A patient with a systemic-to-pulmonary arteriovenous fistula associated with intrapulmonary Hodgkin's disease is presented. Shunting of blood from an internal mammary artery into the pulmonary arterial system via a highly vascular tumor mass was demonstrated.

Pulmonary arteriovenous fistulae are not uncommon. Their etiology is varied and includes congenital, infectious, and iatrogenic causes. Pulmonary arteriovenous fistulae involving chest wall vessels are rare. This report presents a patient with a systemic-to-pulmonary arteriovenous fistula associated with intrapulmonary Hodgkin's disease. It is the eighth report of a pulmonary arteriovenous fistula involving chest wall vessels and the first demonstration of an arteriovenous fistula of the lung secondary to malignant tumor.

**Review of the Literature**

Burchell and Clagett presented the first case of a pulmonary arteriovenous fistula with collateral circulation involving the thoracic wall. Six additional reports of systemic-to-pulmonary fistulae involving chest wall vessels were found in the English literature.

Pulmonary arteriovenous fistulae may be either congenital or acquired in origin, the former being the most common. Of the seven reported cases involving chest wall vessels, five were felt to be congenital and two acquired. Other reports have implicated pulmonary tuberculosis, longstanding cirrhosis, and schistosomiasis. We were unable to find a previous demonstration of pulmonary arteriovenous fistula secondary to malignant tumor. Pierce et al reported a cyanotic patient with multiple pulmonary metastases from a thyroid carcinoma, but were unable to demonstrate an arteriovenous fistula radiographically or surgically and could not exclude the possibility "that multiple pulmonary arteriovenous fistulas existed on a congenital basis."

**Case report**

A 36-year-old physician had experienced good health until two months prior when he developed generalized myalgia, malaise, and a flulike syndrome, which improved with rest. One month later he developed a cough with only slight production of sputum, pleuritic pain of the left anterior chest, with underlying tenderness of the chest wall and temperature elevation. Self-auscultation of the chest at that time revealed a continuous murmur over the left anterior chest which had not been present on previous physical examinations. Chest roentgenograms demonstrated a large anterior mediastinal mass and left hilar density which pulsed at fluoroscopy, suggesting the diagnosis of an arteriovenous fistula.

There was no family history of telangiectasis or arteriovenous malformation. He had not experienced constitutional symptoms prior to the present illness. On physical examination the blood pressure was 126/84 mm Hg, pulse rate 92/min, temperature 36.5°C, with a respiration rate of 18/min. Sclerae and oral mucous membranes were normal with no evidence of telangiectases. There was no cyanosis or clubbing of the digits. The thyroid gland was normal and there was no lymphadenopathy. A grade 2/4 continuous murmur was heard best in the second left intercostal space at the sternal border, with no radiation.

Laboratory data were as follows: Hgb, 11.8 gm percent; Hct, 35.1 percent; platelet level, 480,000/mm³; erythrocyte sedimentation rate, 55; white blood cell count, 19,700/mm³, with a shift to the left. Serum electrolyte, calcium, phosphorus, blood urea nitrogen, creatinine, uric acid, fasting blood sugar, cholesterol, protein, bilirubin, alkaline phosphatase, lactate dehydrogenase and serum glutamic oxalo-

**Figure 1.** Plain chest roentgenogram showing intrapulmonary mass involving anterior segment of left upper lobe.
Acetic transaminase indices were all in the range of normal. Urine analysis, skin tests for antigens, febrile agglutinins and serum electrophoresis values were normal.

Plain chest roentgenograms (Fig 1) suggested an intrapulmonary tumor involving the anterior segment of the left upper lobe. In retrospect, a small mass can be seen in this location on chest roentgenograms obtained six months prior. On left pulmonary arteriography there was washout of opaque material from the left upper lobe vessels during the arterial phase. The size and caliber of the pulmonary arteries and veins were normal. During the levo phase (Fig 2), the mass in the left upper lobe appeared to opacify with contrast material, and a large left internal mammary artery was seen. Selective injections into the left internal mammary artery (Fig 3) demonstrated two large branches, which supplied a highly vascular tumor involving the entire anterior segment of the left upper lobe. There was rapid shunting of contrast medium from the internal mammary artery into the pulmonary arteries with retrograde flow into the remainder of the left pulmonary arterial system. The vessels within the tumor were tortuous and appeared to contain large lakes with pooling of contrast material.

At left thoracotomy, a large mass was found which involved one-third of the left upper lobe and extended medially where it was firmly attached to the mediastinum and chest wall. The mass was extremely vascular, containing large sinusoids. The left internal mammary artery measured 5 mm in diameter and was ligated at its origin from the subclavian artery. Because of involvement of the phrenic nerve and extension over the aorta, the mass could not be totally excised. It was transected and removed with the left upper lobe of the lung. Cross section of the surgical specimens revealed an ill-defined, firm mass 5.0 cm x 4.0 cm x 3.0 cm within the lung parenchyma. Microscopic sections (Fig 4) showed an extensive infiltration by tumor with the characteristic appearance of Hodgkin's disease, nodular sclerosing type, with many Reed-Sternberg cells identified. Several areas of the mass were conspicuously hypervascular with prominent small and large arteries and veins making up the arteriovenous malformation.

After operation the patient felt well. Subsequent lymphangiography showed no abnormalities of the abdominal lymph nodes, and the final diagnosis of nodular sclerosing Hodgkin's disease, stage I-A, was made.

**DISCUSSION**

Pulmonary arteriovenous fistulae may result in either right-to-left or left-to-right shunting of blood. With the rare occurrence of systemic chest wall vessel involvement, i.e., internal mammary and/or intercostal arteries, the higher pressure systemic circulation may result in a left-to-right shunt with little clinical manifestation, depending on the size of the shunt. Table 1 summarizes the findings of the reported cases of systemic-to-pulmonary arteriovenous fistulae. Only two of the patients displayed cyanosis and clubbing, signs which usually accompany fistulae which lack systemic vessel involvement. In all cases, a localized murmur could be detected over the lesion. Other clues to this type of lesion were usually revealed radiologically, such as mass lesions or rib notching.

Burchell and Clagett reported a case of a 20-year-old man who had displayed cyanosis and clubbing since eight years of age. A chest film at 12 years of age had shown an irregular shadow in the right lung, and at 15 years of age he had an attack of "pleurisy" lasting several weeks. Afterward his symptoms had increased in severity.
symptomatic all of her life and chest x-ray films showed multiple pulmonary nodules in the right lung. Post-mortem examination following unsuccessful operation revealed arteriovenous malformations in both lungs, with intercostal and diaphragmatic arteries participating in those of the right upper and right middle lobes. The authors felt that the history of pneumonia in infancy may have been of etiologic significance in the involvement of the chest wall vessels with the arteriovenous fistula.

Davila's patient, a 24-year-old man, had been asymptomatic prior to developing a mycotic infection of the lung and subsequent appearance of a lucent density which persisted until successful operation, when a systemic-to-pulmonary fistula involving intercostal arteries was resected with the lucent segment.

The diagnosis of an arteriovenous fistula in the 31-year-old woman presented by Brain and Kauntze had been made when the patient was 20 years of age after severe hemoptysis. A chest film when the patient was 12 years of age had shown a right lung opacity. Curative right lower lobectomy revealed an arteriovenous fistula with involvement of anomalous branches from the descending aorta as well as intercostal arteries.

The report by Cox et al described a 39-year-old man with a traumatic systemic-to-pulmonary fistula of the lung and chest wall secondary to insertion and removal of a chest tube. This was treated successfully by surgical separation of the fistulous attachment.

The 23-year-old man described by Kiphart et al had relatively recent onset of right chest pain accentuated with activity. The chest film findings were normal; however, a murmur was heard over the right sternal border and subsequent aortography showed a right chest wall arteriovenous fistula fed by the right internal mammary artery. Wedge resection was curative. No speculation as to etiology was made.

Wolarsky and Humphreys presented a 65-year-old man with an extensive systemic-to-pulmonary fistula involving right intercostal arteries, the etiology of which was felt to be a rib fracture at the time of a previous partial pulmonary resection. It was speculated that the vascular plexus lying in the rib bed established pulmonary drainage via a mass of adhesions and fibrothorax secondary to chronic lung disease.

It is difficult to determine the exact etiology of the seven reported cases of systemic-to-pulmonary arteriovenous fistulae. Brain's case most likely represented an entirely congenital anomaly in view of the unusual involvement of vessels originating directly from the aorta. Only Burchell's and Prutzman's patients displayed cyanosis and clubbing and had symptoms of long duration. These cases present good evidence that the lesions were congenital with secondary parasitization of systemic vessels related to infections. Kiphart's case is more difficult to understand; it too probably represents congenital arteriovenous fistula with secondarily acquired systemic involvement, although no contributory history was cited. Davila's case was felt to be a direct result of mycotic infection with no pre-existing anomaly. Cox's and Wolarsky's patients appear to have acquired their lesion entirely iatrogenically.
Burchell and Clagett (1947)

Prutman and Flick (1954)

Davila et al (1968)

Brain and Kauntze (1960)

Cox et al (1967)

Kiphart et al (1967)

Wolansky and Humphreys (1970)

Dunn and Wexler (1974)

+ = abnormal finding  
- = normal finding

It is felt that the new case herein reported represents primary lung parenchymal Hodgkin’s disease, with extension of the mass into the mediastinum. Parenchymal lung involvement in Hodgkin’s disease is not uncommon and has been reported to occur in 15 to 60 percent of patients. However, only 19 cases of primary pulmonary Hodgkin’s disease have been reported. Systemic-to-pulmonary fistulae have not been reported with other malignant lesions of the chest. To our knowledge, arteriographically, and these lesions are not ordinarily well noted.

The presence of a systemic blood supply to an intrapulmonary mass of Hodgkin’s disease is puzzling. Whether the mass growing from the lung against the chest wall and mediastinum “parasitized” a systemic blood supply, or a pre-existing arterial abnormality somehow induced the tumor formation is moot.

REFERENCES

1 Burchell HB, Clagett OT: The clinical syndrome associated with pulmonary arteriovenous fistulas, including a case report of a surgical cure. Am Heart J 34:151-162, 1947


Pulmonary edema confined to one lung is an unusual event and has been described only infrequently. It requires a lesion of the left heart capable of provoking elevated pulmonary capillary pressure in the presence of a difference in the vascular beds of the two lungs. A patient was recently observed at the Jewish Memorial Hospital who presented with unilateral pulmonary edema and in whom the requisite left heart lesion and abnormality of pulmonary blood flow were identified. The left heart decompensation resulted from previous myocardial infarction while pulmonary blood flow abnormalities resulted from Swyer-James syndrome, ie, acquired unilateral hyperlucent lung. This communication reports the case and describes the clinical, pathologic and radiologic features.

**Case Report**

A 79-year-old white man was admitted on April 30, 1973 to the Jewish Memorial Hospital, complaining of severe respiratory distress. This was his fifth admission with the same complaint. Previous history included two myocardial infarctions in 1958 and 1966. He had severe pneumonia during childhood with recurrent attacks of pneumonitis, thereafter. There was a history of chronic cough and moderate mucopurulent expectoration, which had been present throughout most of his adult life. There were no known allergies. The patient had never smoked and there was no history of exposure to industrial dust or other known causes of chronic pulmonary disease. This patient had previously been admitted to another hospital with the same complaint where various studies were performed including tomograms of the right lung, lung scan, pulmonary angiography, pulmonary function studies, and cardiac catheterization studies. These will be detailed below. On admission to the Jewish Memorial Hospital, physical examination showed that the patient was semiconscious, in severe respiratory distress and cyanotic with frothy sputum coming out of the mouth. Blood pressure was 160/90 mm Hg. Pulse rate was 100. Temperature was

**Unilateral Pulmonary Edema in Swyer-James Syndrome**

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We report a case of Swyer-James syndrome which showed the unusual feature of unilateral pulmonary edema in the normally perfused lung occurring during periods of cardiac decompensation. This case complements the two reported by Kieffer and co-workers, where the unilateral pulmonary edema occurred in conjunction with the proximal interruption of a pulmonary artery.

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FIGURE 1. Chest x-ray picture taken during clinical acute pulmonary edema showing unilateral left-sided pulmonary edema with a clear right lung; the heart is enlarged. Note engorged hilar vessels and single wing butterfly pattern of alveolar infiltration.

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