Prolonged Survival Following the Superior Vena Cava Syndrome*

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Bronchogenic carcinoma complicated by the syndrome of superior vena caval obstruction has been considered uniformly fatal. A patient with this syndrome due to small cell undifferentiated carcinoma of the lung has survived for 13 years following radiation therapy without evidence of recurrence. It is recommended that radical irradiation should remain as part of the management of patients with this disease confined to the chest until better chemotherapeutic regimens are developed.

The obstruction of the superior vena cava by compression or infiltration of a malignancy is one of the few emergencies encountered in clinical oncology.1 Although surgical or chemotherapeutic decompression has been proposed for some clinical situations, mediastinal irradiation is more commonly employed as primary therapy for this life-threatening syndrome.2 Patients with the superior vena cava syndrome secondary to Hodgkin disease or malignant lymphoma may be cured by radical irradiation, but few, if any, patients with the syndrome due to bronchogenic carcinoma have had prolonged survival following treatment.3 Because of the commonly held opinion that the syndrome is uniformly fatal for the patient with lung cancer, we report a case of 13 years' disease-free survival following treatment for the superior vena cava syndrome.

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CASE REPORT

A 47-year-old woman presented to Walter Reed Army Medical Center in May 1964 with a three-month history of cough, dysphagia for solid foods, “fullness” in the chest and neck, and a 4-kg (9-pound) weight loss. Physical examination on admission revealed multiple 1-cm lymph nodes in the right cervical and supraclavicular regions, facial plethora and edema, neck vein distension, and prominence of the venous pattern on the chest and upper extremities. The clinical diagnosis of superior vena caval obstruction was confirmed by an elevated upper extremity venous pressure of 324 mm of saline compared to a lower extremity pressure of 117 mm of saline solution. A large superior mediastinal mass was present on chest x-ray film which was noted to compress the esophagus on a barium swallow. Biopsy of a right scalene lymph node was interpreted as “undifferentiated carcinoma consistent with bronchogenic origin.” The patient was treated with 6,000 rads of mediastinal and neck irradiation over a seven-week period. The superior vena cava syndrome resolved with irradiation, and the mediastinal mass decreased in size. In December 1964, a 3-cm mass was noted in the lateral aspect of the right supraclavicular fossa just outside of the original radiotherapy portal. This also resolved following an additional 6,000 rads. The patient has been symptomatic since that time, and when last seen in December 1977, had no abnormalities noted by physical examination, complete blood count, or chemistry profile. A chest x-ray film taken 13 years following treatment revealed paratracheal fibrosis secondary to irradiation but no evidence of tumor (Fig 1).

PATHOLOGY

Hematoxylin and eosin, periodic acid-Schiff, mucicarmine, and reticulin stained sections, prepared from the original paraffin-embedded lymph node removed in 1964, were examined microscopically. Cohesive sheets of tumor cells filled the peripheral sinuses of the node and extended through the paracortical region into the medullary cords. The malignant cells varied in size but were approximately three times larger than the adjacent, displaced lymphocytes on the average (Fig 2). Mitoses were numerous and frequently atypical. The nuclei were

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Figure 1. Posteroanterior view 13 years following treatment showing paratracheal fibrosis but no evidence of tumor.

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irregular in outline and showed coarse clumping of their chromatin. The PAS and mucicarmine stains failed to reveal cytoplasmic mucin. Reticulin fibers were sparse in sections of the node replaced by tumor. These architectural and cytologic features are typical of small cell carcinoma which is most frequently of pulmonary bronchial origin.

Ultrastructural examination was unrewarding in this case because of fixation and embedding artifacts. When "neurosecretory-like" granules can be demonstrated in the cytoplasm of the small malignant cells, it is postulated that such lesions arise from APUD precursors (Kulchitsky cells) within the bronchial mucosa. Frequently, however, evidence of squamous or glandular differentiation is found on electron micrographs. In these areas, it seems reasonable to suggest that bronchial reserve cells are the precursors. It is clear that lesions similar to the one presented, which are currently designated as small cell carcinomas in the WHO and WP-L classifications, comprise a heterogeneous group with respect to morphologic appearances, proposed cell of origin, and malignant potential.

DISCUSSION

The syndrome of superior vena caval obstruction is caused by malignant tumors in 97 percent of all cases. Bronchogenic carcinoma accounts for 75 to 80 percent of these tumors with small cell undifferentiated carcinoma being the most common histologic type. Since the description by Roswit et al of its pathologic physiology in 1953, mediastinal irradiation has been the most common treatment for this condition. In general, treatment to the mediastinum and supraclavicular regions with total doses of 3,000 to 6,000 rads have been recommended. Initial treatment with large daily doses of 400 rads per day have been considered by some to offer more prompt palliation. Although symptomatic relief is noted in 75 to 90 percent of patients following irradiation, most go on to die of distant metastases and/or uncontrolled thoracic disease with a median survival of less than six months. There have been several reports of patients surviving for more than five years following treatment, but most have been patients with lymphoma or patients without biopsy-proven malignancy. LeRoux reported the ten-year survival of a patient with undifferentiated carcinoma of the lung but did not offer review of the pathology.

The patient we report is remarkable both for her long disease-free survival following irradiation for small cell undifferentiated carcinoma of the lung, and for her salvage by irradiation following the development of supraclavicular metastasis. She represents one of the few long-term survivors with well-documented review of the pathology reported in the literature. Although combination chemotherapy has recently been proposed as primary treatment for superior vena caval obstruction due to small cell undifferentiated carcinoma of the lung, we feel that patients presenting with disease confined to the chest may be cured by radical irradiation alone, and therefore, such treatment should be part of their management along with adjuvant chemotherapy.

REFERENCES