Case presentation

SUNY Downstate Medical center
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Case presentation

- 31y/o M who is a construction worker, transferred to the ER from Interfaith on 8/23

- h/o left hand ischemia for 12hrs with no neurological deficits.

- He reported as coolness of his left hand.

- PMHx:
  - was significant for HTN, NIDDM.
  - a similar episode 2 months ago – which resolved spontaneously.

- He was started on heparin drip on admission.
Physical examination

- Alert and oriented
- Chest:
  - S1, S2 N, no evidence of any murmur.
  - B/L BS equal, no crepts
- Abdomen: soft, no tenderness, no guarding
- Upper extremities:
  - B/l equal axillary and brachial pulse
  - No palpable radial or ulnar pulse
  - Doppler biphasic signals +
  - EQUAL BP on both extremities.
- Lower Extremities: NAD
- Rectal: guiac neg
Labs

- CBC – wnl
- Chem – wnl

Other:
- Factor V gene mutation (r506q) – neg
- Protein C – Normal
- Protein S – Normal
- Homocystiene - Normal
- Anticardiolipin Abs IgG / IgM – neg
- Lupus anticoagulant – neg
- Prothombin/ factor II (920210A mutation ) – neg
HD # 2  Diagnostic angiogram
Diagnostic angiogram
Diagnostic angiogram
Hospital course

- TpA initiated at the time of the diagnostic angiogram
  - ECHO – neg
- By HOD #3 patient was fully heparinized.
- Return of pulse (radial) – noted on HOD 4
- A follow up angiogram was performed.
Follow up angiogram – post tPA
Hospital course

- On HOD # 5, patient developed compartment syndrome of the left hand with decreased pulse and neurological symptoms.

- Patient was emergently taken for decompression and 2 compartment fasciotomy. Return of pulse noted at the time of surgery.

- Post-operatively patient made therapeutic on heparin.
Hospital course

- HOD # 9: patient grew staph. Epi in blood cultures.

- HOD # 13: continued to be febrile
  - Blood culture – Klebsiella

- HOD # 17: patient made therapeutic on coumadin.

- HOD # 18: d/c home
Upper Limb Ischemia

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Etiology

- Arterial embolism
  - Cardio-arterial embolization.
  - Arterio-arterial embolization.

- Arterial Thrombosis
  - Arthrosclerosis
  - Congenital anomaly.
  - Infection.
  - Hematological flow disorders.
  - Flow related disorders.
Etiology

- Arterial Trauma.
  - Blunt
  - Penetrating
  - Iatrogenic
- Drug induced vasospasm
- Aortic dissection
- Severe venous thromboplebitis
- Prolonged immobilization.
- Idiopathic
Etiology

- Special causes
  - Upper extremity aneurysms
  - Aberrant subclavian artery
  - Radiation arteritis
  - Thoracic outlet syndrome
  - Fibromuscular dysplasia
  - The Arteritides
    - Takayasu
    - Giant cell
History

- Pain
- History of previous episodes
  - Claudication / Exertional fatigue
- Cardiac disease
- Recent trauma
- HTN, back pain or chest pain.
- Drugs
- Low flow states
Examination

- 5 Ps
- Trophic changes, nail changes
- Complete bilateral pulse exams
- Assessment of limb viability
- Cardiac examination
- EKG
Radiology noninvasive

- CXR
- Abdominal x-ray
- Doppler velocity flow detection
- Duplex ultrasonography
Digital plethysmography or laser Doppler

- Doppler studies, when included with the physical examination, increase the accuracy of detecting occlusions or transections of arterial vessels in patients presenting with minimal signs of injury.

- ABIs are also useful studies that add to the accuracy of detecting arterial injury, especially when combined with Doppler studies.

- According to a study performed by Johansen and colleagues on a series of patients,
  - the negative predictive value for ABIs that exceeded 0.90 was 99%
  - the sensitivity and specificity for ABIs less than 0.90 were 95% and 97%, respectively, for major arterial injury
Radiology
Invasive

- Angiography
  - Indications
    - Determine site of the vascular obstruction
    - Suspect thrombosis
    - Suspect aortic dissection
    - Suspect multiple emboli

- MRA / MRV
Aortic Dissection
Aortic Dissection
Large vessel occlusive disease

- Usually localized to the subclavian and axillary artery
- Commonly associated with atherosclerosis
- Less commonly associated with thrombosis of an aneurysm
Aneurysms of subclavian and axillary

- Chronic subclavian artery trauma
- Radiation
- Post-stenotic dilation
- Thromboembolic
Subclavian aneurysm
# Takayasu disease

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**American College of Rheumatology 1990 Criteria for the Classification of Takayasu Arteritis**

<table>
<thead>
<tr>
<th>Criterion</th>
<th>Definition</th>
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<tbody>
<tr>
<td>Age at disease onset ≤ 40 years</td>
<td>Development of symptoms or findings related to Takayasu arteritis at age ≤ 40 years</td>
</tr>
<tr>
<td>Claudication of extremities</td>
<td>Development and worsening of fatigue and discomfort in muscles of one or more extremities while in use, especially the upper extremities</td>
</tr>
<tr>
<td>Decreased brachial artery pressure</td>
<td>Decreased pulsation of one or both brachial arteries</td>
</tr>
<tr>
<td>Blood pressure difference &gt; 10 mmHg</td>
<td>Difference of &gt; 10 mmHg in systolic blood pressure between arms</td>
</tr>
<tr>
<td>Bruit over subclavian arteries or aorta</td>
<td>Bruit audible on auscultation over one or both subclavian arteries or abdominal aorta</td>
</tr>
<tr>
<td>Arteriogram abnormality</td>
<td>Arteriographic narrowing or occlusion of the entire aorta, its primary branches, or large arteries in the proximal upper or lower extremities, not due to arteriosclerosis, fibromuscular dysplasia, or similar causes; changes usually focal or segmental</td>
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For purposes of classification, a patient shall be said to have Takayasu arteritis if at least three of these six criteria are present. The presence of any three or more criteria yields a sensitivity of 90.5 percent and a specificity of 97.8 percent.

### Distinguishing Features of Giant Cell versus Takayasu Arteritis

<table>
<thead>
<tr>
<th>Finding</th>
<th>Giant cell arteritis</th>
<th>Takayasu arteritis</th>
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</thead>
<tbody>
<tr>
<td>Female-to-male ratio</td>
<td>3:2</td>
<td>7:1</td>
</tr>
<tr>
<td>Age at onset</td>
<td>&gt;50 years</td>
<td>&lt;40 years</td>
</tr>
<tr>
<td>Ethnic ancestry</td>
<td>European</td>
<td>Asian</td>
</tr>
<tr>
<td>Histopathology</td>
<td>Granulomatous</td>
<td>Granulomatous</td>
</tr>
<tr>
<td></td>
<td>inflammation</td>
<td>inflammation</td>
</tr>
<tr>
<td>Primary vessels involved</td>
<td>External carotid artery branches</td>
<td>Aorta and branches</td>
</tr>
<tr>
<td>Renovascular hypertension</td>
<td>Rare</td>
<td>Common</td>
</tr>
<tr>
<td>HLA association</td>
<td>HLA-DR4</td>
<td>HLA-Bw52</td>
</tr>
<tr>
<td>Course</td>
<td>Self-limited</td>
<td>Chronic</td>
</tr>
<tr>
<td>Response to corticosteroids</td>
<td>Excellent</td>
<td>Excellent</td>
</tr>
<tr>
<td>Surgical intervention needed</td>
<td>Rare</td>
<td>Common</td>
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Subclavian steal syndrome

- Subclavian steal, which refers to retrograde flow in the vertebral artery due to an ipsilateral subclavian artery stenosis


- The following year, an association between this phenomenon and neurologic symptoms was noted.

- This has been termed the subclavian steal syndrome, suggesting that blood is stolen by the ipsilateral vertebral artery from the contralateral vertebral artery by way of the basilar artery.
Mechanism of subclavian steal

The subclavian artery stenosis results in lower pressure in the distal subclavian artery.

- Blood flows from the contralateral vertebral artery to the basilar artery, and then in a retrograde direction down the ipsilateral vertebral artery, away from the brain stem.

- Reversed vertebral artery flow, although it may have deleterious neurologic effects, serves as an important collateral artery for the arm in the setting of a significant stenosis or occlusion of the subclavian artery.
Subclavian steal - angiogram

**Angiogram of the aortic arch in the subclavian steal syndrome:** Angiography of the aortic arch demonstrates a significant stenosis of the right innominate artery (panel A). Panel B shows reversal of flow in the right vertebral artery. Courtesy of Peter Sp MD.
Retrograde flow
Management of subclavian steal syndrome

- The approach to therapy of subclavian steal associated with symptoms varies with the clinical setting.
- In many patients, symptoms improve over time without treatment.
- Extrathoracic revascularization has become the most popular form of surgical correction for symptomatic subclavian artery stenosis.
- Overall patency rates of 95 percent at one year, 86 percent at three years, and 73 percent at five years have been reported.
- Patency at five years is significantly higher for procedures utilizing the common carotid artery as the donor vessel compared to those using the contralateral subclavian or axillary arteries (83 versus 46 percent, p<0.01).
Conservative management

- More recently, percutaneous transluminal angioplasty, usually in combination with stent placement has been shown to be effective in patients with subclavian steal syndrome.
Conservative management

- A comparison between stenting and surgery demonstrates equal effectiveness, but shows fewer complications with angioplasty and stenting.

- The long-term patency rates with balloon angioplasty alone are inferior to that of extraanatomic bypass.
Surgical therapy

- Surgical treatment for patients with a subclavian steal and coexisting severe carotid stenosis is more controversial.

- A significant percentage of patients with subclavian steal have concomitant severe extracranial atherosclerotic disease,

- Carotid artery endarterectomy should be performed first and will often resolve all symptoms.

- Symptoms of arm or brain ischemia often subside after surgery.
Surgical therapy

- Symptomatic patients with primary proximal subclavian artery ulcerative disease complicated by embolization into the vertebrobasilar system.
In the absence of other significant cerebrovascular disease, be successfully treated by surgical removal or exclusion of the proximal subclavian site or by anticoagulation.
Axillo-axillary bypass

Axillo-axillary bypass is an alternative method for revascularization in patients at high surgical risk for subclavian steal syndrome.
Thoracic outlet syndrome

**Definition**

- Compression of the subclavian vessels and brachial plexus at the superior aperture of the chest, most commonly against the first rib.

Other terms for this syndrome include scalenus anticus syndrome, costoclavicular syndrome, hyperabduction syndrome, cervical rib syndrome, and first thoracic rib syndrome.
Thoracic outlet syndrome

**Etiology**

- There are many factors which can cause neurovascular compression at the thoracic outlet.

- Bony abnormalities are present in about 30% of patients, and some of these may be visualized on plain chest x-ray.

- Anatomic Factors ·
  - Interscalene compression ·
  - Costoclavicular compression ·
  - Subcoracoid compression.
Thoracic outlet syndrome

- Congenital Factors
  - Cervical rib
  - Rudimentary first rib
  - Scalene muscle abnormalities
    - Fibrous bands
    - Bifid clavicle
    - First rib exostosis
    - Enlarged C7 transverse process
  - Omohyoid muscle abnormalities
  - Anomalous transverse cervical artery
  - Postfixed brachial plexus
  - Flat clavicle

[Image of X-ray of thoracic region]
Other causes

- Traumatic Factors ·
  - Fractured clavicle ·
  - Humeral head dislocation ·
  - Upper thorax crush injury ·
  - Sudden effort of shoulder girdle muscles ·
  - C-spine injuries/cervical spondylosis
Vascular manifestations of TOS

- Pain is usually diffuse and associated with coldness, weakness, and easy fatigability of the hand and arm.

- Unilateral Raynaud's phenomenon in about 7.5% of patients, which can be precipitated by hyperabduction or carrying heavy objects.

- There may be signs of distal embolization, poststenotic dilation or aneurysm of the subclavian artery, or true arterial occlusion.

- Venous obstruction is much less common and is known as "effort thrombosis" or "Paget-Schroetter syndrome". The affected arm is edematous, discolored, and aches.
Thoracic outlet syndrome
Surgical approach

- Transaxillary first rib resection avoids division of major muscle groups, ensures complete removal of the first rib, and has the best cosmetic result.

- Position the patient in the lateral position with the affected arm abducted 90 degrees and loosely suspended (straight up to the ceiling).
Surgical management

Transverse incision in the axilla between pectoralis major and latissimus dorsi.

Dissect along the external thoracic fascia to the first rib.

Divide the scalenus anticus at its insertion on the rib.
Surgical management

- Remove middle and anterior portion of first rib after periosteal elevation.

- Divide costoclavicular ligament and remove posterior portion of first rib.

- Always protect the brachial plexus and vessels. Remove the entire first rib, as any residual portion may cause recurrence.
Small vessel arterial disease

- Occupational hand trauma
  - Hypothenar hammer syndrome
  - Vibration induced white finger
- TAO
- Thrombosis
- Embolic
- Raynaud’s disease / phenomenon
TAO

- Thromboangiitis obliterans is a nonatherosclerotic, segmental, inflammatory disease that most commonly affects the small and medium sized arteries, veins, and nerves of the extremities.

- The use of tobacco is the sine qua non of disease initiation and progression.
There are several criteria proposed for the diagnosis of thromboangiitis obliterans, based upon a clinical, angiographic, histopathological, and exclusionary scoring system.

One set of criteria includes onset before the age of 45 years in the absence of risk factors for atherosclerotic disease other than smoking.
Angiogram of the hand in a patient with Buerger’s disease. The angiogram shows multiple occlusions of the digital arteries, with collaterization “corkscrew collaterals” around the areas of occlusion (arrows).

Treatment

- Vasodilators
- Calcium channel blockers
- Systemic anticoagulation
  - Heparin
  - coumadin
- tPA
- Sympathectomy
- Angioplasty +/- stent placement
- Thrombectomy
The tPA molecule is predominantly an endothelial cell enzyme.

Its release is stimulated by a variety of substances including thrombin, serotonin, bradykinin, cytokines, and epinephrine.

In plasma it circulates as a complex with its natural inhibitor PAI-1 and is rapidly cleared by the liver.
tPA

Mechanism of action

- To restore vessel patency following hemostasis, the clot must be organized and removed by the proteolytic enzyme plasmin in conjunction with wound healing and tissue remodeling.

- Plasminogen, the precursor molecule to plasmin, binds fibrin and tissue plasminogen activator (tPA).

- This ternary complex leads to conversion of the proenzyme plasminogen to active, proteolytic plasmin.
The Fibrinolytic pathway

Activators:
- Urokinase-type plasminogen activator
- Tissue-type plasminogen activator

Plasminogen → Plasmin

Inhibitors:
- Plasminogen activator inhibitors (PAI-1 and PAI-2)
- Alpha-1 antitrypsin
- Alpha-2-antiplasmin
- Thrombin-activatable fibrinolysis inhibitor

Fibrin → Fibrin degradation products
(D-dimer, others)

The fibrinolytic pathway: Plasminogen, the precursor molecule to plasmin, binds fibrin and one of the two plasminogen activators. This ternary complex leads to conversion of the proenzyme plasminogen to active, proteolytic plasmin. Plasmin cleaves the polymerized fibrin strand at multiple sites and releases fibrin degradation products (FDPs). One of the major FDPs is D-dimer, which consists of two D domains from adjacent fibrin monomers that have been crosslinked by activated factor XIII. Plasmin has broad substrate specificity and, in addition to fibrin, cleaves fibrinogen and a variety of plasma proteins and clotting factors. Diagram supplied by Gregory YH Lip, MD.
tPA

Analogous to the prothrombin complex, the rapid generation of plasmin by tPA optimally takes place on a surface, the fibrin clot.

Both tPA and fibrinogen bind to fibrin via recognition of lysine residues in the fibrin clot.

When bound to fibrin, the binding interaction aligns tPA and plasminogen on the fibrin surface so that the catalytic efficiency of tPA is increased several hundredfold.
Complications

- Occlusion and bleeding from thrombosis are common early complications in the postoperative period. These complications necessitate an immediate reoperation.

- Muscle edema causing increased compartmental pressure is another complication of vascular injury, with pain being the most important symptom.

- Decompression of the fascial compartments (fasciotomy) is performed to treat this process.

- Nerve injury causing motor or sensory deficits is another complication that may lead to limb disability.
Complications

- Tissue death and necrosis are complications of prolonged periods of vascular compromise and limb ischemia.

- Amputation of the necrotized part is usually the method of treatment.

- Another serious complication of vascular injury is infection, which requires immediate debridement and antibiotic treatment.

- Late complications of arterial injury include arteriovenous fistulas and false aneurysms. These complications are usually managed by operative repair.
Conclusion

- Although upper extremity ischemia occurs less frequently than its Lower extremity counterpart, very satisfying results are noted with surgical management.

- The operative M &M are low and the long term durability is excellent in properly selected patients.

- Non-operative management is successful in many patients.
  - Therefore unless the viability of the limb is acutely threatened, a period of conservative management and observation should be followed.