

Upper Extremity Occlusive Disease



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Meet Jane

Jane is a 62-year-old female. She is a long time smoker and has hypertension. She presents with a two-year history of Raynaud's phenomenon of her left hand. Since that time she has also noticed that her left arm starts to cramp with activities such as washing her hair and vacuuming, and at her work as a health care aide. Upon examination, she has a blood pressure of 160/90 in her right arm and a blood pressure of 90/60 in her left arm. Peripheral pulse examination of the left arm reveals no brachial, radial or ulnar pulses. Pulses are normal in her right hand. Her left hand looks otherwise normal. She is unable to work as a health care aide due to her symptoms.

Upper extremity occlusive disease is relatively uncommon and accounts for only 5% of all extremity ischemia. Unlike occlusive disease in the lower extremities, which is generally atherosclerotic in origin, the etiology of upper extremity occlusive disease is more varied and can include atherosclerotic disease, arteritis, connective tissue disorders, occupational injuries, and vasospastic diseases (Table 1).¹

Etiology

Upper extremity occlusive disease can involve the large and small arteries of the arm or a combination. Atherosclerosis is the most common cause of large artery disease and generally involves the subclavian artery, but disease may extend more distally, especially in the setting of diabetes. Patient risk

Table 1

Common Conditions Associated with Upper Extremity Ischemia

- Thromboembolism
- Atheromatous Embolization
- Atherosclerosis
- Connective Tissue Diseases
 - Scleroderma
 - Rheumatoid Arthritis
 - Sjögren's Syndrome
 - Systemic Lupus Erythematosus
- Large Artery Vasculitis
 - Takayasu's Disease
 - Giant Cell Arteritis
- Small Artery Vasculitis
 - Thromboangitis Obliterans (Buerger's disease)
- Blood Dyscrasias
- Myeloproliferative Disorders
- Thoracic Outlet Syndrome
- Latrogenic Injury
- Occupational Exposure
 - Hand-arm Vibration Syndrome
 - Hypothenar Hammer Syndrome
- Vasospasm
 - Raynaud's Disease
 - Cocaine Use
 - Amphetamine Use

factors are similar to those of other vascular beds. Tobacco use is almost universal. Hypercholesterolemia, hypertension, diabetes and



age are also important risk factors. Inflammatory diseases (giant cell and Takayasu's arteritis) can also involve the large vessels. Small vessel disease due to diabetes can lead to severe complications such as digital ulceration and gangrene.²

Raynaud's disease is a vasospastic disorder that produces a tri-phasic colour change in the hands or less commonly the feet, in response to cold or emotional stimuli. The hallmark of this disorder is pallor, cyanosis and post ischemic hyperemia of a digit or digits, although all three may not always be present. The disorder is classified as primary when no identifiable cause is evident or secondary when it is due to either a more proximal occlusion or other disease process resulting in digital artery vasospasm (Table 2).³ Patients whose occupations involve the use of vibrational tools or who use their hands as a hammer (especially the hypothenar eminence) can present with Raynaud's or frank digital ischemia from either hand-arm vibration syndrome or hypothenar hammer syndrome.

Occlusive lesions in the arm and hand secondary to scleroderma, rheumatoid arthritis, systemic lupus and other autoimmune disorders can lead to severe upper extremity ischemia manifesting as pain, ulceration and gangrene. Thromboangitis obliterans (Buerger's disease) is an inflammatory disease that can produce pain and ulceration in the fingers and toes.⁴ The patient is usually a young man with a long history of tobacco use.

Rare causes of upper extremity occlusive disease include iatrogenic injury, trauma, malignancy and frostbite.

Table 2

Differentiating Primary From Secondary Raynaud's Phenomenon

Type	Gender	Other Features
Primary	Usually Female	Age <45 Vasospasm of multiple digits Normal vascular examination No skin abnormalities Normal laboratory studies
Secondary	Male or Female	Any age Single digit Abnormal pulse exam Vascular lab studies abnormal Positive autoantibodies

Diagnosis

The initial evaluation for upper extremity occlusive disease starts with the history and physical examination. Unlike lower extremity occlusive disease, which is generally atherosclerotic, there are many pathologic conditions that may affect the upper extremity and a more detailed history is often required. The duration and speed of onset of symptoms is important in determining whether this is a chronic or acute condition. The presence and length of time Raynaud's has been present should also be noted. Symptoms of upper extremity claudication as well as symptoms of connective tissue disorders, such as swallowing difficulties, dry eyes and mouth, and arthritis, are significant. Cardiovascular risk factors along with occupational exposures should be documented.

Physical examination should include palpation of the axillary, brachial, radial and ulnar pulses. Finger cyanosis and tenderness should be noted along with digital ulceration and frank gangrene. Bilateral symptoms may suggest a systemic cause such as a connective tissue disorder. Unilateral symptoms may be from a discrete occlusive lesion. Bilateral blood pressures should also be taken.

Investigations

Patients suspected of having large vessel disease from history and physical examination can be evaluated further by segmental pressures in the vascular laboratory.⁵ A difference in blood pressures greater than 10mmHg compared to the contralateral extremity may be significant. Finally, if large vessel disease is symptomatic, imaging with a CT angiogram is warranted. CT angiograms are excellent at demonstrating proximal lesions, therefore, conventional angiography is generally reserved for treatment with balloon angioplasty and stenting or evaluation of more distal vessels in the forearm and hand.

For patients presenting with Raynaud's, either primary or secondary, it is reasonable to do screening laboratory work for a connective tissue disease (Table 3). While patients with primary Raynaud's by definition have no identifiable cause, approximately 5 to 8% may develop a connective tissue disease in the next 5 to 10 years. If serologic testing is positive, referral to a rheumatologist is indicated for future follow-up. For disease of the fingers, many vascular laboratories can also do finger systolic pressures. A difference of more than 15mmHg between fingers or an absolute pressure less than 70mmHg may indicate significant disease.

Treatment

More proximal disease is generally only treated if the patient is symptomatic. Reconstruction is either with balloon angioplasty/stent or with bypass surgery. Treatment of primary Raynaud's is generally conservative. Patients are told to minimize aggravating stimuli (cold exposure, stress). If the symptoms of primary Raynaud's are very frequent or severe, the first line treatment is calcium channel blockers (Nifedipine XL 30-60mg OD or Amlodipine 10mg OD).

Table 3

Laboratory Evaluation of Upper Extremity Ischemia

- Complete Blood Count
- Chemistry Profile
- Rheumatoid Factor
- Antinuclear Antibody
Anti-dsDNA
Extractable Nuclear Antibody
Anti-centromere Antibody
- Complement (C3,C4)
- C-reactive Protein

These drugs may lessen the number of events, but do not cure the disease. Treatment of occlusive disease secondary to connective tissue disorders is to control the underlying disease process. Ulcers and gangrene of the hands and fingers should be treated with local wound care and correction of any occlusive lesions if possible. Unfortunately, most occlusive lesions in the hand and fingers are not amenable to vascular reconstruction.

Case Conclusion

As Jane was symptomatic and could not work, she was referred to a vascular surgeon. A CT angiogram was performed and revealed a left subclavian artery occlusion. Angiography confirmed the lesion, but it was not amenable to balloon angioplasty. She therefore, underwent a carotid subclavian bypass. She has returned to work.

Take Home Message

1. Upper extremity occlusive disease is uncommon. Unlike lower extremity ischemia which is generally atherosclerotic in nature, the etiology of upper extremity ischemia is more diverse.



2. Treatment of primary Raynaud's is generally conservative. The treatment of secondary Raynaud's is to control and treat the underlying disease process.
3. Patients presenting with non-healing ulcers and gangrene need prompt evaluation and investigation.
4. Large vessel disease is only treated if the patient is symptomatic (claudication, pain, non healing ulcer and gangrene). Small vessel disease in the hands and fingers is usually not amenable to vascular reconstruction.

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Resources

1. Greenfield LJ, Rajagopalan S, Olin JW. Upper Extremity Arterial Disease. *Cardiol Clin.* 2002 Nov;20(4):623-631.
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3. Block JA, Sequeira W. Raynaud's Phenomenon. *Lancet* 2001 Jun 23;357(9273):2042-2048.
4. Olin JW. Thromboangiitis Obliterans (Buerger's disease). *N Engl J Med.* 2000 Sep 21;343(12):864-869.
5. Ouriel K. Noninvasive Diagnosis of Upper Extremity Vascular Disease. *Semin Vasc Surg.* 1998 Jun;11(2):54-59.

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