

Superior Sulcus (Pancoast) tumors of the lung

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Introduction

Superior sulcus tumors (SST) of the lungs are relatively slow growing at the primary site, with distinctive biological behavior and associated symptoms and syndromes, when compared to other non-small cell lung cancer (NSCLC) with similar histology. Once disseminated beyond adjacent organs, however, the distribution and the pattern of metastatic foci are similar to other NSCLC. The estimated occurrence of SST ranges between 3% and 8% of all pulmonary carcinomas⁹. SSTs arise from the pulmonary parenchyma anatomically located at or above the pulmonary superior sulcus, which marks the position of the subclavian artery on the surface of the lungs¹¹. This portion of the lung is closely surrounded by complex structures of chest wall, including C7 and T1 vertebrae, part of the first and second upper ribs, as well as the clavicles bilaterally. Along with the bony structures, the apex of the lungs are adjacent to subclavian arteries and veins, carotid trunks, superior vena cava, as well as nerve roots. This peculiar position might explain the unique symptoms related to this tumor. Pancoast described, in 1924 the typical presentation of the superior sulcus tumors, with the clinical syndrome named after him¹. The direct invasion of the adjacent structures of chest wall and mediastinum² accounts for the characteristic symptoms, including shoulder and upper extremity pain, as well as alterations of regional innervation (head and neck and upper limb). The peripheral position of the SSTs is associated with relatively infrequent signs or symptoms of pulmonary disease, like cough, dyspnea or hemoptysis. Clinical presentation, as well as treatment planning are closely related to the organs affected or invaded by the tumor mass. Pain and Claude Bernard – Horner syndrome are common manifestations of chest wall and nerve invasion. This study presents our experience with the treatment of superior sulcus lung tumors, as well as the long-term outcomes in our patient population. The main objective of the study is to evaluate survival rates of patients with NSCLC tumors located at the superior sulcus of the lungs, and submitted to different treatment approaches.

Methods

This is a retrospective study of all patients admitted with the diagnosis of SST of the lung, with no evidence of distant metastases, between 1990 and 2007. During that period, 19 patients were treated for NSCLC, confirmed by percutaneous fine needle aspiration (n=11) or core biopsy (n=8). Median age at diagnosis was 62 years (range: 59 to 71). Most patients were male (n=14, 73.7%), with median performance status of 90% (range: 80% to 100%). All patients had one or more of the characteristic local symptoms. Pain was the main complaint, present in 100% of the patients. Neurological alterations due to invasion of nerve roots were noted in 14 patients. Full Pancoast syndrome was observed in seven patients (36.8%). Thorough pretreatment staging was performed, including CT scan of the chest and upper abdomen, CT or MRI of the brain as well as whole body bone scan. The last four patients were submitted to PET-CT scans (18F-FDG). After ruling out any detectable distant disease, the patients were treated. The patients were staged as follows: IIb (n=12), IIIa (n=5), and IIIb (n=2). Management included radiotherapy only (n=3), radiotherapy and chemotherapy (n=7), radiotherapy and surgery (n=4),

chemotherapy followed by surgery (n=3), and chemo-radiotherapy followed by surgery (n=2). Patients candidates to surgical resection has histological evaluation of their mediastinum through mediastinoscopy. Surgical resection was considered pathologically complete in eight patients (88.8% of resected patients). En bloc chest wall resection and upper lobectomy was performed on all operated patients, associated with systematic mediastinal lymph-node dissection. Frozen section evaluation of surgical margins was routinely performed. Preoperative radiation ranged between 30 Gy and 45 Gy. Exclusive radiation therapy dose ranged between 50 Gy and 65 Gy. All chemotherapy regimens were platinum based. Follow-up included chest and upper abdomen CT scan every three months.

Statistical analyses were performed on SPSS 11.5 for windows.

Results

Overall objective response rate to radiation or chemoradiation therapy, as well as to neoadjuvant chemotherapy was 68.4%. There was no treatment-associated deaths. Thirty-day postoperative complication rate was 55.6%, with major complications seen in two patients (bleeding that required reoperation and pulmonary embolism, respectively). Median follow up was 22 months (range: 7 to 62 months). Overall median survival rate of all patients included was 34 months (95% CI: 15-53 months - Figure 1), as estimated by Kaplan-Meier method. Patients submitted to surgical resection presented with better, although not significant (p=0.129), overall survival (57% at five years), as compared to the unresected group, with a median survival of 29.2 months, and no patient surviving over five years (Figure 2). Of the patients submitted to surgical resection, five-year disease-free survival rate was 32% (median 39 months).

Discussion

Superior sulcus lung tumor is associated with apparently different prognosis, as well as biological behavior, warranting special treatment strategies. Since 1961, neoadjuvant radiotherapy, followed by surgical resection was considered the treatment of choice in operable patients³. Several prognostic factors (like lymph node metastases, vascular and vertebral body invasion) were subsequently identified to help defining the patient population more likely to benefit from this aggressive approach^{4,5}. Unfortunately, some patients with SSTs still present with unresectable disease, with complete resection only reached in 65% to 70% of patients. Several technical modifications were introduced to increase operation rates, including vascular replacement and vertebral body resection^{6,7}. Neoadjuvant treatment nowadays include induction chemoradiotherapy followed by an attempt to complete resection. Most studies used a combination of drugs with a platinum compound. Patients submitted to neoadjuvant treatment with complete resection, presented with 53% five-year survival rate^{8,9}. Multimodality treatment, including chemotherapy as well as radiotherapy and surgery, was established as the treatment of choice since late 1990's. In patients with invasion of adjacent subclavian vessels or vertebral bodies, operative technique should include a team of vascular and

orthopedic surgeons with experience in similar situations. On the other hand, patients with unresectable disease or unfit for complete resection, are considered for radiotherapy or combined chemoradiotherapy, with five-year survival rates ranging from 12% to 25%.

Data from the present study, where we present our experience with SSTs, confirm previous reports that demonstrated the effectiveness of aggressive approach to superior sulcus lung tumors. Initial treatment with chemotherapy and/or radiotherapy resulted in overall objective response rate (RECIST) of 68.4%. Resection was only possible in 9/17 patients, after induction therapy. Five-year survival rate of 57% for completely resected patients is encouraging. The small number of patients included in this study probably accounts for the lack of statistically significant difference between the groups of patients resected, as compared to the patients with unresected disease.

This study, although retrospective, shows that multimodality approach for patients with SSTs of the lungs should be considered as treatment of choice, mainly in specialized centers. Results of this treatment are similar to those reported by prospective studies. Thorough staging, including PET-CT evaluation, should be routinely employed in this group of patients, to avoid unnecessary major operations^{10,11}.

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Figures

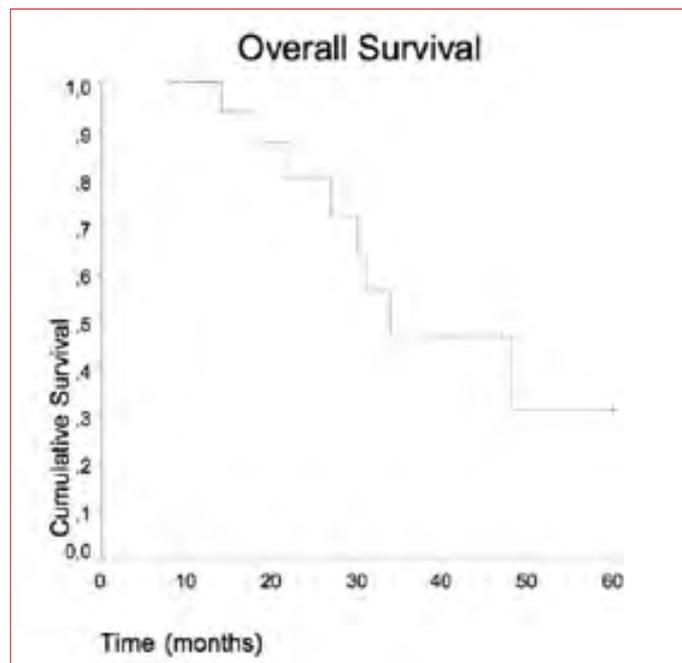


Figure 1- Overall survival of all patients included

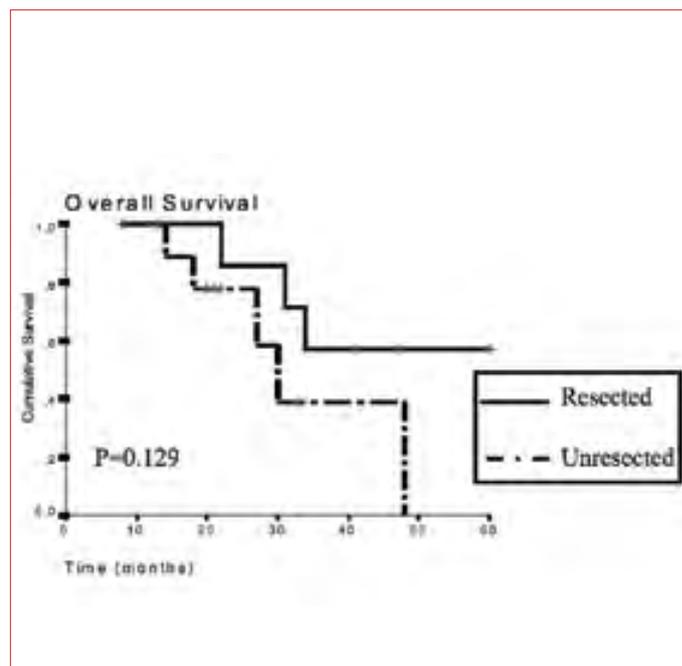


Figure 2- Overall survival of patients related to operative procedure