Recurrent Bronchial Carcinoid Tumor*

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Bronchial carcinoid tumors are considered to be of low grade malignancy, and if completely resected, to be cured. A patient with resection of a bronchial carcinoid presented 18 years later with superior vena caval obstruction, and carcinoid syndrome due to a recurrence. There was an excellent response to radiation. (Chest 1989; 95:693-94)

Bronchial adenomas, of which 85 to 95 percent are carcinoid tumors,1-3 comprise 1 to 4 percent of all lung tumors. These have in the past been considered to be of low grade malignancy, and if completely resected, were thought to be cured. Carcinoid syndrome occurs in about 5 percent of all bronchial carcinoid tumors.4 When it does occur, it is usually at recurrence, rather than primary presentation, and almost invariably with hepatic metastases. The tumor is generally considered resistant to irradiation.

This report is of a patient who had a noninvasive bronchial carcinoid tumor completely resected by pneumonectomy, who presented 18 years later with tumor recurrence. She had superior vena caval obstruction, carcinoid syndrome without hepatic metastases, and a response to irradiation. These unique features bring into question some of our "conventional wisdoms" about this tumor.

CASE REPORT

In 1968, a 35-year-old nonsmoking white woman underwent right thoracotomy for bronchial carcinoid adenoma of the typical variety (Fig 1). The tumor was in the right main bronchus take off, and for technical reasons, a right pneumonectomy was necessary. The margin of resection and hilar nodes were free of tumor. The postoperative course was complicated by a bronchopleural fistula, empyema, and subsequently, she underwent a right thoracoplasty. Pathologic specimen from the second procedure showed no evidence of tumor.

She did well except for chronic shortness of breath. Between 1971 and 1986, she had five episodes of hemoptysis, underwent bronchoscopy repeatedly, and all that was found was a healed bronchial stump with granulation tissue. In 1986, her vital capacity was 0.7 L with an FEV1 of 0.4, representing severe restrictive and obstructive lung disease.

In November 1986, she presented to the Chest Clinic because of swelling of both breasts. She was found to have facial puffiness, especially in the morning, and bilateral hand edema. She complained of loose bowel movements and episodes of flushing and sweating. On examination, she was found to have distended external jugular veins, bilateral breast edema, and distention of the veins on her chest and abdomen. The chest roentgenogram was unchanged from those through the years, revealing only the right thoracoplasty (Fig 2). The CT scan, however, showed a right thoracic mass at the level of the resection of the right main bronchus. The superior vena cava was narrowed greatly. Superior vena cavaogram showed 90 percent occlusion (Fig 3). Fine needle aspiration performed under CT guidance of the right thoracic mass revealed cells characteristic of carcinoid adenoma.

Twenty-four urine 5HIAA was increased to 70 mM/dL (normal range 5 to 35). Liver function test results, including SGOT, alkaline phosphatase, LDH, and isotope scan, were normal.

Because of the patient's severely impaired respiratory status, she was treated with irradiation; 5,200 rads were administered to her mediastinum. Over the course of the next month, the manifestations of superior vena caval obstruction regressed. The CT scan showed regression of the tumor mass, and the urinary 5HIAA dropped to 30 mM/dL.

Nineteen months after the completion of radiotherapy, there was no clinical evidence of recurrence.

DISCUSSION

The patient is reported because of the combination of several unusual features in her clinical course among which are the time and manner of recurrence. The literature states that if the tumor has been completely resected, the outlook are generally good and ten-year survival rates vary from 85

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FIGURE 2. PA chest roentgenogram showing right pneumonectomy and thoracoplasty. Unchanged since 1968.

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to 100 percent.\textsuperscript{3,5-8} 15-year survival rates vary from 80 to 85 percent,\textsuperscript{3,8} and 25-year survival rates approximate 65 percent.\textsuperscript{3,7} However, late reappearance of tumor has been noted, and in one series in which there was a five to 30 year follow-up in 57 patients, there were two recurrences at nine and 16 years.\textsuperscript{3}

There are other case reports of late recrudescence of tumor in the literature, but those had evidence of metastasis at time of original surgery.\textsuperscript{10,11} One remarkable case reported by Altschule et al\textsuperscript{12} had a pneumonectomy in 1943 for bronchial carcinoid which was infiltrating into adjacent lung tissue and metastasized to regional lymph nodes. In 1975, 32 years later, the patient presented with superior vena caval obstruction and increased urinary 5-HIAA without symptoms of carcinoid syndrome.

Carcinoid syndrome has been reported in 1 to 7 percent\textsuperscript{6,7} of bronchial carcinoids, and almost invariably indicates liver metastases. The review by Ricci et al\textsuperscript{13} revealed that five of 66 patients were reportedly free of metastases; however, there was a satisfactory search for hepatic metastases in only one of those five patients. It has been pointed out\textsuperscript{6,7} that "carcinoid syndrome" may, however uncommon, be present in patients with bronchial carcinoid without liver metastases.

Superior vena caval obstruction at the time of original presentation or subsequently as a manifestation of metastatic disease has been reported in only one other case in the literature,\textsuperscript{10} and the case reported here is the only case in the literature in which the primary tumor was completely resected, and in which the presenting complaint was swollen breasts.

The difficulty in early diagnosis by usual means is unusual. Chest roentgenogram reveals abnormalities in 90 to 95 percent of patients with bronchial carcinoid.\textsuperscript{1,3,5-9} but in this patient, it was unchanged. Because the tumor often presents in the main bronchi, bronchoscopy is a sensitive method of identifying the lesion in 80 to 90 percent of cases.\textsuperscript{1,3,5,7,9} However, repeated bronchoscopy missed the recurrence in this patient. A CT scan, however, did reveal evidence of recurrence with compression of the superior vena cava.

Despite the claims\textsuperscript{5-10} that radiotherapy is not useful in the treatment of this tumor, except perhaps in the palliation of liver disease, it is noteworthy that this patient was successfully palliated with radiotherapy. Not only did she have resolution of her symptoms secondary to superior vena cava obstruction, but also remission of her carcinoid syndrome and a drop to normal values of 5HIAA in her urine.

There are only two other patients reported\textsuperscript{26} in whom there was a successful response to radiotherapy as demonstrated by diminution in tumor size.

In conclusion, a patient has been reported with unusual features of a carcinoid tumor which recurred after 18 years, presenting with superior vena caval obstruction and carcinoid syndrome which has responded to irradiation. This appears to be unique in the literature.

**References**