THE MANAGEMENT OF SUPERIOR SULCUS TUMORS

Ritsuko KOMAKI*1, James D COX*1, Joe B PUTNAM, Jr*2, Garrett WALSH*2, Ara VAPORCIYAN*2, Zhongxing LIAO*1, Craig STEVENS*1, Frank V FOSSELLA*3, Jin S LEE*3, Waun K HONG*1, Jack ROTIT*2

Abstract: Superior sulcus tumors are a rare type of lung cancer arising in the apex of the lung above the sulcus and cause specific symptoms and signs depending on the location and whether the tumor extends into the surrounding structures. Because of the closeness of critical structures to the tumor (e.g., the subclavian artery for anterior lesions, the brachial plexus for lesions in the middle location, and the sympathetic stellate ganglion causing Horner's syndrome [Pancoast's tumor], the vertebral bodies, nerve foramen, and spinal cord for posterior lesions), superior sulcus tumors were often considered marginally resectable or unresectable. Therefore, for many years, preoperative radiation therapy was considered routine treatment for those tumors. However, with the evolution in our understanding of these tumors and modern imaging techniques such as computerized tomography (CT) and magnetic resonant imaging (MRI) and surgical techniques, there is now considerable debate about the roles and timing of surgical resection, radiation therapy, and chemotherapy in the treatment of patients with these tumors. If mediastinoscopy reveals microscopic mediastinal lymph node involvement, the patient can be treated with preoperative concurrent chemoradiotherapy followed by surgery. If there was a gross mediastinal lymph node involvement (N2) on CT, N3 or T4 lesions, the patient can be treated with concurrent chemoradiotherapy with a curative intent; the outcome of such treatment appears to be better than that of sequential chemotherapy followed by radiation therapy. Whenever possible, without compromising the patient's quality of life, surgery should be considered to improve outcome.

Key words: Multidisciplinary management, Lung, Superior sulcus tumors

INTRODUCTION

Superior sulcus tumors were first described in the medical literature in 1838. In that year, Edwin Hare1 documented the case of a patient who presented with a history of pain, tingling, and numbness in the distribution of the left ulnar nerve, Horner's syndrome, and a palpable mass in the "inferior triangular space" on the left side of his neck. The mass continued to grow until the patient became paraplegic, developed urinary retention, and eventually died of the disease. The postmortem examination revealed a hard tumor extending superiorly toward the origin of the brachial plexus and involving the carotid artery, the cervical sympathetic nerves, the vagus and phrenic nerves, the spine, and intervertebral foramina. In 1932, Pancoast defined the superior sulcus tumors as bronchogenic carcinomas that developed in the apex of the lungs and invade the superior pulmonary sulcus2-4. They are situated in the thoracic inlet and invade the lymphatics of the endothoracic fascia and extend to the lower roots of the brachial plexus, the intercostal nerves, the stellate ganglion, the sympathetic chain, and adjacent ribs and vertebral bodies. The resulting severe pain and Horner's syndrome (pupillary constriction, ptosis of the upper eyelid, slight elevation of lower lid, sinking in of the eyeball, narrowing of the palpable fissure, anhidrosis and flushing of the affected side of the face) have been given the name "Pancoast's syndrome" because of his description as mentioned above.

Initially, only radiation treatment was attempted since these tumors were thought to be unresectable. The first successful removal of superior sulcus tumor was performed in 1950 by Chardack and MacCallum5. In brief, they resected the upper lobe en bloc along with the first and second ribs and roots C7, C8, and T1; they followed this with postoperative radiation therapy to eradicate microscopic foci of the residual tumor. One of their patients treated in this way lived for 5 years and 10 months after treatment, and postmortem examination revealed no evidence of recurrence or metastasis6.

Shaw and colleagues treated7 superior sulcus tumors in 18 patients by preoperative radiation therapy followed by en bloc surgical resection 4-6 weeks later. Although not randomized, this study demonstrated a high resectability rate and good palliation, which popularized preoperative radiation therapy for superior sulcus tumors. However, this approach masked the exact local extension of the tumor as well as nodal metastases that might otherwise have been successfully treated by resection first and definitive radiation therapy second. Recent improvements in diagnostic tools (e.g., CT, MRI, and mediastinoscopy) have made possible more accurate staging of local extension and nodal disease and now allow better treatment selection8.

Published reports9 have noted 5-year survival rates of 15% to 56% for patients receiving combinations of radiation therapy and surgical treatment. In some of these studies, patients received additional postoperative radiation therapy. Few studies, however, have had adequate numbers of patients to allow examination of the effects of distinct treatments such as

1 Departments of Radiation Oncology, The University of Texas M. D. Anderson Cancer Center (1515, Holcombe Blvd., Box 97, Houston, TX 77030)
2 Thoracic and Cardiovascular Surgery, The University of Texas M. D. Anderson Cancer Center
3 Thoracic and Head and Neck Medical Oncology, The University of Texas M. D. Anderson Cancer Center
as varying doses of radiation with surgery, radiation therapy with chemotherapy, or radiation therapy alone. A further difficulty is that some studies have identified univariate predictors of survival\textsuperscript{10-15}, but only one identified multivariate (independent) predictors of survival\textsuperscript{11}. None has examined independent predictors of survival by stage of disease. The identification of predictors of survival along the continuum of care could reveal subgroups of patients who require new combined treatment strategies.

**DIAGNOSIS**

Diagnosis of superior sulcus requires a physical examination followed by radiographic examination, including CT and MRI, histologic confirmation, and cytologic confirmation.

1. **Symptoms**

Superior sulcus tumors may occur in three locations, which are shown in Figures 3, 4 and 5. Symptoms and signs will be related to the location of the tumor: anterior, where the tumor invades major blood vessels such as the subclavian artery; middle, where it mainly invades the brachial plexus; and posterior, where it invades satellite ganglia.

The pain due to superior sulcus tumor usually occurs in the shoulder and along the vertebral body of the scapula. It is due to tumor localized at the apex of the lung and involving parietal pleura. Subsequently, the pain extends down the ulnar distribution of the ipsilateral arm to the elbow (which indicates involvement of T1) and eventually to the ulnar surface of the forearm and to the fourth and fifth fingers of the involved side of the hand (which indicate ulnar nerve involvement). Once the sympathetic chain and the stellate ganglia are involved by the tumor, patients usually develop Horner’s syndrome as described above. The direct extension into the first or second rib or vertebral bodies usually causes severe pain, and if the spinal canal or cord is involved, patients might be paralyzed. Patients usually do not show the pulmonary symptoms of hemoptysis, shortness of breath, and cough usually associated with endobronchial lesions.

In a series reported by Komaki and Colleagues\textsuperscript{16}, most patients were treated for osteoarthritis or bursitis of the shoulder with nonsteroidal analgesics for an average of 5 to 7 months before the diagnosis. Physical examination of these patients revealed increased pain with abduction of the involved side of the arm, muscle weakness, atrophy of the muscle of the hand, loss of the triceps reflex, and Horner’s syndrome. Some patients complained of tenderness where the tumor had invaded into the ribs or vertebral body. In cases where the tumor extended to the supraclavicular lymph nodes directly or by metastasis, physical examination revealed palpable lymph nodes. In cases where the tumor invaded into the spinal canal, physical examination revealed paraplegia or paralysis. If the tumor had metastasized into the aorto-pulmonary (AP) window lymph nodes, patients presented with hoarseness due to recurrent laryngeal nerve compression or invasion. Occasionally, patients presented with superior vena caval obstruction when the mediastinal lymph nodes were involved. Other symptoms and signs are weight loss or fatigue.

2. **Radiographic Evaluation**

1) **Chest Radiographs**

The pitfalls of making a diagnosis of superior sulcus tumor were pointed out by Simon and his colleagues\textsuperscript{17}. The first is that on regular chest radiographs, a soft tissue mass without bony destruction can be missed or misinterpreted as pleural thickening. The shadow extending medially from the pleural thickening toward the hilum is a sign of more aggressive malignant tumor. The apical lordotic or slightly oblique views show the apical lesions much better. The second pitfall is that an apical lordotic view did not show bony destruction. Additional anteroposterior views of the lower cervical/upper dorsal spine or tomogram may be needed to show bony destruction.

2) **Computed Tomography**

CT scans are better than conventional radiographs at showing the relationship of superior sulcus tumors to the anterior structures (i.e., subclavian artery and vein and trachea) and posterior structures (i.e., chest wall, ribs and vertebral bodies). The information gathered by CT scan provides more information in regard to operability. CT of the thorax is important for staging purposes to show any mediastinal lymph node involvement, pleural effusion, as well as CT of the upper abdomen to reveal any adrenal gland, hepatic or renal metastasis\textsuperscript{18}.

3) **Magnetic Resonance Imaging**

Compared with CT scans, MRI scans more accurately
display the anatomy of superior sulcus tumors (63% versus 94%)\(^\text{19}\). A sagittal section obtained by MRI can show extension of a superior sulcus tumor to the posterior wall of the subclavian artery as well as to mediastinal lymph nodes (Fig. 1). An axial scan cannot do this as easily since MRI is not as accurate at demonstrating invasion of tumor into the foramen, spinal cord, and major blood vessels (Fig. 2). In any case, MRI is required to demonstrate resectability of the superior sulcus tumors. MRI of the brain is important for showing any micrometastasis; patients who have superior sulcus tumors often present with brain metastases because of the location of the tumor and the fact that more of these patients have adenocarcinoma or large cell carcinoma of the lung which are highly metastatic\(^\text{20}\). When combined, MRI and CT can reveal extension of the superior sulcus tumors into the great vessels at the thoracic inlet primarily subclavian artery and vein; involvement of the trachea or esophagus; invasion into the brachial plexus; invasion of the chest wall; invasion into the vertebral bodies, foramen, and spinal cord; mediastinal lymph node involvement by metastasis or direct extension; and extrathoracic metastasis into the brain and upper abdomen. Although the diagnosis of superior sulcus tumors by diagnostic imaging is accurate in more than 95% of cases\(^\text{21}\), cytological or histological confirmation of the definitive diagnosis is important since it might affect treatment. For example, one study found that 3% of superior sulcus tumors were undifferentiated small cell lung cancers, which can be treated with chemotherapy and radiation therapy alone\(^\text{22}\).

3. Bronchoscopy and Fine-Needle Aspiration

According to Hepper and colleagues\(^\text{23}\), accurate diagnosis of superior sulcus tumors by bronchoscopy occurs in only 16% of cases. In comparison, Attar and colleagues\(^\text{24}\) found that fiberoptic bronchoscopy was cytologically diagnostic in 13 of 43 patients (30%), and Miller and colleagues\(^\text{25}\) found sputum cytology and bronchoscopy diagnostic in 31% of their patients. Because of the location of superior sulcus tumors, needle biopsy and, more recently, fine needle aspiration (FNA) are practical approaches to a positive diagnosis. For instance, McGoon\(^\text{26}\) described a transcervical technique that took a supraclavicular approach lateral to the sternal head of the sternocleidomastoid muscle passing through the scalene fat pad. Siderys and Pittman\(^\text{27}\) reported the direct insertion of a Vim-Silverman needle through the second and third intercostal space into the posterior chest of a patient in prone position. Walls and colleagues\(^\text{28}\) reported 26 of 27 patients with superior sulcus tumors by CT-guided posterior FNA. However, accurate diagnostically CT-guided biopsies are relatively costly compared with fluoroscopically and ultrasonically guided biopsies\(^\text{29}\).

4. Staging

Staging is key to deciding on treatment of superior sulcus tumors. Paulson\(^\text{30}\) emphasized the importance of mediastinoscopy since his patients did very poorly if the mediastinal or hilar lymph nodes were involved: only 3 out 17 patients with hilar or mediastinal nodal involvement survived 1 year and none survived 2 years. In contrast 44% of the patients with no nodal involvement survived 4 years or longer. After preoperative radiation therapy and subsequent surgery, Paulson also advocated scalene node biopsy if the scalene nodes were palpable prior to initiation of treatment. Attar and colleagues\(^\text{31}\) affirmed that adequate preoperative assessment is extremely important since those patients who had positive mediastinal lymph nodes died shortly after surgery. In his Toronto experience, Ginsberg\(^\text{32}\) documented that stringent preoperative assessment of patients with superior sulcus tumor ruled out surgery in 62 of 72 patients. As these reports illustrate, it is important to document the exact location of the superior sulcus tumor as well as extent of the disease since location will influence the resectability of the tumor and the radiation therapy dose arrangement.

If resection is contraindicated, tumors can be treated with concurrent chemotheraphy and radiation therapy. Depending on the location of the tumor, radiation therapy can be given anteriorly or posteriorly. The contraindications of surgery include extensive invasion of the brachial plexus, subclavian artery; vertebral bodies and mediastinal involvement (particularly perinodal); venous obstruction, and distant metastases\(^\text{33}\). Superior sulcus tumors located more anteriorly tend to invade the subclavian artery (Fig. 3). Superior sulcus tumors located medially tend to involve the brachial plexus and extend more superiorly, thus contraindicating surgery or indicating sacrifice of the brachial plexus if necessary (Fig. 4). Superior sulcus tumors located more posteriorly (i.e., typical Pancoast's tumors), may produce Homer's syndrome due to the involvement of the stellate ganglia or vertebral bodies with or without extension of the tumor into the foramen (Fig. 5). If the superior sulcus tumor is more anteriorly located, an anterior boost of radiation can be given without compromising tumor control or damaging more normal tissue. If the tumor is located more posteriorly, a posterior boost can be arranged without subjecting the spinal cord or more anterior structures to high doses of radiation (Fig. 6).

Fig. 2 Axial scan showing invasion of the tumor into the foramen, spinal cord, and major blood vessels. Reprinted courtesy Komaki, et al: Semin Surg Oncol 18: 152-164, 2000.
TREATMENT AND RESULTS

1. Preoperative Radiation Therapy and Surgical Resection

The combination of preoperative radiation therapy and subsequent surgical resection to treat superior sulcus tumors was first reported by Shaw and his colleagues in 1961. Paulson used this approach and updated it on several occasions. In brief, the superior sulcus tumor, chest wall, and superior mediastinum were given, bilaterally, 300 cGy of radiation per day in 10 fractions over 12 days up to a total tumor dose of 3000 cGy. Three weeks after completion of radiation therapy, patients underwent en bloc resection of the tumor along with the involved chest wall including the entire first rib and posterior portion of the second and third ribs. In addition, the resection sometimes included part of the first three transverse processes of the thoracic vertebrae and nerve roots, the 8th cervical nerve root, the lower trunk of the brachial plexus, part of the stellate ganglion, and the dorsal sympathetic chain. The involved lung was resected by either lobectomy or segmental resection. The entire procedure was accompanied by dissection of the regional hilar and mediastinal lymph nodes. More recently, patients

![Fig. 3](image-url) More anteriorly located superior sulcus tumor invading the subclavian artery. Reprinted courtesy Komaki, et al: Semin Surg Oncol 18: 152-164, 2000.

![Fig. 5](image-url) Superior sulcus tumor located posteriorly (typical Pancoast's tumor). Reprinted courtesy Komaki, et al: Semin Surg Oncol 18: 152-164, 2000.

![Fig. 4](image-url) Medial location of the superior sulcus tumor involving the brachial plexus extending more superiorly where surgery might be contraindicated. Reprinted courtesy Komaki, et al: Semin Surg Oncol 18: 152-164, 2000.

![Fig. 6](image-url) If the tumor is located posteriorly, a posterior boost can be arranged without giving high dose to the spinal cord or more anterior structure. Reprinted courtesy Komaki, et al: Semin Surg Oncol 18: 152-164, 2000.
in our institution have had mediastinoscopy prior to this radical dissection. If they are found to have any positive contralateral mediastinal lymph nodes or high mediastinal lymph nodes, they usually receive chemotherapy and radiation therapy rather than radical surgery.

In a series reported by Paulson\(^3\)\(^3\), 131 patients were started on preoperative radiation therapy to be followed by en bloc surgical resection. Of these, 78 patients (60%) completed preoperative radiation therapy followed by radical dissection. The operative mortality rate was 2.6%, and the overall survival was 31% at 5 years, 26% at 10 years, and 22% at 15 years. Three of 17 patients who had involvement of either the hilar or mediastinal lymph nodes survived 1 year, and none who had positive lymph nodes survived beyond 2 years. Among the 61 patients who had no nodal involvement at the time of surgery, the 5-year survival was 44%, the 10-year survival was 33%, and the 15-year survival was 30%. Miller and his colleagues\(^3\)\(^3\) administered preoperative radiation therapy at doses of 2000-4000 cGy over time spans of 4 days to 4 weeks. Attar and his colleagues\(^3\)\(^4\) administered it at doses of 5500-6000 cGy preoperatively over 1 month. However, both groups noted a high rate of morbidity and mortality and so set the dose at 3000 cGy. Beyer and Weisenburger\(^3\)\(^3\) administered a preoperative dose of at least 55 cGy, which induced a better response and appeared to increase survival. Devine and associates\(^3\)\(^4\) used two different dose levels: 3000-3500 cGy over 2 weeks in 25 patients and a higher dose of 4500-5000 cGy over 5-5 1/2 weeks in 15 patients. They noted no difference in the resectability or survival between the two dose levels. Miller and colleagues\(^3\)\(^5\) reported on 26 patients who had preoperative radiation therapy followed by en bloc resection and noted a 5-year survival of 32%. Stanford and colleagues\(^3\)\(^5\) treated a similarly treated group and noted a 5-year survival rate of 49.7%. Beyer and Weisenburger\(^3\)\(^3\) treated 28 patients with superior sulcus tumor. In 15 patients treated with preoperative radiation therapy, the 5-year survival was 45%. In 13 patients treated with radiation therapy alone, the 5-year survival was much worse. The overall 5-year survival was 22%. Ginsberg\(^3\)\(^1\) reported on 72 patients with Pancoast's tumor: 50 of them had tumors deemed inoperable after screening, and only 10 patients underwent preoperative radiation therapy (3000 cGy over 2 weeks) followed by resection. The 2-year survival of those 10 patients was 40%.

Martini\(^3\)\(^6\) reported on 145 patients treated at the Memorial Sloan-Kettering Cancer Center in New York City over a 36-year period. Sixty-eight out of 148 patients had surgery alone, and 48 patients had preoperative radiation followed by resection. Only 9% of the 68 patients who had surgery alone had completely resected tumors, compared to 23 patients who received preoperative radiation followed by complete resection. Martini also claimed that, without CT or MRI, it was difficult to evaluate the resectability during that time. Devine and his colleagues\(^3\)\(^4\) reported on a series of patients who completed preoperative radiation therapy and noted a 2-year survival of 29% and a 5-year survival of 14%.

Overall, patients with stage IIB tumors had a 5-year survival of 47%. However, patients who had surgical resection followed by radiation therapy had a 5-year survival of 56%, and patients who received both preoperative and postoperative radiation therapy with chemotherapy had a 5-year survival of 87% (although there were only 8 patients among this group). Surgery was an important prognostic factor in stage IIB patients. In comparison, patients with stage IIIA and stage IIIB tumors had 5-year survivals of 14% and 16%, respectively. Surgical resection was not an important prognostic factor in these patients since all had adequate CT and MRI studies of the chest as well as a metastatic work-up and mediastinoscopy before initiation of treatment.

2. Preoperative and Postoperative Radiation Therapy

Shahian and his colleagues\(^3\)\(^7\) reported on 18 patients who were treated with preoperative radiation therapy followed by resection (14 of these patients also underwent supplemental postoperative radiation therapy to treat positive lymph nodes or positive margins at the resection site or both). The overall 5-year survival for the entire series was 56%. Shahian and colleagues\(^3\)\(^8\) believed that postoperative radiation therapy, in addition to preoperative radiation therapy for patients with unfavorable findings such as positive lymph nodes or positive margins, would improve the survival. Martini and his group\(^3\)\(^9\) found that, if a tumor was not resectable at the time of surgery after preoperative radiation therapy, then a combination of radon seed implants and external radiation therapy (40 Gy in 4 weeks) would provide better local control and a survival rate comparable to that for external radiation therapy alone. The median survivals were 12 months versus 6 months, respectively.

At M. D. Anderson Cancer Center\(^3\)\(^0\), 143 patients with superior sulcus tumors had single-modality or combined treatment. Those patients with T3N0M0 tumors who had preoperative and postoperative radiation therapy, with or without chemotherapy, did extremely well and had a 5-year survival of 87% (however, there were only 8 patients in this group).

3. Surgical Resection Alone

Patients with superior sulcus tumor are not usually treated with surgery alone. At M. D. Anderson\(^3\)\(^1\), only 5 of 143 patients (3%) with superior sulcus tumors had surgery alone. In studies by Martini\(^3\)\(^6\) and Hilaris\(^3\)\(^8\), 9% of the total 68 patients had resectable tumors as compared with 23% of the total 48 patients who had preoperative radiation therapy. However, some patients with well-localized disease and fairly small tumors can be treated with radical surgery that includes adequate margins. Nevertheless, this is not the usual protocol for treating superior sulcus tumors, which usually occur in patients who have been followed up after treatment for a primary in the head and neck or at any other site by routine chest x-ray or CT of the chest. Attar and colleagues\(^2\)\(^9\) reported a 3-year survival rate of 60% in 5 such patients, similar to results reported by Komaki and colleagues\(^2\)\(^2\).

4. Surgical Resection Followed by Postoperative Radiation Therapy

Postoperative radiation therapy does not improve survival in patients with carcinoma of the lung who had complete
surgical resection without gross or microscopic residual tumor or regional lymph node involvement including hilar or mediastinal lymph nodes. This applies to superior sulcus tumors as well, even though their resection may leave positive margins.

Marini reported on 170 patients with superior sulcus tumors treated at the Memorial Sloan-Kettering hospital from 1938 to 1978. Of these patients, 127 underwent surgery; although the staging work-up was not complete and inoperable patients were treated more palliatively, the 5-year survival rate was 17% for these patients versus 3.4% for the inoperable patients. Among the 127 patients who underwent surgery, 20 patients had curative surgery after preoperative radiation therapy; their 5-year survival rate was 29%. The remaining 107 patients who underwent surgery received postoperative brachytherapy; their 5-year survival was 14%.

A report from M. D. Anderson Cancer Center on 85 patients with superior sulcus tumors noted that combined surgery and radiation therapy was more effective controlling the tumors and improving survival than either modality alone. In that study, 43 patients were categorized as having stage IIIA disease and 42 patients as having stage IIIB disease (according to the old American Joint Committee on Cancer [AJCC] classification used in 1992). Surgery was a significant factor in improving local control and survival among the stage IIIA patients. Those patients who had stage IIIB unresectable non-small cell lung cancers arising from the superior sulcus were treated by chemotherapy and radiation therapy. The 2-year survival was 46.5% for patients with stage IIIA disease versus 21% for patients with stage IIIB disease. When surgery was included in the treatment, 52% of patients (13 of 25) lived longer than 2 years compared with 22% (13 of 60) when the lesion was unresectable.

At the University of Texas M. D. Anderson Cancer Center, we updated a retrospective study of 143 patients with superior sulcus tumors to identify outcome predictors for patients treated by a multidisciplinary approach between 1975 and 1994. In this study, we examined the 5-year survival rate by pretreatment tumor and patient characteristics and by the treatments received. Strict criteria were used to define superior sulcus tumors. Actuarial life-table analyses and Cox proportional hazard models were used to compare survival rates. Overall predictors of 5-year survival were weight loss (p < 0.01), supraclavicular fossa (p = 0.03) or vertebral body (p = 0.05) involvement, stage of the disease (p < 0.01), and surgical treatment (p < 0.01). Five-year survival for patients with stage IIIB disease was 47% compared to 14% for stage IIIA, and 16% for stage IIIB. For patients with stage IIIB disease, surgical treatment (p < 0.01) and weight loss were significant independent predictors of 5-year survival (p = 0.01). According to our recent retrospective study for patients with stage IIIB disease, the only independent predictor of survival was a right superior sulcus location, which was associated with a worse 5-year survival rate than that for patients with tumors in the left superior sulcus (p = 0.02). More patients with adenocarcinoma than with squamous cell tumors experienced cerebral metastases within 5 years (p < 0.01). Patients without residual disease after surgical resection who received postoperative radiation therapy with total doses of 55 to 64 Gy had a 5-year survival rate of 82% as compared with the 5-year survival rate of 56% in patients who received 50 to 54 Gy. Twenty-three patients survived for longer than 3 years. Of these, 4 patients (17%) received radiation therapy alone or in combination with chemotherapy without surgical resection. The other 19 patients (83%) had resection combined with radiation therapy and/or chemotherapy. Interestingly, there was no significant 5-year survival difference between stage IIIA (N2) and stage IIIB (T4 or N3) if patients had unresectable lesions. Our conclusions from this study confirmed the importance of the new AJCC staging system, separating T3N0M0 (stage IIB) from stage IIIA, since there was significant difference in the 5-year survival between patients with T3N0M0 and T1-3N2M1 (p < 0.01). This study suggests that surgery is an important component of the multidisciplinary approach to patients with superior sulcus tumors if their nodes were negative. Disease that is minimally invading surrounding normal structures can be resected followed by radiation therapy in doses of 55 to 64 Gy.

Darteville and his colleagues reported on a group of patients in which none received preoperative radiation therapy, 14% had surgery alone and 86% had postoperative radiation therapy. The 2-year survival was 50%; the 5-year survival was 31%. The median follow-up was 2.5 years. The surgical approach in this series was aggressive. Some patients underwent a large L-shaped anterior cervical incision for removal of the medial half of the clavicle, dissection or resection of the subclavian vein, division of the anterior scalenus muscle, and resection of the cervical portion of the phrenic nerve if it had been invaded by the tumor. The subclavian and vertebral arteries were exposed, the brachial plexus was dissected up to the spinal foramen, the invaded ribs were resected, and the chest wall and primary superior sulcus tumor were removed en bloc, either directly or by extension of the surgical incision into the deltoid pectoral group. This approach has been used occasionally at M. D. Anderson.

5. Inoperable Superior Sulcus Tumors

Patients whose superior sulcus tumors are considered to be medically inoperable or surgically resectable should be considered candidates for curative or palliative radiation therapy with or without chemotherapy. Morris and his colleague reported on 26 cases in which superior sulcus tumors were treated with high-dose radiation therapy (7000 cGy / 7 weeks or higher).

Ahmad and colleagues reported on 48 patients treated by radiation therapy alone using either cobalt-60 or cesium-137 teletherapy up to a total tumor dose of 5000-6000 cGy over 5-6 weeks. The actuarial 3-year survival rate was 28% and the 5-year survival rate was 21%. There were no severe complications among the patients treated with radiation therapy alone, except for some fibrotic changes recognized in the radiographs, although this did not cause any symptoms in the patient.

Van Houtte and colleagues reported on 31 patients with superior sulcus tumors treated with external high-energy radiation therapy up to a total tumor dose of 2000-7000 cGy.
The overall 5-year survival rate was 18%. The doses below 5000 cGy and bone invasion were associated with a higher local recurrence rate.

Komaki and colleagues reported on 36 patients with superior sulcus tumors who were treated with external radiation therapy between 1963 and 1977 at the Medical College of Wisconsin. Local control correlated positively with field size and median survival. All patients who survived beyond 2 years exhibited local control of the tumor. No patient survived beyond 2 years if treatment failed locally. Between 1978 and 1983, an additional 32 patients with inoperable superior sulcus tumors were studied. Relief of pain was achieved in 91% of all patients who presented with pain. Three fourths of the patients with Horner's syndrome responded to the radiation therapy. The disease-free survival rates were 65% at 12 months, 38% at 24 months, 25% at 36 months, and 15% at 48 months. Again, no patient survived beyond 2 years if treatment failed locally. The patterns of failure showed that the brain was the most common site of distant metastasis after the completion of radiation (23 of 68 patients, 34%).

Further investigation of treatment strategies combining high dose radiation therapy (≥ 66 Gy) with chemotherapy is indicated for patients with unresectable and/or node-positive (N2) superior sulcus tumors.

SUMMARY

Resectable superior sulcus tumors that have not distantly metastasized or involved regional lymph nodes (as determined by an adequate staging system, including mediastinoscopy) can be treated with combined surgery and radiation therapy with or without chemotherapy. At present, there is no significant difference between the results of preoperative and postoperative radiation therapy for resectable superior sulcus tumors. At M. D. Anderson Cancer Center, a metastatic work-up, MRI of the upper thorax and lower neck, and mediastinoscopy are used to evaluate the extent of the disease into the spinal canal, and to select patients with stage IIB (T2N0M0) tumors for immediate surgery followed by postoperative radiation therapy (1.2 Gy BID fractionation) and chemotherapy (oral VP-16 and cisplatin). Those patients who exhibit a complete response are offered prophylactic cranial irradiation. At present, 23 patients are being treated on this protocol; the 2-year overall survival is 65% and pain control has been excellent. In most cases of treatment failure, the patients had a distant metastasis (i.e., in the adrenal gland or lung) or a second malignancy in the lung.

Patients who have medically inoperable or surgically unresectable superior sulcus tumors need to be treated with concurrent chemotherapy and radiation therapy. This provides rapid relief of their symptoms and more effective treatment. In patients whose tumors have been adequately staged as stage IIB (T3N0M0), who have a good performance status, and who have had a less than 5% weight loss, preoperative chemotherapy and/or radiation therapy will probably not improve local control, long-term survival, or pain relief when compared with immediate surgery followed by definitive radiation therapy with or without chemotherapy. If the surgical specimen reveals no microscopic disease or nodal involvement (N1 or N2), postoperative radiation therapy is not indicated. However, radical surgery is necessary for adequate margins. If the tumor is marginally resectable (as determined by adequate MRI and CT scans), tumors may be treated with concurrent radiation therapy (45 Gy over 5 weeks) and concurrent chemotherapy (VP-16 and cisplatin) followed by surgery 1 month after their completion. Whether this will be better than definitive concurrent chemotherapy and radiation therapy alone at improving quality-of-life, long-term local control, and survival is unknown. Patients who have positive margins or microscopic N2 disease need postoperative radiation therapy. According to a recent randomized study, postoperative concurrent chemotherapy for N2 disease did not improve survival. Patients who have not had preoperative radiation therapy, but whose tumor margins are grossly positive need a definitive dose of radiation therapy (66 Gy without chemotherapy or 60-63 Gy with concurrent chemotherapy). Since most patients with superior sulcus tumors present with adenocarcinoma or large cell carcinoma, prophylactic cranial irradiation needs to be considered early in the course of treatment.

The management of superior sulcus tumors has been improving, along with the use of MRI and CT to stage and delineate the primary tumor and lymph node involvement and the application of spiral CT to planning optimization. Since superior sulcus tumors at the apex of the lung are less mobile than tumors at the base of the lung, superior sulcus tumors can now be treated more effectively using sophisticated 3-dimensional conformal or intensive moderating techniques of radiation therapy. In fact, the improvement in local control means that treatment failures now occur less often at the primary tumor site and more often at sites of distant metastasis. In the future, newer tools such as positron emission tomography scanning might even be used to detect lymph node and other distant metastases in patients with superior sulcus tumors and thus improve treatment even more.

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