Carcinoma of the Larynx

Incidence

Cancer of the larynx represents less than 2% of all carcinomas. Squamous cell carcinomas comprise 95-98% of all malignant neoplasms of the larynx. In this regard the American Cancer Society estimates approximately 10,900 new patients with laryngeal cancer annually; with 9100 male and 1800 female new cases for 1982. At the time of diagnosis 62% will have local disease, 26% regional disease, and 8% distant disease. The estimated death rate is 3100 males and 600 females annually. There appears to be no racial tendency although the incidence is considerably higher in males (5:1), heavy smokers, and alcohol consumers. Epidermoid carcinoma of the larynx may be experimentally produced in hamsters exposed to chronic cigarette smoke inhalation.

Diagnosis

There are six methods of examining the larynx and no laryngeal examination, where disease is suspected, is complete unless five of these are used. They are: (1) palpation of the cervical soft tissues, laryngeal cartilage, and base of tongue; (2) indirect mirror examination of the larynx; (3) direct laryngoscopy and biopsy; (4) plain radiography of the neck and chest; (5) specialized radiologic tests - polytomography, xeroradiography, laminography, and computed axial tomographic (CAT) scanning of the larynx; and (6) laryngography - contrast study of the larynx.

Clinical diagnosis usually is made by mirror examination. Laryngeal tomography may be helpful in assessing limited portions of the larynx such as the true cords and immediate subglottic space. Xeroradiography may provide similar limited information. When airway obstruction is threatened by tumor size, contrast laryngography, which provides more comprehensive examination, may be hazardous and therefore contraindicated. Nevertheless, when possible, laryngography may provide an excellent demonstration of difficult-to-examine areas such as the laryngeal surface of the epiglottis at the anterior commissure and subglottic tumor extension. During laryngography the modified Valsalva's maneuver will facilitate examination of the pyriform sinuses, whereas the Müller's maneuver (reverse EE) will aid in evaluating the ventricles of Morgagni by drawing the true cords subglottically, thus dilating the ventricular mucosa. The subglottic arches also may be evaluated by this technique which provides additional opportunity to assess vocal cord mobility on deep inspiration and phonation. Laryngography should generally precede direct laryngoscopy and biopsy since surgical defects or reactive edema produced by the latter render radiologic interpretation difficult if not impossible.
Symptoms

Major

1. Hoarseness: It is the cardinal symptom of laryngeal cancer. It is due to inappropriate vocal cord approximation, increased vocal cord bulk, alteration of vibrating properties of the vocal cords, or vocal cord paralysis. Paralysis usually is due to invasion of the vocalis muscles.

2. Dyspnea and stridor: It is a late symptom associated with airway obstruction. The stridor is generally inspiratory and expiratory.

3. Pain can be of two types. One is a vague scratchy sensation in the throat. The other is referred pain to the ipsilateral ear via the vagus nerve. The latter is more common with aryepiglottic fold and pyriform invasion.

4. Dysphagia or difficulty in swallowing is associated with extralaryngeal involvement usually at the base of the tongue, hypopharynx, and supraglottis.

5. Odynophagia or painful swallowing is associated with invasion of the extralaryngeal musculature.

6. Chronic cough and haemoptysis are signs of large fungating tumors, hypopharyngeal invasion, overflow of secretions, or superior laryngeal nerve paralysis.

Minor

1. Weight loss: Due to poor nutrition.

2. Halitosis: Due to necrosing tumor.


4. Laryngeal tenderness: Tumor necrosis or suppuration.

Signs

Signs of laryngeal carcinoma can be divided into two groups: (1) those which are found on neck examination, and (2) those which are seen by visualization of the larynx.

Neck Examination

These are nonspecific signs which occur late in the course of the disease:

1. Lump in neck - metastatic lymph node.

2. Broadening of the larynx on palpation denotes tumor extension to the thyroid cartilage and inner perichondrium and extensive invasion of the larynx.
3. Tenderness of larynx denotes tumor invasion of inner perichondrium and cartilage or laryngeal chondritis secondary to tumor invasion.


5. Fullness of cricothyroid membrane to palpation denotes subglottic and usually extralaryngeal tumor extension.

6. Fullness in the thyroid membrane implies tumor invasion of the preepiglottic space, base of the epiglottis, and probably extralaryngeal tumor extension.

7. Digital palpation of the base of the tongue may show induration and submucosal tumor extension into the vallecula and base of the tongue.

**Laryngeal Examination**

The gross appearance of squamous cell cancer of the larynx varies greatly with size and location.

1. Supraglottis: Tumors in this region are often fungating with heaped-up edges and multiple areas of central ulceration.

2. Glottis: The characteristic lesion has a whitish "cauliflower" appearance which is friable to palpation and is surrounded by an area of hyperemia. Glottic lesions tend to be proliferative rather than ulcerating.

3. Subglottis: Tumors in this area are more diffuse and have superficial ulceration.

4. Marginal lesions: Tumors in this area are proliferative, and tend to be fungating with smal central ulcerations and heaped-up margins.

5. Pyriform tumors: These are larger bulky tumors which are not ulcerated. They have heaped-up margins and appear dark red and fleshy.

**Anatomic Classification**

The larynx is divided into three regions and these may be subdivided into a number of sites as shown in Fig. 19-1.

1. Supraglottis: Tip of epiglottis including its free borders to and including the false cords, and laryngeal ventricles.

2. Glottis: Floor of ventricle including the true cord, 1 cm infraglottic from the edge of the true cord including the anterior and posterior commissure.
3. Infraglottic: Arise 10 mm below the free edge of the vocal cord to the inferior border of the cricoid cartilage.

4. Transglottic: Lesions that cross the ventricle or involve the larynx above and below the true cords, and extend 10 mm below the free edge of the true vocal cord.

**Associated Regions**

1. Marginal: Lesions involving the aryepiglottic folds.

2. Superior hypopharynx:
   
a. Vallecula: Vallecula surface of epiglottis, base of tongue posterior to circumvallate papilla and medial to glossoepiglottic folds.
   
b. Posterior pharyngeal wall: Zone between posterior projection of the tip of the epiglottis and inferior border of cricoid.

3. Inferior hypopharynx:
   
a. Pyriform sinus: An area bounded superiorly by the glossoepiglottic fold, inferiorly by the apex of the pyriform sinus, laterally by the thyroid cartilage, and medially by the aryepiglottic fold and arytenoid cartilage.
   
b. Postcricoid: Mucosal area covering the rostrum of the cricoid cartilage.

**The Biopsy**

One cubic centimeter of tissue contains $10^9$ cells. In general a carcinoma is detectable palpably when it attains a diameter of 1 cm. Therefore $10^9$ cells/cc must be malignant to allow gross detection. Histopathologic detection is possible when $10^6$ cells/cc (1:1000) are malignant. A lesser ratio of malignant to normal cells per cubic centimeter of tissue go entirely unnoticed histopathologically, emphasizing the occasional difficulty in obtaining histologic confirmation of cancer.

**Treatment Modalities**

Before deciding on the mode of treatment, it is well to note that, in good hands, the mortality for laryngectomy with radical neck dissection varies from 2-5%. The risk is higher after the patient has received radiotherapy. MacComb reported that the mortality of all composite resections about the head and neck is 4.4% without preoperative irradiation and 8.5% after preoperative irradiation. We do not intend to promote any specific mode of therapy. To date, surgery, radiotherapy, and combined therapy are the therapeutic modalities of choice.
TNM Classification and Staging - Definitions:

(American Joint Committee on Cancer Staging and End Results Reporting, 1980)

TNM Classification

Primary Tumor (T)

\( T_x \) Tumor that cannot be assessed by rules.
\( T_0 \) No evidence of primary tumor.

**Supraglottis**

\( T_{is} \) Carcinoma in situ.
\( T_1 \) Tumor confined to site of origin with normal mobility.
\( T_2 \) Tumor involving adjacent supraglottic site(s) or glottis without fixation.
\( T_3 \) Tumor limited to larynx with fixation and/or extension to involve postcricoid area, medial wall of pyriform sinus, or pre-epiglottic space.
\( T_4 \) Massive tumor extended beyond the larynx to involve oropharynx, soft tissues of neck, or destruction of thyroid cartilage.

**Glottis**

\( T_{is} \) Carcinoma in situ.
\( T_1 \) Tumor confined to vocal cord(s) with normal mobility (including involvement of anterior or posterior commissures).
\( T_2 \) Supraglottic and/or subglottic extension of tumor with normal or impaired cord mobility.
\( T_3 \) Tumor confined to the larynx with cord fixation.
\( T_4 \) Massive tumor with thyroid cartilage destruction and/or extension beyond the confines of the larynx.

**Subglottic**

\( T_{is} \) Carcinoma in situ.
\( T_1 \) Tumor confined to the subglottic region.
\( T_2 \) Tumor extension to vocal cords with normal or impaired cord mobility.
\( T_3 \) Tumor confined to larynx with cord fixation.
\( T_4 \) Massive tumor with cartilage destruction or extension beyond the confines of the larynx, or both.

**Nodal Involvement (N)**

\( N_x \) Nodes cannot be assessed.
\( N_0 \) No clinically positive node.
\( N_1 \) Single clinically positive homolateral node 3 cm or less in diameter.
\( N_2 \) Single clinically positive homolateral node more than 3 cm but not more than
6 cm in diameter, or multiple clinically positive homolateral nodes, none more than 6 cm in diameter.

N2a Single clinically positive homolateral node more than 3 cm but not more than 6 cm in diameter.
N2b Multiple clinically positive homolateral nodes, none more than 6 cm in diameter.
N3 Massive homolateral node(s), bilateral nodes, or contralateral node(s).

N3a Clinically positive homolateral node(s), one more than 6 cm in diameter.
N3b Bilateral clinically positive nodes (in this situation, each side of the neck should be staged separately; that is, N3b: right, N2a; left, N1).
N3c Contralateral clinically positive node(s) only.

Distant Metastasis (M)

Mx Not assessed.
M0 No (known) distant metastasis.
M1 Distant metastasis present Specify.

Stage Grouping

<table>
<thead>
<tr>
<th>Stage</th>
<th>T1N0, M0</th>
<th>T2N0, M0</th>
<th>T3N0, M0</th>
<th>T1-3N1, M0</th>
<th>T4N0, M0</th>
<th>Any T N2-3, M0</th>
<th>Any T, any N, M1</th>
</tr>
</thead>
</table>

Five-Year Survival by 1972 TNM Classification

The American Joint Committee for Cancer Staging published in July 1972 the following 5-year survival rates on 1632 laryngeal carcinoma, 1061 of which were of glottic origin, 552 supraglottic, 19 subglottic, and one of unknown site of origin (Table 19-1). It would appear that in large series, radiation produces comparable results to surgery when the tumor is small but appears at a disadvantage as tumor size increases. The pool of radiation successes in this combined series is even smaller than indicated when survival rates are corrected for surgical salvage following radiation failure.

Vermund's 1969 series is corrected for the 1972 re-classification. His 5-year survival statistics appear in Table 19.2.
Table 19-1. Five-Year Survival by 1972 TNM Classification

<table>
<thead>
<tr>
<th>N0 Glottic</th>
<th>N1a Supra Glottic</th>
<th>N1b Supra Glottic</th>
<th>N2b Glottic Supra</th>
<th>N3 Supra Glottic</th>
<th>M Glottic Supra</th>
</tr>
</thead>
<tbody>
<tr>
<td>T1a</td>
<td>94%</td>
<td>61%</td>
<td>65%</td>
<td>100%</td>
<td>45%</td>
</tr>
<tr>
<td>T1b</td>
<td>93%</td>
<td>62%</td>
<td>45%</td>
<td>(no case)</td>
<td>20%</td>
</tr>
<tr>
<td>T2</td>
<td>85%</td>
<td>53%</td>
<td>36%</td>
<td>0%</td>
<td>20%</td>
</tr>
<tr>
<td>T3</td>
<td>65%</td>
<td>40%</td>
<td>40%</td>
<td>10%</td>
<td>8%</td>
</tr>
<tr>
<td>T4</td>
<td>40%</td>
<td>37%</td>
<td>40%</td>
<td>10%</td>
<td>8%</td>
</tr>
</tbody>
</table>

Table 19-2. Laryngeal Carcinoma Five-Year Survival in Relation to Treatment Modalities

<table>
<thead>
<tr>
<th>Tumor</th>
<th>Radiation and Surgical Salvage</th>
<th>Surgery</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Glottic</td>
<td></td>
</tr>
<tr>
<td>T1</td>
<td>86%</td>
<td>65%</td>
</tr>
<tr>
<td>T2</td>
<td>55</td>
<td>69%</td>
</tr>
<tr>
<td>T3</td>
<td>29%</td>
<td>55%</td>
</tr>
<tr>
<td>T4</td>
<td>14%</td>
<td>35%</td>
</tr>
<tr>
<td></td>
<td>Supraglottic</td>
<td></td>
</tr>
<tr>
<td>T1</td>
<td>73%</td>
<td>71%</td>
</tr>
<tr>
<td>T2</td>
<td>44%</td>
<td>62%</td>
</tr>
<tr>
<td>T3</td>
<td>29%</td>
<td>55%</td>
</tr>
<tr>
<td>T4</td>
<td>10%</td>
<td>56%</td>
</tr>
<tr>
<td></td>
<td>Subglottic</td>
<td></td>
</tr>
<tr>
<td>T1-4</td>
<td>36%</td>
<td>42%</td>
</tr>
</tbody>
</table>

Conservation Surgery

1. According to Som the indication for horizontal supraglottic laryngectomy is a tumor limited to the supraglottic region 3-5 mm from the anterior commissure with normal vocal cord mobility. Contraindications are involvement of the vallecula, arytenoid, or pyriform fossa. On the other hand, the criteria proposed by Sisson include:

   a. A margin of 5 mm must exist between the inferior border of the tumor and the anterior commissure.
b. The true vocal cords must be mobile.

c. Only one arytenoid can be removed.

d. There is no clinical or x-ray evidence of extension into the thyroid cartilage. (Theoretically, supraglottic lesions should not involve the thyroid cartilage).

e. There is no evidence of anterior neck invasion. Induration and the presence of enlarged nodes in the suprahysoid space or the thyrohyoid membrane are contraindications to conservation surgery.

f. In the case of tongue lesions there must not be fixation of the tongue or extension on the lingual surface to within 5 mm of the circumvallate papillae or foramen caecum.

g. There must be no extension to either the postcricoid or the interarytenoid space. Prevertebral fascia fixation is a contraindication.

h. Laryngograms and direct endoscopy must prove the apex of the pyriform sinus to be free of disease.

i. Generally, a lesion over 3 cm in diameter or fixed cervical nodes are contraindications to partial resection.

j. Relatively normal pulmonary function.

k. Under age 60-65.

2. Indications for vertical hemilaryngectomy in non-radiated tumors of the true cord are: (a) at least one vocal cord or arytenoid must be mobile; (b) less than 30% involvement of the anterior contralateral cord. Contraindications are: (a) superior involvement of the ventricle or false cord; (b) subglottic extension greater than 10 mm anteriorly. Vertical hemilaryngectomy may be used successfully in salvage of radiation failures of tumors involving the membranous true cord providing strict criteria are met. Contraindications to this procedure in radiated tumors of the true cord are therefore: (a) any contralateral cord involvement; (b) greater than 10 mm anterior subglottic extension; (c) bilateral cord fixation.

3. Indications for anterior commissure technique are: (a) horseshoe lesions of the membranous vocal cords crossing the anterior commissure; (b) less than 10 mm anterior subglottic extension; (c) no arytenoid involvement; (d) normal vocal mobility of at least one vocal cord or arytenoid.

**Discussion of Specialized Areas**

**Glottis**

Anterior Commissure. Anterior commissure lesions per se are rare and should not be categorized with cordal tumors that cross the midline anterior commissure. Radiation therapy provides 80% survival rates or 40% 5-year cure rates according to Kirchner who therefore
concludes radiation to be a relatively ineffective treatment modality in this situation. Som provides a 68% surgical cure rate by anterior commissure technique and 81% overall cure rate produced by further salvage surgery.

Ogura noted that 1% of glottic lesions are pure anterior commissure lesions and 30% are glottic tumors which extend into the anterior commissure. Thirty-one percent had greater than 5 mm subglottic extension and 8% had positive lymph node metastasis. The absolute overall survival was 74% for conservation surgery, 62% for total laryngectomy, and 61% for radiation. Survival by stage was stage I, 83%; stage II, 74%; and stage III and IV, 62%. Most of the recurrences (34%) occurred in the neck. Local recurrences was 1%.

Hemilaryngectomy. The overall survival for patients treated with hemilaryngectomy is 91% (primary cure and salvage). The survival by stage is as follows: stage I, 87%; stage II, 82%; stage III, 74%. The salvage rate by surgery and irradiation is 74%. In the most recent series of 111 cases, the survival is 90% for primary surgery, 64% salvage rate, and overall survival is 96%.

Tumor extension into the arytenoid area on one side did not affect survival. In 79 cases the overall cure rate is 90%. Glottic carcinoma with subglottic extension occurs in 20% of the cases. The survival depends upon the length of the subglottic extension. Stage II lesions with less than 10 mm extension have a cure rate of 92%. Stage II and III lesions with more than 10 mm extension have a cure rate of 69% and 50% respectively. The overall cure rate for surgery is 83% and for radiation 63%.

Total Laryngectomy. The overall survival for the combined total laryngectomy and irradiation group is 70%: with stage I, 95%; stage II, 69%; stage III, 68%; stage IV, 45%. The salvage rate for irradiation is 51% for an overall cure rate of 80%.

Radiation for Early Lesions. The use of radiotherapy has the following cure rates shown in Table 19-3.

Table 19-3. Reported Cure Rates Following Radiotherapy for Laryngeal Carcinoma

<table>
<thead>
<tr>
<th>Study</th>
<th>T1</th>
<th>T2</th>
<th>Overall</th>
<th>Primary Therapy and Salvage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Perez et al</td>
<td>82%</td>
<td>77%</td>
<td>62%</td>
<td></td>
</tr>
<tr>
<td>Horiot et al</td>
<td>90.5%</td>
<td>86%</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Goepfert et al</td>
<td>88%</td>
<td>74%</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Wang et al</td>
<td>90%</td>
<td>80%</td>
<td></td>
<td></td>
</tr>
<tr>
<td>(membranous cord 92%, anterior commissure 82%, arytenoid 76%, vc fixation 63%)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Stewart et al</td>
<td>95%</td>
<td>72%</td>
<td>(T3 50%, T4 25%)</td>
<td></td>
</tr>
<tr>
<td>Hibbs et al</td>
<td>-</td>
<td>76%</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Boles et al</td>
<td>-</td>
<td>74%</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Lederman et al</td>
<td>-</td>
<td>65%</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Carcinoma in Situ. According to A. Miller carcinoma in situ carries a 15% probability of invasive change within 3-8 months. Vocal cord stripping, the initial treatment of choice, provides 75% cure rate. Hemilaryngectomy is recommended for recurrent carcinoma in situ.
Irradiation as the initial treatment is contraindicated because it carries the highest rate of recurrence and invasion.

Verrucous Carcinoma. This carcinoma is characterized by an exophytic warty appearance, slow growth, local invasion, scarcity of metastases, and a benign histology. Biller et al reported 15 cases of this variant of squamous cell carcinoma. They advocated adequate but conservative surgical excision as opposed to radical radiation therapy.

Supraglottic Lesions

A 3-year determinate cure rate for supraglottic tumors is: stage I, 82.5%; stage II, 79%; stage III, 69%; stage IV, 50%. The overall cure rate is 76%. The salvage rate by other modalities is 47%. The overall survival is 82% for conservation surgery.

The 3-year determinate survival for large T3 and T4 lesions with combined total laryngectomy and neck dissection and radiation is 66%. Those treated by irradiation alone is 33%. The 5-year survival after total laryngectomy only is 45-60%, and full-course radiation only is 7-56% with an average of 20% on review of the literature.

Transglottic Carcinoma

According to Wang transglottic carcinoma carries a 24% 5-year survival by radiation therapy alone and 53% 5-year survival rate when treated by total laryngectomy.

Aryepiglottic Fold Carcinoma

The overall 3-year determinate survival is 53%. The survival by stage is: stage I, 66%; stage II, 50%; stage III, 52%; stage IV, 28%. Smaller lesions treated by partial laryngopharyngectomy had a 59% and larger lesions 27% 3-year survival.

Pyriform Sinus Carcinoma

This carcinoma is rightfully a hypopharyngeal lesion but so often invades the laryngeal framework that its respiratory symptoms, hoarseness, aspiration, and pain justify its brief discussion in this chapter. The 3-year overall cure rate is 40%. When metastases are present the cure rate drops to 34%. If there are no positive lymph nodes the cure rate is 48%. With bilateral neck metastasis, the cure rate is 17%. The 3-year cure rate by stage is as follows: stage I, 66%; stage II, 65%; stage III, 40%; stage IV, 30%. The cure rate by irradiation is 40%. The main cause for therapeutic failure is persistent neck disease. Once lymph node metastases are present the cure rate drops to below 35%.

Treatment of Nodal Metastasis

There is little disagreement that radical neck dissection is of value in the treatment of the clinically palpable node. However, elective neck dissection remains a controversial issue. The procedure is generally justified when the incidence of occult metastasis for any given tumor approaches 25%. The kind of data that may be helpful in the preoperative decision making process is therefore based on histologic examination of neck specimens in large series.
**Incidence of Nodal Metastasis**

A. Norris

<table>
<thead>
<tr>
<th>Location</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Epiglottic</td>
<td>42%</td>
</tr>
<tr>
<td>False cord</td>
<td>29%</td>
</tr>
<tr>
<td>True cord</td>
<td>6%</td>
</tr>
<tr>
<td>Subglottic</td>
<td>16%</td>
</tr>
</tbody>
</table>

B. McGavran

<table>
<thead>
<tr>
<th>Location</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Supraglottic</td>
<td>33%</td>
</tr>
<tr>
<td>Transglottic</td>
<td>52%</td>
</tr>
<tr>
<td>Subglottic</td>
<td>19%</td>
</tr>
</tbody>
</table>

On the other hand, others might argue that there is in fact no statistical evidence for improved survival rates by elective neck dissection over that performed subsequent to the emergence of a palpable node. Furthermore, there is growing evidence that radiotherapy alone may be effective in controlling occult regional metastases. Bagshaw suggests a 27% 5-year control rate for N2, N3 adenopathy and in this regard a 95% 5-year control rate for occult metastasis.

Node fixation is universally an ominous sign. Here radical neck dissection according to Santos provides no substantial role in improving survival in such patients. The effectiveness of standard radical neck dissection is equally questionable when node involvement occurs in the posterior cervical triangle or along the paratracheal chain, the latter notably resulting from a subglottic primary. In this instance Harrison recommends thyroid lobectomy and superior mediastinal dissection of the paratracheal lymphatics.

**Neoplasms of the Ear**

The majority of ear malignancies involve the auricle (85%) while 10% of such malignancies involve the external auditory canal and only 5% involve the middle ear and mastoid.

**Symptoms and Signs**

The earliest symptoms of carcinoma of the external auditory canal is otorrhea with pain (50%). The pain is intense and is out of proportion to the pathologic and clinical findings. Other symptoms include bleeding, fullness in the ear, and a conductive hearing loss (29%). The major findings include a vascular friable polyp or ulceration of the posterior and inferior wall of the external auditory canal. Later symptoms include perceptive deafness (15%), vertigo (13%), and facial nerve paralysis (13-35%). The incidence of metastasis at initial presentation is 13-30%.
External Ear

Most tumors of the external ear are squamous cell carcinoma. The incidence is one case in 16,000 ear complaints. In Conley's series, 62% were squamous cell and 31% were basal cell epitheliomas. The treatment for basal cell carcinoma of the auricle is wide excision. Miller advocates en bloc resection of most of the auricle with the underlying mastoid cortex when cancer (basal cell or squamous cell) lies within 1 cm of the external meatus. Squamous cell carcinoma of the helix can be resected widely and primarily reconstructed. Within the canal, the posterior canal wall near the annulus is a common site of malignancy. The lymphatics of this area drain in the direction of the preauricular, mastoid, subparotid, and subdigastric nodes. Lesions of the external auditory canal, particularly of the bony canal, should be treated with "superficial" temporal bone resection leaving intact the facial nerve and the labyrinth. The prognosis for helical and antihelical lesions (squamous cell as well as basal cell) is 90-95% 5-year survival. Lesions near the concha and those of the external auditory canal have a 5-year survival rate of about 30-40%. Lesions that invade the middle ear or produce facial nerve paralysis have a cure rate of less than 25%. Lesions of the anterior canal may metastasize to the pretragal or parotid nodes through the fissures of Santorini. In general, surgical resection provides a better prognosis than radiotherapy. En bloc radical neck dissection is not necessary in the absence of clinically palpable nodes. If the parotid region is involved, en bloc parotidectomy is to be performed.

Middle Ear Mastoid

The most common tumor of the middle ear is squamous cell carcinoma while glomus jugulare tumor ranks second in prevalence. The most common sarcoma of the middle ear is embryonal rhabdomyosarcoma. Adenoidcystic carcinoma is rare. It is derived from the ceruminous gland duct epithelium of the posterior meatus.

Symptoms of Malignancy

1. Deep, unremitting pain due to invasion of bone by the tumor.

2. Bleeding from the external auditory canal may be the first and only symptom of the disease.

3. Hearing loss may be of the conductive type if the tumor obstructs the external auditory canal or destroys the middle ear transformer mechanism. Perceptive hearing loss may result if the inner ear is progressively involved.

4. Vertigo may indicate inner ear destruction.

5. Facial nerve paralysis may occur when disease extends medially to involve the middle and inner ear.

6. Therefore, the only symptom that differentiates carcinoma from chronic otitis media or cholesteatoma is pain, and its presence should alert the physician to possible malignancy.
Diagnosis

Routine mastoid and skull films show bone destruction in 40% of the patients. Anteroposterior and lateral polytomography are helpful in delineating the extent of bone erosion. Carotid angiography is essential when dural involvement is suspected.

Treatment

Early attempts to treat cancer of the temporal bone consisted of radical mastoidectomy followed by radiation therapy which resulted in a 5-year cure rate of less than 25%. Osteoradionecrosis and radiation injury to the brain stem often brought devastating sequelae. Temporal bone resection has increased the 5-year cure rate to 36-44%. Preoperative radiation should be used cautiously especially when dural involvement is present since the incidence of CSF leaks and meningitis greatly increases when dural grafting follows radiation therapy. In this regard, when the dura is involved with carcinoma, radiation used postoperatively avoids life-threatening complications of the dural graft repair.

Lesions involving the middle ear therefore call for subtotal temporal bone resection, a method which Montgomery beautifully describes. Hilding in 1967 described an approach for total resection of the temporal bone. In this regard the facial nerve is sacrificed, a total parotidectomy is performed, and a facial to hypoglossal anastomosis is recommended. Radical neck dissection is recommended when clinical adenopathy is present.

Carcinoma of the Paranasal Sinuses

Cancer of the paranasal sinuses is relatively rare. According to Martin malignancies here represent 0.2% of all cancer and 3% of cancer of the upper respiratory tract. Carcinoma of the maxillary sinus constitutes 80% of sinus malignancies and has been linked to previous diagnostic introduction of thorium oxide (Thorotrast) contrast medium into the sinus cavities.

Maxillary Sinus

This is the largest paranasal sinus. Its boundaries are:

1. Medial wall: Nose.
3. Anterior: Face.
5. Posteromedial: Pterygopalatine fossa.
6. Roof: Orbit.
7. Floor: Alveolar process of the maxilla which holds the three molars and the second premolar. In the adult, the floor of the maxillary sinus is lower than that of the nose, whereas
in a child the reverse is true.

Tabb and Barranco reported a male/female ratio of 2:1 with an age distribution greatest in the sixth and seventh decades of life. No racial tendency is reported.

The earliest sign of cancer involving the antrum of Hihgmore is unilateral nasal obstruction. This is associated with ipsilateral nasal discharge which may be purulent. The next most common symptom is cheek or palate swelling. Nasal bleeding is less commonly encountered. Hypesthesia in the distribution of the infraorbital nerve is an ominous sign which indicates a high and often posterior lesion. Trismus is a sign of pterygoid involvement and ophthalmoplegia points to orbital or intracranial extension.

Most topographic classifications of tumors of the maxillary sinus are based on Ohngren's line, an imaginary line drawn diagonally through the maxillary sinus seen on lateral projection from the inner canthus to the mandibular angle. Tumors anterior to this line share a better prognosis (71% 3-year survival) as opposed to a tumor posterior to this line (28% 3-year survival). It is an established fact that tumors lying posteriorly are much more lethal by virtue of their ability to gain access to the anterior cranial fossa through the cribriform plate and into the middle cranial fossa via the foramen rotundum. The AJCC (1980) has staged and classified the maxillary sinus tumors in the following way:

The TNM Classification and Staging: Definitions

(American Joint Committee on Cancer Staging and End Results Reporting 1980)

TNM Classification

Primary Tumor (T)

T_x Tumor that cannot be assessed by rules.
T_0 No evidence of primary tumor.
T_1 Tumor confined to the antral mucosa of the infrastructure with no bone erosion or destruction.
T_2 Tumor confined to the suprastructure mucosa without bone destruction, or to the infrastructure with destruction of medial or inferior bony walls only.
T_3 More extensive tumor invading skin of cheek, orbit, anterior ethmoid sinuses, or pterygoid muscle.
T_4 Massive tumor with invasion of cribriform plate, posterior ethmoids, sphenoid, nasopharynx, pterygoid plates, or base of skull.

Nodal Involvement (N)

N_x Nodes cannot be assessed.
N_0 No clinically positive node.
N_1 Single clinically positive homolateral node 3 cm or less in diameter.
N_2 Single clinically positive homolateral node more than 3 cm but not more than 6 cm in diameter or multiple clinically positive homolateral nodes, none more than 6 cm in diameter.
N2a  Single clinically positive homolateral node more than 3 cm but not more than 6 cm in diameter.
N2b  Multiple clinically positive homolateral nodes, none more than 6 cm in diameter.
N3   Massive homolateral node(s), bilateral nodes, or contralateral node(s).
N3a  Clinically positive homolateral node(s), one more than 6 cm in diameter.
N3b  Bilateral clinically positive nodes (in this situation, each side of the neck should be staged separately; that is, N3b; right, N2a; left, N1).
N3c  Contralateral clinically positive node(s) only.

**Distant Metastasis (M)**

Mx  Not assessed.
M0  No (known) distant metastasis.
M1  Distant metastasis present.
    Specify.

**Stage Grouping**

- Stage I  T1N0; M0
- Stage II T2N0; M0
- Stage III T3N0; M0
- Stage IV T4N0 or N1; M0
  Any TN2 or N3; M0
  Any T, any N; M1.

The treatment of choice for maxillary sinus malignancy is en bloc resection of the maxilla including the ethmoid sinus and lateral nasal wall. If indicated, the orbital contents, frontal sinus, and cribriform plate also are removed en bloc. Preoperative radiation improves the ability to encompass the tumor by sterilizing its margins and reducing mechanical spillage at the time of surgery. The average 5-year survival rate for maxillary carcinoma treated by radical surgery alone is therefore 62%.

In a recent series of 96 patients Lee and Ogura noted that 69% of patients had primary tumor control with combined radiation and surgery and 14% with radiation alone. The overall cure rate was 49%. Tumor recurrence was found in 20% of the cases in the neck, 16% at the primary site, and 8% at distant sites. Overall, the 5-year NED survival by stage was T1, 60%; T2, 45%; T3, 28%; and T4, 38%.

A review of the literature indicates that combined therapy gives an overall survival in maxillary cancer of 29-48% and radiotherapy 0-34% (see Table 19-4). Thus the former is the treatment of choice.
Table 19-4. Maxillary Sinus Carcinoma Survival

<table>
<thead>
<tr>
<th>Study</th>
<th>No of patients</th>
<th>Five-Year Overall (%)</th>
<th>Surgery and Radiation (%)</th>
<th>Radiation Alone (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Holsti</td>
<td>62/219</td>
<td>28</td>
<td>34</td>
<td>15</td>
</tr>
<tr>
<td>Boone</td>
<td>52/121</td>
<td>43</td>
<td>30</td>
<td>44</td>
</tr>
<tr>
<td>Badib</td>
<td>48/344</td>
<td>14</td>
<td>29</td>
<td>10</td>
</tr>
<tr>
<td>Hamberger</td>
<td>148/591</td>
<td>25</td>
<td>45</td>
<td>19</td>
</tr>
<tr>
<td>Lewis</td>
<td>191/677</td>
<td>28</td>
<td>33</td>
<td>20</td>
</tr>
<tr>
<td>Cheng</td>
<td>17/50</td>
<td>34</td>
<td>48</td>
<td>22</td>
</tr>
<tr>
<td>Lee and Ogura</td>
<td>44/96</td>
<td>49</td>
<td>69</td>
<td>14</td>
</tr>
</tbody>
</table>

The maxillary sinus drains into the retropharyngeal and parapharyngeal nodes. Hence, a routine elective neck dissection is of little benefit. However, if a patient presents with a palpable neck node, neck dissection should be performed although en bloc dissection is not possible.

The following factors should be considered contraindications to radical surgery of the maxillary sinus:

1. Destruction of the base of the skull.
2. Extension of cancer into the nasopharynx.
3. Inoperable regional metastasis.
5. Patient refusal to accept treatment.

**Frontal, Ethmoid, and Sphenoid Sinuses**

Primary carcinoma of these sinuses is extremely rare. Brownson and Ogura reported five cases of malignancy of the frontal sinus and reviewed the literature for another 28 cases. They concluded that epidermoid carcinoma is the most common form of malignancy in the frontal sinus. The most frequent presenting symptoms are swelling, pain, and proptosis. The prognosis is extremely poor. The surgical approach described in their paper recommended that the dura should be sacrificed when the posterior wall is eroded. The ethmoid sinuses should be resected en bloc. The orbit and the contralateral frontal sinus are to be sacrificed when in doubt as to the margin of invasion.

Carcinoma of the ethmoid has a similarly discouraging prognosis. It is interesting to note that ethmoidal carcinoma is unusually common among woodworkers. Ketcham has recently reported a series of 54 patients who underwent surgical removal of ethmoid sinuses by a combined intracranial transfacial approach resulting in a 56% 5-year survival rate. Advantages of this combined approach were: (1) intracranial tumor extension could be established with certainty; (2) the brain could be adequately protected during tumor
mobilization; (3) en bloc removal was possible; (4) CSF fistula could be avoided. Preoperative radiation was not used as an adjuvant to surgical resection because of greatly increased morbidity associated with failures of dural grafts and subsequent cerebrospinal fluid leaks and meningitis. Bridger noted cures in nine of 15 patients after this approach.

Carcinoma of the sphenoid sinus is very rare with unfavourable prognosis. Most tumors invade the sphenoid from the nasopharynx, posterior ethmoid air cells, or via the periorbital tissues. On rare occasions nasal tumors may erode into the sphenoid sinus. Because of the inaccessible anatomic location, complete resection is impossible. Consequently, the treatment of choice is radiotherapy. Clinically palpable nodes can be treated with radiation or radical neck dissection.