Case 3005

A case of thromboangiitis obliterans or Buerger’s disease.
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Section: Cardiovascular
Imaging Technique: Ultrasound-Colour Doppler
Case Type: Clinical Cases
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Patient: 40 years, male

Clinical History:

A 40-year-old male smoker presented with a history of Raynaud’s phenomenon, two-month ischemic pain followed by gangrene of distal portion of the left index finger and the right index and medium fingers.

Imaging Findings:

A 40-year-old man with a two-year history of Raynaud’s phenomenon of both the hands presented with gangrene of the distal phalanxes of the left third finger and the right second and third fingers, preceded by a two-month ischemic pain. The patient smoked 20–30 cigarettes per day for 25 years. On physical examination, it was found that the radial and ulnar pulses were absent on both arms, while the brachial pulse was present bilaterally. Blood pressure was recorded as 125/75 mmHg. Laboratory tests were found to be normal, particularly plasma levels of glucose, lipid, complement, homocystein, protein C, protein S and antithrombin III were found to be within normal limits. Antinuclear antibodies, rheumatoid factor, anti-Scl 70 and anti-phospholipid antibodies were absent. Echocardiography performed on the patient showed that embolic source was absent in the heart. Color-Doppler sonography (CDS) revealed segmental occlusions of the distal radial and ulnar arteries on both sides, while subclavian, axillary and brachial arteries found to be normal. Digital subtraction angiography of the upper extremities confirmed CDS findings and showed multiple occlusions of digital branches. The patient stopped smoking and was treated with amiodipine, low-molecular-weight heparin and endovenous iloprost administration. Even if ischemic symptoms regressed, amputation of gangrenous distal phalanxes was required.

Discussion:

Thromboangiitis obliterans (TAO) or Buerger’s disease is a non-atherosclerotic, segmental, occlusive inflammatory disease of the small and medium-sized arteries and veins. This disease mainly affects the extremity vessels and rarely affects the visceral and cerebral ones. TAO is more prevalent in the Far East and Middle East than in North America and Western Europe. It usually occurs in 20–50-year-old male smokers, but its prevalence among women is increasing. The etiology of TAO is unclear, however the usage of tobacco has a central role in its initiation and progression. In fact, it is well known that almost all the patients had a history of smoking, and that remission, relapses and worsening correlate with the patient’s smoking habits. HLA associations, vascular endothelial dysfunction and high titers of anti-endothelial-cell antibody have also been reported. Pathologically, TAO is characterized, in the acute phase, by occlusive, highly cellular, inflammatory thrombi with lymphocytic infiltration of the intima and media. Internal elastic lamina and structures of vascular wall are relatively preserved. In the chronic phase, organized thrombi with partial recanalization and mild vascular wall fibrosis are observed. The patient generally presents with claudication of distal extremities. This disease often involves more than one extremity and the lower extremities are more frequently affected than the upper ones. With the progression of the disease, signs
and symptoms such as ischemic pain at rest, ischemic ulcerations and gangrene occur on the toes, feet or fingers. Superficial thromboflebitis, Raynaud’s phenomenon and sensory abnormalities are not uncommon. On physical examination, it was found that proximal pulses were strong, while distal pulses were absent. Laboratory tests revealed that blood cell counts were normal and that plasma levels of glucose, lipid, acute-phase reactants, complement, homocystein, protein C, protein S and antithrombin III were also normal. Antinuclear antibodies, rheumatoid factor and cryoglobulins were absent. Digital subtraction angiography revealed multiple segmental occlusive lesions affecting the distal arteries, associated with corkscrew collaterals. Typically, proximal arteries were normal. These findings, even if suggestive of TAO, are not pathognomonic, because they may be present in scleroderma, systemic lupus erythematosus, rheumatoid vasculitis and in other vascular diseases. The diagnosis of TAO is based on clinical findings and on exclusion of other ischemic diseases, like atherosclerotic occlusive disease, collagen diseases, hypercoagulopathy and distal embolization. Angiography and biopsy with histopathological examination may contribute to make diagnosis. The most effective therapeutic choice to prevent the progression of the disease and to avoid amputation is complete smoking cessation. Medical treatment is generally based on intravascular or oral administration of prostaglandin analogues. Anticoagulant, antiplatelet and thrombolytic agents and vasodilators were also used in the treatment of the disease. The distal nature of the disease often does not allow surgical revascularization. The role of sympathectomy in preventing amputation or in treating pain remains unclear. Amputation may be required in advanced cases and where medical or surgical therapy has failed.

**Differential Diagnosis List:** Thromboangiitis obliterans or Buerger’s disease.

**Final Diagnosis:** Thromboangiitis obliterans or Buerger’s disease.

**References:**

Figure 1

Description: The scan showing the presence of a severe caliber reduction and of an obstruction in the right ulnar artery at the wrist. Origin:
Description: Angiogram confirming the severe caliber reduction and the obstruction of the distal ulnar artery. The palmar arch, supplied by collateral artery from the interosseous artery, presents with multiple segmental obstructions. Origin:
Description: Angiogram showing several occlusions of digital arteries. Corkscrew collaterals are also evident. Origin:
Description: The image demonstrating the distal occlusion of the radial artery. Origin:
Figure 3

Description: Angiogram depicting obstruction of ulnar and radial arteries at the level of the wrist. Palmar arch is supplied by ulnar artery collateral. Origin:
Description: Several occlusions of digital arteries with corkscrew collaterals are shown. Origin: