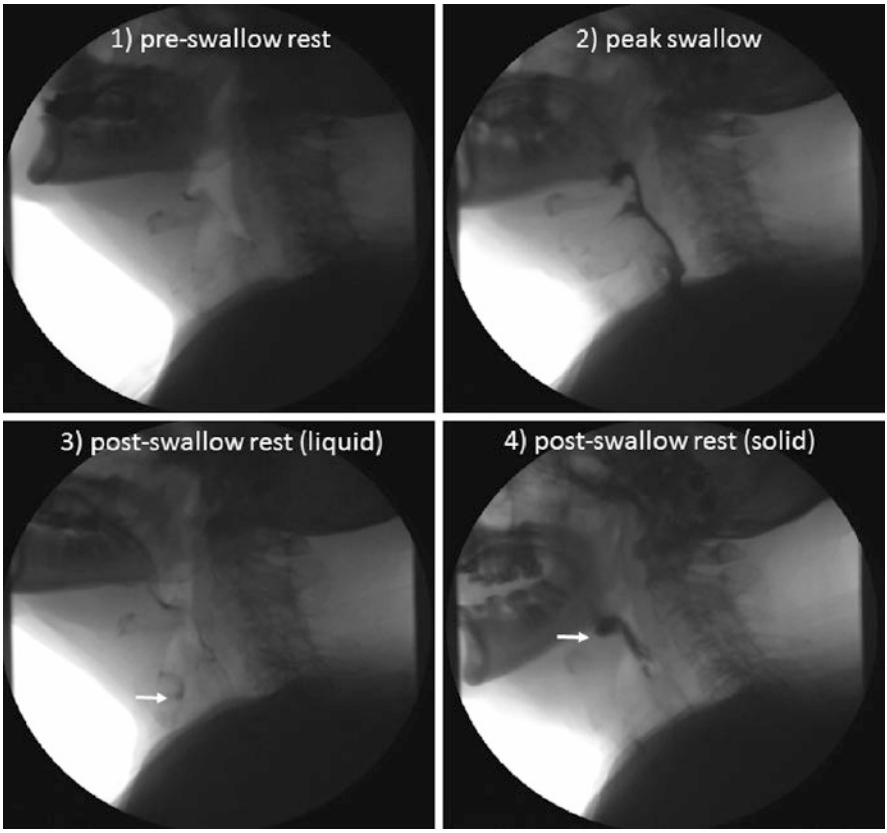

Fig. 11.2 Example images from a disordered modified barium swallow (MBS) study. Image series demonstrate disordered swallow function with incomplete pharyngeal constriction and laryngeal vestibule closure peak swallow (frame 2) then laryngeal vestibule residue aspirated post-swallow (frame 3) and solid food residue in valleculae and pharynx (frame 4)

Patient-Reported Outcome Measures of Dysphagia

Patient-reported outcome (PRO) measures including quality of life (QOL) instruments may also be included in a comprehensive battery to integrate a patients' perception of their swallowing function into a rehabilitation program [16]. A variety of relevant swallowing-related PRO tools are available, including the Performance Status Scale for Head and Neck (PSS-HN) [18], Eating Assessment Tool-10 [19], MD Anderson Dysphagia Inventory (MDADI) [20], and the SWAL-QOL [21]. These instruments provide detailed information about the patient's current perception of swallowing function.

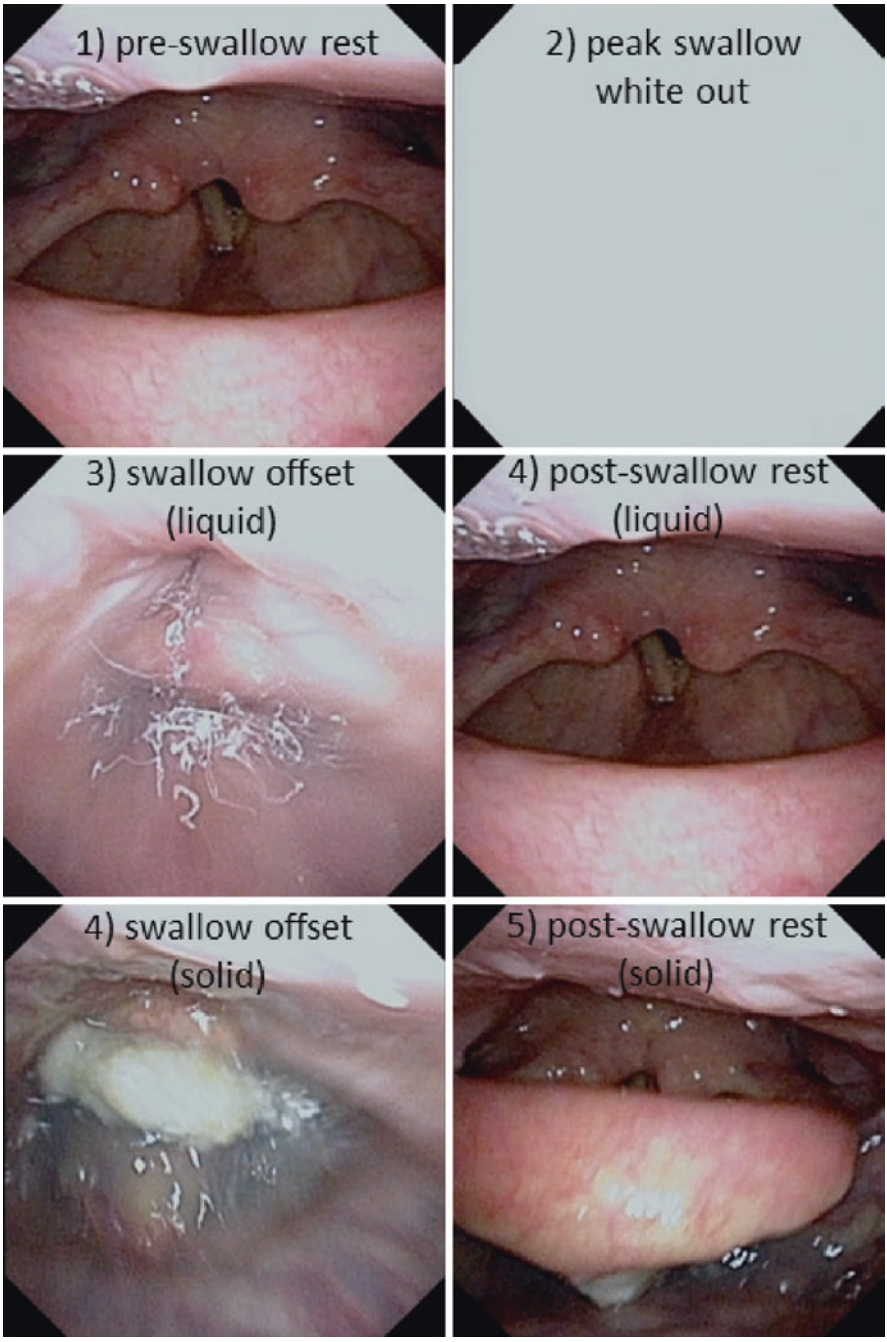

Fig. 11.3 Example images from a normal fiberoptic endoscopic evaluation of swallowing (FEES). Image series demonstrate normal pharyngeal swallow function without penetration or aspiration with typical white out at peak swallow as the pharynx is maximally constricted around the endoscope (frames 1–4). Frames 4 and 5 demonstrate mild vallecular residue on the cracker bolus visualized both at swallow offset and post-swallow rest

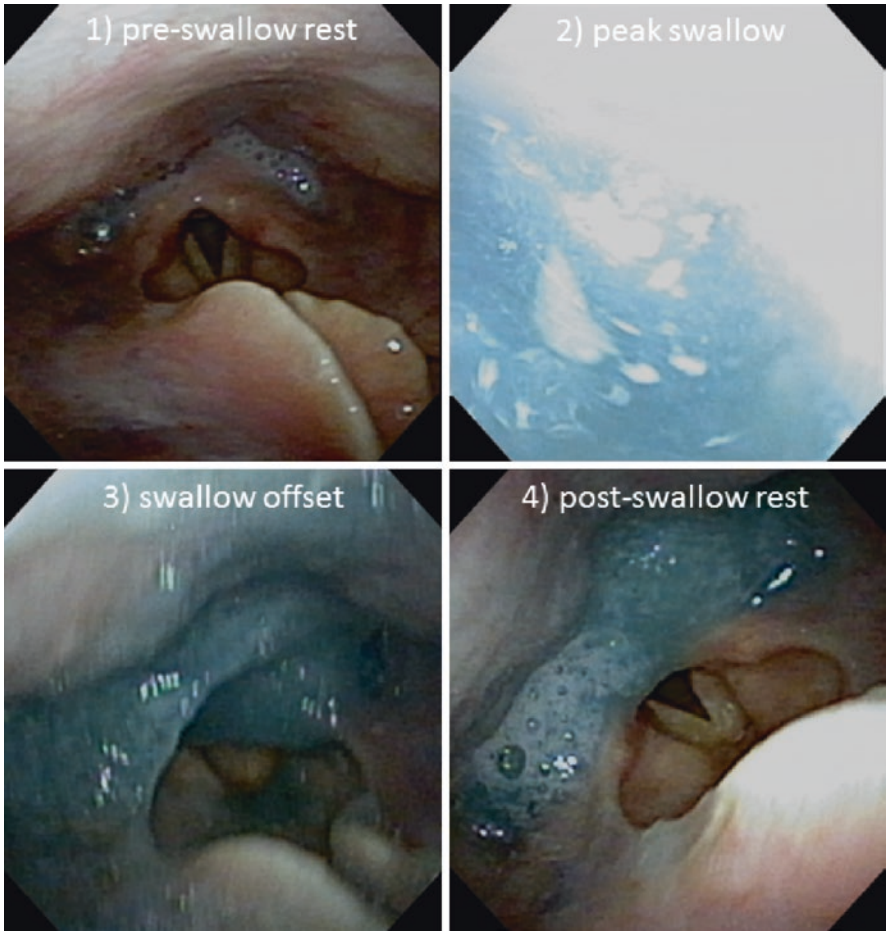

Fig. 11.4 Example images from a disordered fiberoptic endoscopic evaluation of swallowing (FEES). Image series demonstrate abnormal pharyngeal swallow function with pooling in piriform sinuses and supraglottic fibrosis visible pre-swallow (frame 1), absence of typical white out at peak swallow indicating incomplete pharyngeal constriction (frame 2), and severe post-swallow residue (frames 3 and 4)

Dysphagia Rehabilitation

Rehabilitation of swallowing deficits may include compensatory and/or restorative techniques. Compensatory strategies are used to improve bolus flow or eliminate aspiration via diet modifications (e.g., thickened liquids, pureed food), swallowing techniques (e.g., supraglottic swallow) or postural changes (e.g., head turn, chin tuck maneuver). Compensations can facilitate short-term function while targeting long-term improvement. Restorative therapy targets improved strength or coordination of swallowing based on physiologic deficits identified on instrumental

assessment [15, 16]. There are a variety of therapy techniques including: traditional swallowing exercises (e.g., effortful swallow), McNeil Dysphagia Therapy [22–24] program that involves a hierarchical arrangement of foods to in mass practice, biofeedback via FEES or surface electromyography, and device facilitated exercise therapy such as expiratory muscle strength training [25]. Instrumental assessment and patient-centered goals should drive rehabilitation plan of care.

Voice

Neuroanatomy and Physiology

Voice production requires coordination of laryngeal and respiratory muscles to produce sound for speech, referred to as phonation. During exhalation, subglottic pressure is controlled to initiate and maintain vocal fold vibration. Sufficient abduction and adduction of the true vocal folds must occur to produce adequate voicing and regulate breathing. Vagus nerve (CN X) innervation is integral for voice production. The superior laryngeal branch of the laryngeal nerve allows change in pitch through shortening and elongating the cricothyroid muscle. The recurrent laryngeal nerve branch provides motor innervation to the intrinsic muscles of the larynx allowing for vocal fold movement.

Voice Outcomes

It is estimated that 10% of patients undergoing CPA surgery demonstrated unilateral vagal palsy, with high rates of postsurgical recovery [11, 15]. Even transient postoperative unilateral recurrent laryngeal nerve injury may adversely affect both swallowing and voice production. The glottic gap that results from incomplete vocal fold approximation causes a weak and breathy voice as well as a weak cough. Although rare, bilateral vocal fold paresis can occur as a potentially life-threatening complication that may require tracheostomy if the airway is inadequate.

Voice Evaluation

The cornerstone of voice assessment is endoscopy. Laryngoscopy is a diagnostic tool often completed in the clinic but also possible at the bedside. A flexible endoscope is passed through the nose or rigid endoscope through the mouth to visualize laryngeal anatomy, pathology, airway aperture, and true vocal fold mobility. Comprehensive voice assessment also includes the patient's self-assessment such as

the Voice Handicap Index (VHI) to quantify the psychosocial consequences of a voice disorder [26]. Clinical assessment measures of respiratory patterns, maximum phonation time, and auditory perceptual rating of the voice by the speech pathologist assess overall grade of dysphonia, roughness, breathiness, asthenia or weakness, and strain to accompany the imaging result. Acoustic assessment using computer software objectively profile parameters of vocal loudness, pitch, and quality.

Vocal Rehabilitation

Rehabilitation of vocal fold paresis and paralysis depends on several factors. Primarily, this includes laterality (unilateral vs. bilateral paresis), airway aperture, degree of dysphonia, aspiration, and cough function. For unilateral paralysis resulting in glottic gap, vocal fold augmentation with injection or surgical medialization thyroplasty can provide temporary or permanent correction of glottic incompetence. An injection or implant moves the paralyzed vocal fold closer to midline to facilitate easier closure by the mobile contralateral cord. Vocal fold medialization is reportedly used in 10–29% of cases with unilateral paralysis after skull base surgery with improvements noted in both voice and swallow [11, 15]. In one study, the majority of patients achieved complete glottic closure with good to excellent voice quality after postoperative injection [27]. Expected swallowing improvements after medialization include less aspiration and better cough strength to clear aspirate resulting from improved glottic closure. Nonsurgical voice therapy can also be of benefit including exercise paradigms (e.g., PHoRTE and respiratory muscle strength training) [25, 28].

Speech

Neuroanatomy and Physiology

Speech production requires sensorimotor regulation of the muscles involved in respiration, phonation, articulation, resonance, and prosody. Motor regions of the cortex, basal ganglia, brainstem, and cerebellum innervate the diaphragm, larynx, tongue, lips, and soft palate to influence precision, intelligibility, quality, and rate of speech. Injury resulting weakness or incoordination of the speech muscles causes a speech disorder, or dysarthria. There are many different types of dysarthria depending on the area of the brain or periphery that is injured.

Dysarthria Outcomes

Up to 30% of patients after skull base surgery present with speech deficits or dysarthria [29]. Patients with posterior fossa or CPA tumors commonly present with flaccid and ataxic dysarthria. Flaccid dysarthria results in imprecise articulation with reduced speech intelligibility secondary to weakened speech muscles from cranial nerve injury or brainstem compression. Specific qualities depend on the cranial nerve or supratentorium injured. Facial nerve (VII) injury impacts labial closure and strength that can reduce speech intelligibility. Glossopharyngeal (IX) and pharyngeal branch of the vagus nerve (X) injury results in velopharyngeal insufficiency that causes resonance disturbance. Hypoglossal (XII) injury results in tongue weakness that reduces speech intelligibility by way of imprecise articulation of lingual speech sounds. Direct or indirect damage to the cerebellum from CPA tumors can cause ataxic dysarthria. The cerebellum helps to coordinate the muscles of speech including respiration, phonation, and articulation. Incoordination due to cerebellar damage causes prominent fluctuations in speech prosody, loudness, and articulation. Perceptually, a patient's speech sounds "drunk."

Speech Evaluation

The underlying processes of speech are assessed through an auditory perceptual evaluation by a speech pathologist. Rate, rhythm, and precision of speech production are evaluated during a standardized set of speech tasks including repeating sounds in isolation and rapidly in a sequence, reading tasks, and conversational speech. Respiration, phonation, articulation, resonance, and prosody are assessed. Standardized assessments such as the Assessment of Intelligibility of Dysarthric Speech may be used to quantify intelligibility and track progress [30]. Patient perception of their communication effectiveness can be assessed using tools such as the Communicative Effectiveness Scale.

Speech Rehabilitation

Rehabilitation of dysarthria includes impairment-based treatment and compensatory management. Non-speech oromotor exercises are not typically recommended but instead behavioral speech practice. Diaphragmatic breathing and pacing techniques may be trained for a patient with ataxic dysarthria. Simple compensatory strategies may be taught to increase intelligibility in flaccid dysarthria, such as slow rate of speech and overarticulation. In severe cases, alternative and augmentative communication devices may be necessary.

Cognitive-Communication

Neuroanatomy and Physiology

Cognitive and communication skills, including higher mental functions such as reasoning, memory, and language, are organized within the cerebral hemispheres. The four major cortical lobes, frontal, temporal, parietal, and occipital and their cortical gyri operate as modules dedicated to specific cognitive or behavioral functions [31]. These modules are interactive networks that interconnect regions of the brain and with the subcortical centers. A variety of cognitive-communication impairments may arise from cortical damage depending on the area of the cortex impacted by skull base tumors and their treatment.

Cognitive-Communication Impairment Outcomes

Cognitive-communication impairment in patients with skull base tumors are rare but relevant. Patients who underwent resection for large anterior skull base meningiomas have shown frontal lobe damage, specifically within the ventromedial prefrontal cortex. Deficits were seen in verbal memory, information processing, adaptive functions, and real-life decision making as well as acquired personality changes and poorly modulated emotional reactions [32, 33]. Children after posterior skull base tumor resection have presented with a profound communication deficit termed cerebellar mutism syndrome (CMS). Children typically present with a transient mutism 12–96 hours after surgery that typically resolves weeks or months later [34].

Cognitive-Communication Evaluation

A variety of standardized assessments can assess cognition and language skills when impairment is suspected. Neuropsychologists and speech pathologists are the primary providers for this testing. Cognitive testing measures a patient's memory, executive functions, visuospatial skills, and attention. Language testing measures expressive and receptive skills during speech, listening, reading and writing tasks. Social pragmatics is also assessed, especially for patients with frontal lobe damage, including personality changes, emotional regulation, impulsivity, initiation, turn-taking, and eye contact. A thorough evaluation must also include functional interviewing to establish patient and family centered goals.

Cognitive-Communication Rehabilitation

Cognitive-communication rehabilitation involves three main approaches: restorative, compensatory, and metacognitive goals. Restorative goals attempt to decrease impairments in cognitive processes by incorporating the principles of neuroplasticity through mass practice of salient tasks (e.g., spaced retrieval training for memory loss). Compensation aims to decrease impairments in cognitive processes, such as training of the use external aids (e.g., memory book). Metacognitive therapy involves training self-monitoring skills to judge performance and implement strategies as needed (e.g., goal-attainment scaling). Therapy goals are established based on cognitive domains identified on testing, as well as family and patient input on activities and participation barriers.

Lymphedema

Lymphatic System

The healthy, intact lymphatic system is a one-way transportation system for fluid and proteins to travel from the interstitial space back to the circulatory system. The principal conduits in the lymphatic system include the capillaries, collecting vessels, lymph nodes, trunks, and ducts. Lymphatic fluid (LF), or lymph, forms when the interstitial fluid moves into the lymphatic capillaries. Lymph drains from the capillaries into the collecting vessels. The vessels enter the lymph node(s) where lymph is filtered. Lymphatic fluid is then carried by the exiting vessel to larger trunks, leading to lymph ducts. The ducts deliver the lymph back to the bloodstream, completing the circuit of fluid transport [35].

Lymphatic drainage patterns in the head and neck are predictable but variable. The most common drainage pattern of the nasal cavity and midface involve lymph transport via the facial vessels inferiorly to the neck lymph nodes of level I–II as well as the retropharyngeal nodes (Fig. 11.5). Lymphatic drainage patterns while primarily used to predict regions at risk for regional spread of disease can also aid in predicting regions of lymphedema [36, 37].

Lymphedema

Secondary, or acquired lymphedema, is a common morbidity that can result from head and neck cancer treatment. Based on lymphatic drainage patterns and field of oncology treatment, sites of lymphedema are highly predictable. Lymphedema results from disruption of lymphatic fluid transportation to the central circulatory system, which

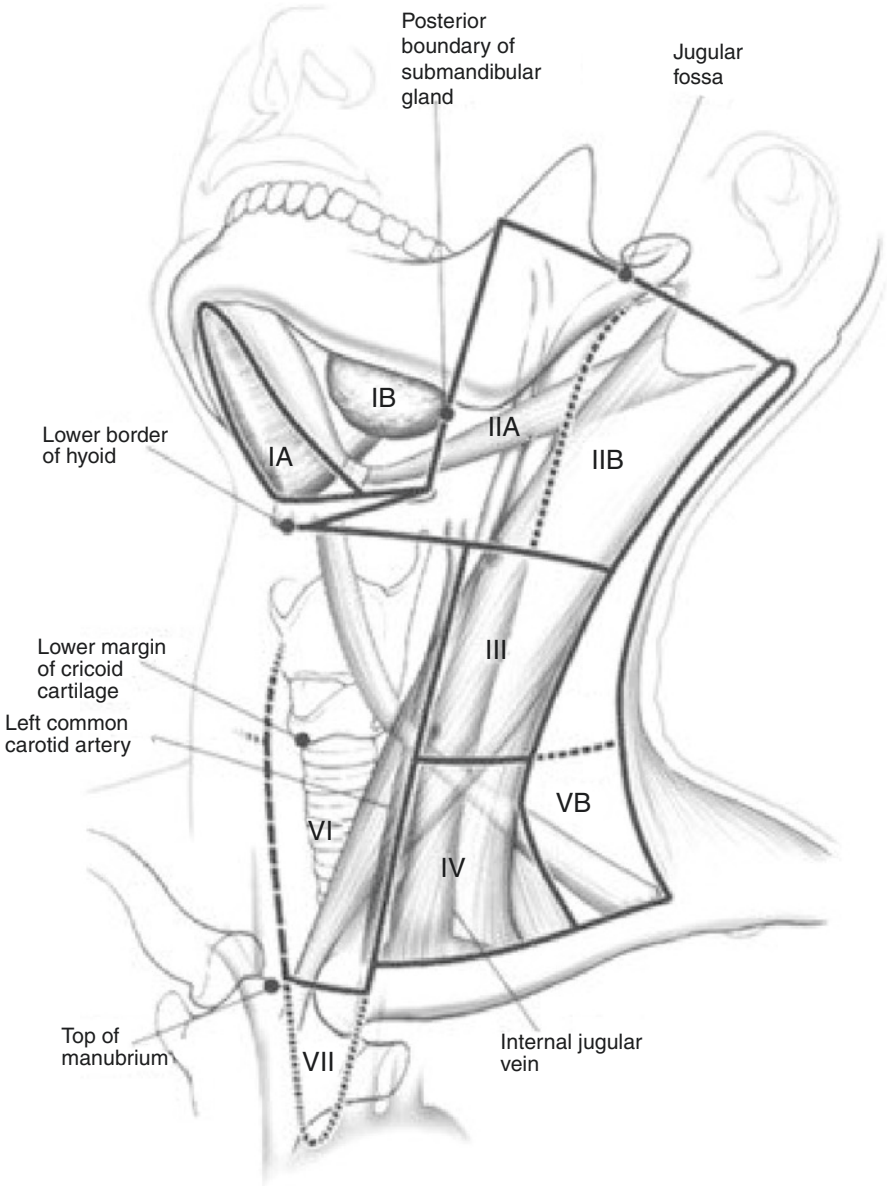

Fig. 11.5 Cervical lymph node levels. This figure by the American Joint Committee on Cancer shows an illustration on the classification of cervical lymph nodes (LN) levels. The following regions represent LN levels: submental and submandibular, levels 1a/b; Upper internal jugular chain, levels 2a/b; Middle internal jugular chain, level 3; Lower internal jugular chain, level 4; Spinal accessory chain and transverse cervical chain, levels 5a/b; Anterior cervical LNs, levels 6 and 7, are often omitted from the head and neck but commonly referred to as superior mediastinal nodes. (Reprinted with permission from Som et al., American Journal of Roentgenology. 2000;174: 837–844. 10.2214/ajr.174.3.1740837)

can cause lymph fluid retention or backup. This disruption can result from tumor burden, surgery, and/or radiation +/- chemotherapy. Within the surgical or radiation field, external lymph congestion manifests as visual or palpable swelling while most internal lymphedema can only be seen with imaging. Head and neck lymphedema (HNL) is highly prevalent, reported in up to 75% of patients who undergo surgery and/or radiation [38]. Functional, physical, psychological, and decreased quality of life symptoms are associated with internal and external head and neck lymphedema [39]. Lymphedema is particularly of concern among patients with sinonasal malignancies.

Lymphedema Evaluation

Lymphedema assessment in head and region typically includes: an oral mechanism examination, motor speech profile, swallow assessment, speech assessment, 2-D and 3-D images of the head and neck, physical examination as well as circumferential face and neck measurements (Fig. 11.6). Various methods exist to grade HNL based on physical examination. By palpation and functional assessment, the

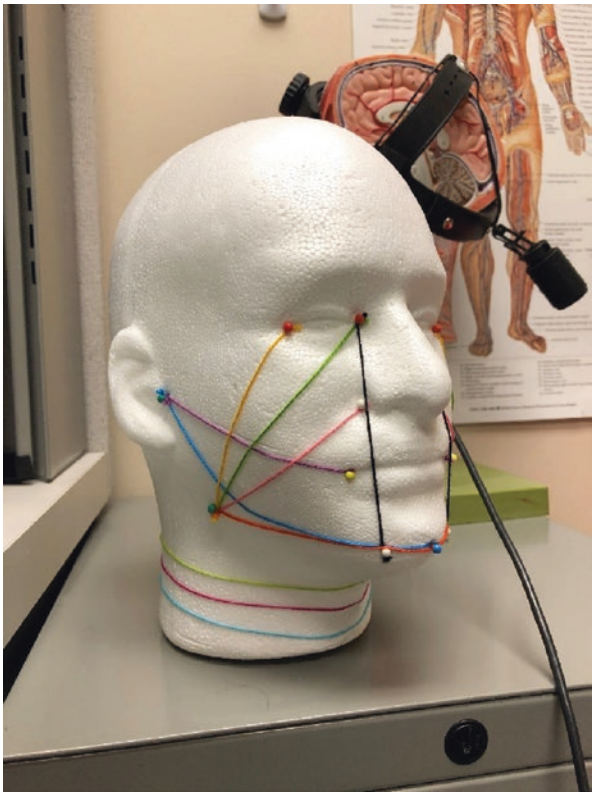

Fig. 11.6 Measurement of head and neck lymphedema. Example of surface tape measurement for quantifying the degree and volume of facial edema as first described by Piso et al. [53], with added circumferential neck measures as described by Smith et al. [42]

examiner may quantify the degree of lymphedema (and fibrosis) according to the following:

- American Cancer Society Lymphedema Scale
- Stages of Lymphedema (Földi's Scale) [40]
- Common Terminology Criteria for Adverse Events (CTCAE) [41]
- MD Anderson Cancer Center Head and Neck Lymphedema Scale (an adaptation of Földi's system that is specific to the head and neck) [42]
- The Head and Neck External Lymphedema and Fibrosis (HN-ELAF) Assessment Criteria [43]

Standardized lymphedema assessments can be repeated longitudinally. Timelines for serial assessment vary based on change in medical status, number of outpatient treatments, and compliance with treatment protocols at home. In general, serial evaluations are completed every 2–3 months until lymphedema has resolved or stabilized [42, 44, 45].

Lymphedema Treatment

Complete decongestive therapy (CDT) is widely regarded as the "gold standard" and is the most researched therapeutic intervention for lymphedema. CDT is comprised of 4 components, including skin care, manual lymphatic drainage (MLD), compression bandaging or garments, and exercise. Protocols of treatment call for an acute "intensive" phase in clinic or at home, followed by a long-term, maintenance program at home [42, 44–49].

Meticulous skin care is recommended indefinitely. Patients are educated to moisturize and protect their skin to decrease the risk of infection. MLD is a lightweight, highly directional massage technique used to increase lymphokinetic activity and stimulate the movement of lymphatic fluid. Compression reduces ultra-infiltration of soft tissues, improve lymph movement as well as joint and muscle pumping. Lymphedema exercises are intended to further assist with lymphatic fluid movement, especially while wearing compression which provides additional resistance for joint and muscle pump efficiency. Contraindications to treatment include cellulitis or other infection in the head and neck, local fistula, deep vein thrombosis, cardiac edema, kidney failure, and dermal metastasis [42, 47, 49]. Physician clearance is prerequisite before initiating treatment.

Alternative treatments for HNL are emerging. Surgical management has included lymphovenus anastomosis (LVA) in a small number of patients with refractory HNL that failed conservative treatment. Studies report a reduction in HNL circumference and improvement in appearance following invasive surgical interventions [48]. Liposuction in the submental region is also used to treat HNL. Brake [50] and Alamoudi [51] report improvements following liposuction based on patients' self-perception of appearance without objective measurements [48–50]. Oral administration of selenium yields mixed results in the reduction of HNL [48]. Non-invasive

procedures including various Kinesiotaping methods mimic skin stretch and soft tissue lift have been offered as a complement to conventional CDT to, in theory, assist with lymphatic drainage. Research supporting alternative treatments, like most lymphedema research, is limited by small sample size and reproducibility.

Lymphedema Case Examples in Sinonasal Malignancy

Case examples in Table 11.1 and Fig. 11.7 demonstrate the presentation, treatment, and outcomes of head and neck lymphedema in a heterogeneous population of patients with sinonasal malignancy.

Table 11.1 Case examples of lymphedema among patients with sinonasal malignancy

	Patient 1	Patient 2	Patient 3
Age, gender, race	51, M, AA	64, M, C	59, F, C
Disease stage, cell type, location	Unknown, squamous cell carcinoma, left maxillary sinus	T4 N2C M0 squamous cell carcinoma, nasal cavity	Multiply recurrent sarcomatoid left, maxillary sinus
Oncology treatment	Chemoradiation	Induction chemo, surgery, external beam radiation, surgery	4 partial resections; postoperative radiation; level1B recurrence— left neck dissection, postoperative radiation; persistent disease— immunotherapy, surgery
Lymphedema stage and location	Soft nonpitting Left upper/lower eyelid, left cheek, left jowl	Soft pitting bilateral: submental, anterior neck, midface, upper lip; lower lip, right intraoral cheek/cheek/jowl	Soft pitting bilateral: submental; left jowl, cheek, midface, lip, pre-auricular, submandibular, anterior+ lateral neck
Treatment recommendations	MLD; non-compliant Kinesio taping	MLD during radiation, CDT post radiation	CDT
Outcome	Discharge 14 weeks post-initial evaluation; 3.2% reduction in neck, 2.9% reduction face	2.7% increase neck during XRT; at discharge 18 month post-initial evaluation: 6% reduction neck; 2.5% face	3 months post CDT initiation: 4.1% reduction face; 3.6% reduction neck, active/ongoing CDT

Abbreviations: *AA* African America, *C* Caucasian, *CDT* complete decongestive therapy, *MLD* manual lymphatic drainage

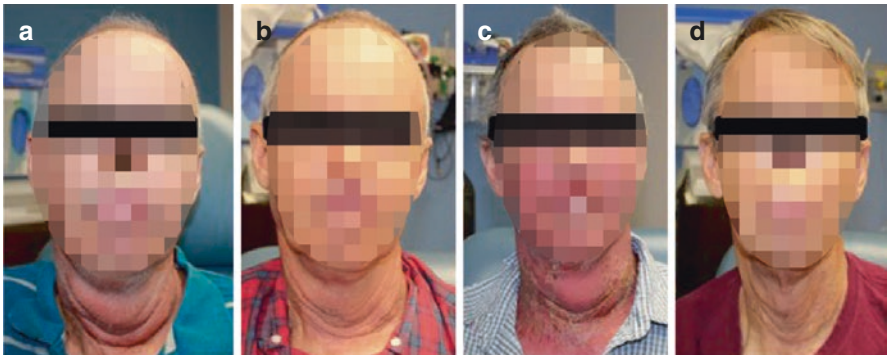

Fig. 11.7 Case example of head and neck lymphedema longitudinally through surgery and radiotherapy for sinonasal malignancy. Patient with a history of T4 N2C M0 squamous cell carcinoma of the nasal cavity presenting (**a**) postoperatively (**b**) 4 weeks post-complete decongestive therapy (CDT) program prior to external beam radiation. (**c**) Lymphedema increased during adjuvant radiotherapy, as evidenced by 2.7% increase in circumferential neck measurements. Lymphedema therapy continued during adjuvant radiation using manual lymphatic drainage and skin care only; compression was withheld during radiation. Patient resumed a full regimen of CDT 8 weeks post-radiation with the final panel (**d**) showing presentation at discharge from lymphedema therapy, 18 months post-radiation at which time he demonstrated 6.0% total reduction in neck measurements from initial evaluation

Summary

The goal of treatment is to maximize survival while minimizing patient morbidity. Given the proximity of skull base and sinonasal tumors to vital structures, a multidisciplinary team is necessary to maximize function for these patients. Supportive care is nuanced and challenging in this population. An multidisciplinary team of providers including speech language pathologists, oral oncologists, physical therapists, dietitians, social work, neuropsychologists, and other allied health-care providers are integral to provide critical care before, during, and after oncology intervention to improve functional outcomes. This involves a thorough evaluation, often including instrumental assessments to establish a baseline function, presurgical counseling from supportive care teams to educate patients on anticipated of deficits and recovery patterns, and therapeutic intervention as appropriate. Rehabilitation requires a team approach with a variety of expert clinicians to meet the complex needs of this population [52].

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