Left Upper Extremity Edema, Rash, and Venous Varicosities in a 52-Year-Old Man*

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A 52-year-old man with a 20-pack-year smoking history presented with 4 days of nonproductive cough and dyspnea. Six months earlier, he had noted upper extremity swelling and an erythematous, non-pruritic rash predominantly on his left side. He was otherwise healthy except for a previous episode of peptic ulcer disease treated with a histamine H2 blocker. The patient denied occupational exposures.

Physical Examination

Vital signs: temperature, 37°C; pulse, 108/min (irregularly irregular); respirations, 18/min; BP, 110/60 mm Hg. General: alert and oriented. Skin: macular, erythematous, truncal rash that blanched with pressure; venous varicosities over the left shoulder. Chest: bibasilar crackles predominantly in the left lower lobe without wheezes. Cardiac: normal S1 and S2, grade 3/6 holosystolic ejection murmur at the apex with radiation to the left axilla. Extremities: 5-mm increase in the left upper arm circumference and 3-mm increase in left forearm circumference compared to the right arm.

Laboratory Findings

WBC, 6,300/µl with 64 percent polymorphonuclear leukocytes and 20 percent bands; hematocrit, 42.6 percent; platelets, 103,000/µl; total bilirubin, 2.2 mg/dl. ECG: atrial fibrillation. Chest radiograph: enlarged cardiac silhouette with prominent central vessels, especially on the left; moderate pulmonary congestion and small pleural effusions. Digital subtraction images from a left subclavian arteriogram are shown in Figure 1.

What is the vascular abnormality? What underlying syndrome explains the patient's clinical presentation?
Diagnosis: Subclavian artery-pulmonary artery arterioarterial malformation (possible variant of Klippel-Trenaunay-Weber syndrome)

Multiple congenital and acquired conditions are associated with abnormal communications between the pulmonary and systemic arterial circulations. Congenital malformations connecting the pulmonary arteries with intercostal, internal mammary, and bronchial arteries appear to develop when branches of the primitive dorsal aorta and the bronchial pulmonary plexus fail to regress during embryonic formation of the pulmonary artery.

Acquired intrathoracic arterioarterial communications are more common than congenital communications as causes of vascular malformations involving the pulmonary circulation. They typically result from disruption of thoracic vascular beds by inflammatory conditions that include actinomycesis, fungal infections, tuberculosis, and schistosomiasis, as well as malignant conditions such as Hodgkin’s disease and metastatic carcinoma.

Abnormal vascular communications involving the subclavian artery are distinctly uncommon. Most instances present as arteriovenous fistulas subsequent to vascular injury from indwelling catheters placed into the internal jugular or subclavian veins. To our knowledge, arterioarterial communications from the subclavian artery to the pulmonary artery, as occurred in the present patient, have not been previously reported.

The Klippel-Trenaunay-Weber syndrome is a unique condition associated with vascular malformations. Klippel and Trenaunay first observed, at the turn of the century, an unusual triad of venous varicosities, limb hypertrophy, and cutaneous hemangiomas. In 1907, Weber expanded these observations by noting the rare addition to the triad of arteriovenous fistulae. Although most contemporary instances of the syndrome affect the lower extremities, a single patient with an extrathoracic upper extremity arteriovenous fistula has been previously reported. Arterioarterial malformations may be an additional feature of the syndrome, as suggested by the present patient.

Vascular imaging is the cornerstone of diagnosis for patients with arterioarterial malformations affecting the pulmonary circulation. Doppler sonography can exclude abnormalities of extrapulmonary venous structures, such as intravascular thrombosis. Arteriography, however, remains the definitive study. Contrast infusion can visualize the malformation, and oximetry can detect a step-up in oxygenation within the pulmonary arteries.

No effective medical treatment exists for pulmonary arterioarterial malformations. Surgical management with excision of the malformation has been advocated in patients with large shunts, malformations under-going progressive enlargement, multiple well-localized lesions, and malformations with systemic blood supplies. Therapeutic embolization using woolen coils or balloon embolization has also been used to obliterate pulmonary arterioarterial malformations in patients at high risk for surgical resection.

The present patient underwent a left subclavian arteriogram that demonstrated rapid enhancement within the left upper lobe pulmonary arterial branches (Fig 1, left); later intense opacification of dilated, tortuous vessels in the left upper lobe (Fig 1, center); and still later retrograde filling of the left main pulmonary artery (Fig 1, right). These observations were consistent with an arterioarterial malformation between the left subclavian and axillary arteries and the pulmonary artery.

Cardiac catheterization determined the right ventricle and pulmonary artery pressures to be elevated with a step-up in oxygen saturation at the level of the left pulmonary artery. A two-dimensional echocardiogram with M-mode Doppler showed a dilated left ventricle and left atrium with normal systolic function, mitral regurgitation, and a narrow high-velocity jet flow from the left atrium to the right atrium through a small septum secundum atrial defect.

The patient refused surgical or embolization therapy. His dyspnea improved with diuresis and rate control of the atrial fibrillation.

Clinical Pearls

1. Systemic to pulmonary arterioarterial communications are unusual forms of intrapulmonary vascular malformation.

2. The unusual triad of venous varicosities, limb hypertrophy, and cutaneous hemangiomas combined with abnormal vascular communications defines the Klippel-Trenaunay-Weber syndrome.

3. The majority of systemic-to-pulmonary vascular malformations are acquired and result from infectious or neoplastic disorders.

4. Surgery is recommended for large shunts and malformations with progressive enlargement or systemic blood supplies. Embolization has been used in patients at high risk for surgery.

Suggested Reading

Klippel M, Trenaunay P. Neavus variqueux osteohypertrophique. Arch Gen Med 1900; 3:641-72