Pulmonary blastoma: Report of Two Cases*

Abdul Ghaffar, M.D.;** S. V. Vaidynathan, M.D.;**
Alberto Elguezabal, M.D.;† and Bernard S. Levowitz, M.D.;‡

Pulmonary blastoma is a rare primary malignancy of the lung originating from multipotential pulmonary blastema. Two cases of this tumor are reported emphasizing lack of correlation between the microscopic appearance and the clinical course. There are no specific clinical features that differentiate this tumor from the more common lung neoplasms. Surgical resection is the preferred treatment; the merits of radiotherapy and chemotherapy have not yet been established. Prognosis after adequate resection appears no better than for other pulmonary malignancies.

Pulmonary blastomas are a group of rare malignant neoplasms first described by Barnard1 in 1952 as embryomas of the lung because of their histologic similarity to fetal lung. In 1961 Spencer2 reported three new cases and used the term "pulmonary blastoma" for a tumor thought to be derived from pluripotent immature pulmonary primordium. Despite their striking resemblance to nephroblastomas, which are primarily tumors of infancy and childhood, the vast majority of the reported cases of pulmonary blastoma have occurred in adults. This presentation documents two cases of pulmonary blastoma appearing at different ages with dissimilar clinical courses. The first case is of special interest for it represents the youngest patient with pulmonary blastoma yet recorded.

Case Reports

Case 1

A nine-year-old Puerto Rican boy was admitted to our hospital on November 14, 1972 with a history of intermittent fever and left-sided chest pain of eight weeks' duration. The patient's mother had been aware of his reduced growth rate, small stature, and tendency to fatigability over the last two years.

Physical examination revealed dullness to percussion and diminished breath sounds over the left side of the chest. Chest x-ray films and tomograms showed a large, well circumscribed opacity occupying the upper three-quarters of the left chest. The mass displaced the heart and the mediastinum slightly to the right, compressed the left lower lobe, and caused elongation of the left main stem bronchus (Fig 1). Echogram suggested the presence of fluid within the lesion. Thoracentesis yielded 40 ml of serosanguinous fluid which showed no neoplastic cells on cell block study and was sterile on culture.

Left pneumonectomy was carried out on December 9, 1972. The left upper lobe was completely replaced by a soft, white, friable tumor that spared only a small portion of the lingula segment. Adhesions of varying thickness bound the mass to the parietal pleura. There was extensive involvement of the pericardium and, at the conclusion of the resection, residual tumor was present in the mediastinum. Because of histologic similarities between the pulmonary lesion and Wilms' tumor, nephrotomograms and bilateral selective renal arteriograms were performed to exclude a suspected renal embryoma. These studies were negative as were the findings at exploratory laparotomy performed on January 12, 1973.

The child was subsequently given radiation (3500 R) to the left hemithorax and mediastinum and several cycles of chemotherapy with methotrexate and bleomycin. Despite vigorous treatment, he demonstrated metastases to the right lung, jaw and a large recurrent tumor mass in the left pleural cavity. Terminally he developed a massive left pleural effusion and finally died on October 17, 1973. At autopsy, there was recurrent tumor in the left pleural cavity with local invasion of the diaphragm, mediastinum, pericardium and the ribs. Metastases were found in the pancreas, brain, right lung and vertebrae.

Pathology. The surgical specimen consisted of left lung weighing 620 grams. The upper lobe was enlarged and totally replaced by a bulging, solid mass which measured 14.5 x 8...
FIGURE 2. Gross section of the tumor reveals fleshy character, central necrosis and cavitation. Compressed left lower lobe attached to tumor mass (Case 1). x 6 cm. The cut surface was grayish white and fleshy with irregular central necrosis and cavitation (Fig 2). The lower lobe was collapsed and free of tumor. Microscopically the lesion demonstrated spindle cells and tubular structures dispersed in a stroma of delicate fibrous tissue. The spindle cells were the predominant component and showed distinct nuclear hyperchromasia, pleomorphism and scattered mitotic figures. The tubular structures of variable size had well formed lumina and the lining cells varied in thickness from a single cell to several layers of cells (Fig 3). No mitoses were noted among these cells. Hilar nodes were uninvolved by the tumor.

CASE 2

A 48-year-old black man, a nonsmoker, was admitted to our hospital on April 3, 1971 following the incidental discovery of an opacity in the left lung on a chest roentgenogram. The patient was asymptomatic except for a history of hypertension controlled with medication. Physical examination and routine laboratory studies were within normal limits. The chest roentgenogram revealed a mass in the periphery of the left lower lobe.

Left lower lobectomy was performed on March 5, 1971 for a 4 cm nodular mass of cartilagenous consistency that projected from the pleural surface and was reported as a spindle cell tumor on frozen section. The patient's postoperative course was uneventful and he has been well and completely free of recurrence at follow-up examination three years later.

Pathology. Inspection of the surgical specimen revealed a 4.5 x 3.5 x 1.5 cm, smooth, grayish-white, irregular solid mass protruding from the pleural surface. The cut surface was homogenous, smooth and glistening. The surrounding lung tissue, as well as the remainder of the left lower lobe, was unremarkable except for emphysema (Fig 4). Microscopically the tumor was composed of spindle cells and scattered tubular structures, the histology for the most part being similar to the previous case (Fig 5). In this instance, the stroma was more abundant in some areas and there were foci of early chondroid metaplasia. The lymph nodes were negative for tumor.

DISCUSSION

Pulmonary blastoma is a rare malignant neoplasm which generally arises in the peripheral portion of the lung and demonstrates variable clinical behavior. Twenty of these lesions have been previously recorded in patients whose ages ranged from 15 to 77 years. Males outnumbered females by 4.5 to 1. Symptomatology varied from patients who were asymptomatic to those with symptoms of cough, hemoptysis, and chest pain. The prognosis for pulmonary blastoma is generally poor, with a reported 5-year survival rate of 10-20%.

FIGURE 3. Micrograph of section illustrates epithelial tubular structures surrounded by cellular sarcomatous stroma with hyperchromasia, pleomorphism, and scattered mitotic figures (Case 1) (H & E × 250).

FIGURE 4. Gross specimen showing solid homogenous tumor attached to the left lower lobe (Case 2).

FIGURE 5. Micrograph illustrates spindle cells and tubules distributed in a loose fibrous stroma (Case 2), (H & E × 100).
who exhibited cough, hemoptysis, chest pain and shortness of breath. There were no clinical features to distinguish blastomas from other pulmonary neoplasms.4

The commonly employed diagnostic measures (chest x-ray films, cytologic studies of sputum, bronchoscopy and scalene node biopsy) were of limited value in establishing the diagnosis. Identification of the tumor was made only at thoracotomy or autopsy. In operable lesions wedge resection, lobectomy or pneumonectomy were employed. Surgical resection was the preferred treatment, although in two patients with metastases satisfactory palliation had been achieved with radiotherapy.3,5,6 The role of postoperative radiation and/or chemotherapy has not yet been defined. An aggressive therapeutic approach to our first case employing these modalities after pneumonectomy, did not prove beneficial.

Although a few long-term survivals have been reported, the overall prognosis in pulmonary blastoma is unpredictable and should be regarded as unfavorable. Spencer’s7 recorded follow-up of Barnard’s4 case revealed that the patient was well and free of disease 15 years after resection. On the other hand, of 17 definitive cases reviewed by Karcgiolu and Someren,8 11 had died within one year after treatment, five were between one and three years, and two had survived beyond five years. Among three cases in their first or second decade of life, including our Case 1, two were reported as surviving beyond one year. On the basis of histologic observations and autopsy records showing locally invasive characteristics and metastases, the malignant potential of these lesions is clearly evident and demands, whenever possible, an aggressive surgical approach. Treated by lobectomy, our second case has been well and free of recurrent tumor during a three year follow-up period.

Histologically, the blastoma bears close resemblance to fetal lung and Wilms’ tumor of the kidney. In a discussion of their two patients, Parker, Payne and Woolner9 referred to a case where occurrence of pulmonary blastoma in an 11-year-old boy necessitated extensive surgical exploration to rule out a nephroblastoma as in our first case. The dimorphic histologic pattern includes scattered tubular elements surrounded by cellular connective tissue stroma. Spencer2 noted differentiation of the primitive stroma to form fibrous tissue and smooth muscle. A tumor with a striated muscle component has been described,2 but epidermoid derivatives have never been reported. While fibrous tissue capsules and pseudocapsules are often present, the tumor is locally invasive and gives rise to regional and distant metastases. These have been reported to involve liver, brain, adrenals, pancreas and retroperitoneum.8 The histologic appearance of these tumors cannot be correlated with their clinical behavior with any degree of accuracy.

The histogenesis of pulmonary blastoma is still uncertain and several theories concerning its origin have been proposed. According to Waddell,4 the peripheral respiratory portion of lung is derived from mesenchyme while the distal air passages are formed by canalization of cords of mesenchymal cells which grow in apposition to the terminations of the laryngotracheal bud. Accepting Waddell’s concept of embryogenesis of the lung, Spencer2 postulated an origin of this neoplasm from pulmonary blastema, similar to the manner in which a nephroblastoma is derived from pleuripotent immature renal blastema. Appearance of a pulmonary blastoma at a later age than the nephroblastoma has been attributed by Spencer to the fact that the lung continues to develop for at least ten years after birth while the renal parenchyma is mature at the time of birth. Adult mesenchymal tissue may contain undifferentiated cells which are capable of reverting to an embryonic state and thereafter produce both epithelial and mesodermal elements.

There has been some confusion in distinguishing these tumors from carcinosarcomas because of the mixed cellular pattern of both tumors. Carcinosarcomas usually consist of squamous carcinoma in a sarcomatous stroma and bear no resemblance to fetal lung.10 In contrast, pulmonary blastoma has a glandular epithelial component surrounded by abundant immature sarcomatous stroma, a pattern quite similar to that found in fetal lung. Several reports have emphasized the origin of pulmonary blastoma from the peripheral portion of the lung, whereas carcinosarcoma often originates in a major bronchus. Despite these apparent differences, pulmonary blastomas have been considered a distinctive group of the carcinosarcomatous tumors resembling primitive pulmonary blastema.4,10

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