nant neoplasms, which are known to metastasize to heart.\(^{20}\)

Hypersecretion without a clinical carcinoid syndrome has been recognized in association with bronchial adenoma of carcinoid type.\(^{21}\) In the present case neither the heart nor the lungs showed the lesions frequently seen in the carcinoid syndrome, ie the characteristic endocardial thickening or intimal changes in the pulmonary veins. Rather, a small area of a nonspecific endocardial fibrosis was seen. Conspicuous narrowing of the right ventricular outflow tract by the tumor as well as the large pericardial effusion may have contributed to the patient's dyspnea and eventual death. Therefore, this patient's fulminant course was not caused by an excess of circulating serotonin or related compounds, but it can be ascribed to a highly malignant neoplasm, a bronchial carcinoid.

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REFERENCES
5 Foster-Carter AF: Bronchial adenoma. Quart J Med 10:139-174, 1941

Alveolar Soft-Part Sarcoma

Report of a Case Presenting as Asymptomatic Pulmonary Nodules

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Alveolar soft-part sarcoma is an unusual tumor of uncertain histogenesis which usually presents as a painful mass in the right thigh in young women and progresses slowly but inexorably to death. A search of the English literature revealed only 65 cases, none of whom presented with asymptomatic pulmonary nodules. The authors recently had the opportunity to see a young man with multiple pulmonary nodules whose biopsy diagnosis revealed alveolar soft-part sarcoma. It is the purpose of this report to add this tumor to the list in the differential diagnosis of pulmonary nodules.

Alveolar soft-part sarcoma, described by Christopherson et al\(^1\) in 1952, as an unusual tumor of uncertain histogenesis, usually presents as a painful mass in the right thigh in young women. We have recently had the opportunity to study the first reported case of this tumor presenting as multiple pulmonary nodules in a routine chest roentgenogram in a totally asymptomatic young man. It is the purpose of this report, therefore, to add this tumor to the differential diagnosis of asymptomatic pulmonary nodules.

CASE REPORT

An asymptomatic 24-year-old white man on active duty with the United States Navy was found to have multiple pulmonary nodules on a routine chest roentgenogram (Fig 1). He was a one-pack-per-day cigarette smoker and had had one bout of pneumonia at the age of three. He had lived in the mid-west briefly, but had spent most of his life in

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ALVEOLAR SOFT-PART SARCOMA

Figure 1. Multiple pulmonary nodules, right lower lobe.

California. A roentgenogram two years before this admission was said to have been normal but was unavailable. The physical examination and all laboratory studies gave negative or normal findings including studies for mycobacterial and fungal infections and a primary source of carcinoma.

All attempts at a definitive diagnosis by noninvasive techniques failed and at exploratory thoracotomy many nodules were found scattered throughout both lung parenchyma. Easily enucleated, these nodules were classified pathologically as alveolar soft-part sarcoma (Fig 2).

The patient's postoperative course was uneventful and he returned to his home area for follow-up. A report six months after surgery indicated that he remained asymptomatic and his local physicians had elected to withhold chemotherapy.

Discussion

Alveolar soft-part sarcoma is an unusual tumor of uncertain histogenesis, but is always associated with skeletal muscle. It tends to occur in young adult women with a true laterolization to the right side of the body with the primary usually found in the thigh. It is inexorably fatal despite an indolent course in some patients. Death is always from disseminated disease with metastases involving lung, bone, and brain in 42 percent, 19 percent, and 15 percent of cases respectively. Prolonged survival after excision of the primary tumor mass, however, is not unusual and approximately 50 percent of patients are alive at five-year follow-up. An occasional patient may exceed a 15 year survival. Patients tend to remain in relatively good general health with few, if any, systemic symptoms until the terminal stage of their disease.

The primary therapeutic modality has been surgical excision. The tumor is only moderately radiosensitive, thus limiting radiotherapy for local relief of pain from bony metastases. Very few patients have been treated with chemotherapy and there are no controlled studies. Asvall et al reported a case in 1969 in whom thio-TEPA caused disappearance of pulmonary metastases for a period of four years. Occasional use of prednisone, anabolic steroids, cyclophosphamide, and actinomycin-D has not appeared to influence the course of the tumor. Chemotherapy was withheld at this time in this asymptomatic patient.

Our patient presented with obvious pulmonary metastases, but was completely asymptomatic. An extensive search short of a laparotomy, but including soft tissue radiography by a radiologist alerted to the diagnosis, failed to reveal the primary lesion. A bone scan utilizing radioactive strontium, however, did reveal another area of metastatic involvement of the ileum. No similar case could be found in the literature. We feel, therefore, that this case report serves to call attention to this diagnostic possibility in the case of asymptomatic pulmonary nodules of obscure etiology.

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References


Figure 2. High power detail (100x) of the biopsy section in this patient. Normal but compressed lung may be seen above the tumor cells arranged in a pseudoalveolar pattern.