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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Pulmonary edema confined to one lung is an unusual event and has been described only infrequently.1-5 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, i.e. acquired unilateral hyperlucent lung.6 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...
36.5°C rectally and respiratory rate was 30 per minute. Skin, mucous membranes and nails were deeply cyanotic. Pupils were constricted. There was marked jugular venous distention. Coarse rales were heard over the left lung only, while the right lung revealed only diminished breath sounds. Heart sounds were distant, poorly heard, and no gallop was discerned. There was no peripheral edema. This disparity in physical findings between right and left lungs, on review of previous examinations, proved to be a constant feature.

Laboratory Data

Chest x-ray film shown in Figure 1 confirmed the physical findings and demonstrated unilateral left-sided pulmonary edema with a clear right lung. Moderate cardiac enlargement was present. Characteristic engorgement of hilar vessels, distention of pulmonary venous radicals and diffuse alveolar infiltration in a single wing butterfly distribution are present.

Electrocardiogram showed regular sinus rhythm and complete left bundle branch block pattern. A complete blood count showed that the hematocrit was 43 percent with a hemoglobin of 14.6 gm. Total white blood count was 31,000 with 75 percent segmented neutrophils. Sputum culture showed group A Streptococcus. Remainder of laboratory data was noncontributory.

Hospital Course

The patient was treated with intravenous digitalis preparations, furosemide and ampicillin. He rapidly became alert; his dyspnea improved; cyanosis remitted and abnormal physical findings following a pronounced diuresis, remitted almost entirely. Chest x-ray film taken 12 hours later and shown in Figure 2 demonstrates dramatic clearing of the pulmonary edema pattern previously seen in the left lung. The appearance of the right lung is unchanged. There are no pleural effusions noted.

Figure 2. Chest x-ray film taken 12 hours after Figure 1 during clinical remission of edema showing complete clearing of the left pulmonary edema.

Figure 3. Lung scan shows absence of perfusion of the entire right lung field except for a small area of the middle portion.

Specialized Studies (Performed at Roosevelt Hospital, NYC, during a previous admission)

Tomograms of the right lung revealed marked diffuse emphysema of the entire right lung with decreased caliber of the blood vessels. No localized bullae were seen. Lung scan shown in Figure 3 demonstrated absence of perfusion of the entire right lung except for a tiny area in the mid-portion. One frame of the pulmonary angiography is shown in Figure 4. This revealed a normal-sized main pulmonary artery and normal-sized main left pulmonary artery with normal secondary and tertiary left pulmonary arterial branches. By contrast, the right main pulmonary artery is attenuated beginning at about 2 cm beyond its main trunk. The initial portion of the main right pulmonary artery is of normal caliber. All secondary branches of the right pulmonary ar-

Figure 4. Pulmonary angiography reveals normal left pulmonary circulation, normal origin of the right main pulmonary artery but marked attenuation of all secondary branches with minimal blood flow.
Pulmonary function studies revealed a reduced vital capacity at 73 percent of predicted with a decreased maximum breathing capacity and a decreased timed vital capacity (FEV1; 56 percent and FEV1/VC 77 percent of vital capacity). The total lung capacity was reduced to 85 percent of predicted and the RV/TLC ratio was 50 percent. Following administration of bronchodilators there was an increase in vital capacity to 88 percent of predicted but no increase in timed vital capacities.

These results are compatible with chronic obstructive lung disease with a bronchospastic component. Combined with the history of chronic cough and sputum production in a non-smoker, it would suggest the presence of air trapping and bronchiectasis as would be expected in Swyer-James syndrome. Cardiac catheterization of the right heart revealed a mean pressure in the right atrium of 8 mm Hg and at rest, pulmonary artery pressure of 40/18 mm Hg.

**DISCUSSION**

The diagnosis of unilateral pulmonary edema in this case is considered justified because of the sudden onset of respiratory distress, rales in the left lung and characteristic x-ray appearance which then cleared entirely within 12 hours following treatment with diuretics and digitals. Any unilateral process such as this may be caused by other conditions such as aspiration pneumonia, pneumonia or possibly pulmonary neoplasm. These latter conditions will enter the differential diagnosis and must be excluded before the diagnosis of acute pulmonary edema is made. In this particular case, the history, clinical onset, and hospital course effectively excluded all of the above.

The reason for the unilateral distribution of the pulmonary edema in this case probably depends upon the destruction of the pulmonary vascular bed of the right lung by a condition called "Swyer-James syndrome" or "MacLeod syndrome." Other explanations for localized pulmonary edema such as that of Gleason and Steiner who postulated that posture and gravity may have an influence, or that local variations in pulmonary venous pressure may occur, are not applicable in the present case since the repeated presentations were always with the same pattern on the left side. Unilateral pulmonary edema was also reported by Albers and Nadas following systemic pulmonary edema for cyanotic congenital heart diseases. Unilateral distribution can then be explained on the basis of local abnormalities and variations in pressures resulting from such a shunt.

Unilateral hyperlucent lung is caused by one of two conditions: (1) the acquired type of unilateral hyperlucent lung (unilateral pulmonary emphysema) Swyer-James or MacLeod syndromes; (2) congenital absence or hypoplasia of a pulmonary artery. The acquired type is attributed to (childhood) bronchiolitis, and bronchiolitis obliterans with dilatation and destruction of lung parenchyma occurring in various anatomic distributions within the lung, and there is usually a history of repeated respiratory tract infections in childhood. The basic etiology is infection perhaps due to an adenovirus.

X-ray findings include: (1) a significant difference in the radiolucency of the two lungs caused by decreased vascular perfusion in the affected lung; (2) decrease in the peripheral lung markings indicating narrowing and attenuation of vessels; (3) diminutive ipsilateral hilum. The hilum is present but small, a feature of great value in the differentiation from proximal interruption of the pulmonary artery (congenital absence).

The volume of lung is either normal or diminished depending on the age of patient at the time the infectious insult occurred, and the younger the patient at the time of pneumonia the smaller the lung since the insult prevents further maturation of the lung.

There is air-trapping during expiration which indicates the presence of bronchial obstruction. During expiration the mediastinum swings to the normal lung and excursions of the hemidiaphragms are markedly asymmetric. being diminished on the affected side. The density of the normal lung is greater because the blood flow is virtually the total output of the right ventricle.

Pulmonary angiography shows diminutive hilar vessels on the affected side. Bronchography shows a characteristic deformity of the bronchial tree, with segmental bronchi irregularly dilated and ending abruptly in a squared or tapered configuration at the fifth or sixth division. Clinically, Swyer-James syndrome patients may be asymptomatic or present with a variable clinical picture. Some complained of dyspnea on exertion or of experiencing repeated lower respiratory tract infections. Bronchspirometry shows reduction in ventilation by as much as 90 percent on the affected side and oxygen uptake is reduced to as low as 6 percent of normal.

The congenital absence of the pulmonary artery to one lung (pulmonary artery agenesis, also called proximal interruption of the pulmonary artery) is due to the interruption of the proximal portion of the primitive right or left sixth aortic arch. It is called proximal interruption since the vessels within the lung are usually intact and patent. The ipsilateral lung is hypoplastic, hyperlucent and of reduced volume and its arterial supply is derived from a hypertrophied bronchial circulation. X-ray examination in full expiration does not show the air-trapping seen in Swyer-James syndrome. The interrupted pulmonary artery is usually on the side opposite the aortic arch. When the proximal interruption is on the left, there is a high incidence of associated congenital anomalies especially tetralogy of Fallot, septal defects, and right-sided aortic arch. Associated congenital defects are much less frequent when the pulmonary artery anomaly is on the right.

**REFERENCES**

Leiomyosarcoma of the Inferior Vena Cava; An Unusual Cause of Pulmonary Embolism*

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A patient with leiomyosarcoma of the inferior vena cava is presented. The diagnosis was suggested by caviography and confirmed at necropsy. To the best of the authors’ knowledge, this is the first reported case with the clinical picture of pulmonary embolism.

Leiomyosarcoma is a rare malignant tumor which often arises from soft tissue, but it can also arise from viscera, particularly from the media of arteries and veins.1

In 1871, Perl2 published the first description of a sarcoma of the inferior vena cava. So far, only 20 cases have been reported in the literature.3,4 This report is concerned with the description of a further case present-

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Figure 1. Inferior ileo-caviography (anteroposterior incidence). Note hypoplasia of inferior vena cava in its proximal portion, the distal dilatation with enormous endoluminal formation reaching the atrium and the collaterality through renal and perivertebral veins.