Although tobacco use has declined in recent decades in many parts of the world, smoking is still common in a number of countries. Many smokers are unaware of the link between tobacco smoking and cancerous ulcers. Almost 5 out of every 10 cases of Buerger's syndrome can be attributed to tobacco use.

Introduction to Buerger’s Disease

In 1908, a unique disease was described by Buerger. A detailed analysis based on his pathological conclusions is still used today. Buerger’s disease is a chronic disease caused by thrombosis, i.e., severe inflammation and blood clotting, especially in the veins and arteries. The bodily parts most affected by this painful disease are the hands and feet.

Buerger’s disease is also known as thromboangiitis obliterans. It is a very unusual disease that interrupts blood circulation along specific arteries or veins of the legs and arms.

It is common for Buerger's disease patients to smoke cigarettes or other types of tobacco.

Definition of Buerger’s Disease
Buerger’s disease as inflammation of the blood vessels

Buerger’s disease is persistent. It is characterized by fragmental inflammation and thrombosis, which causes severe damage of small- to medium-sized arteries and veins, eventually affecting the peripheral structure of the lower and upper limbs.

Blood clotting inflames the patient’s blood vessels, causing swelling and blockage in the vascular structure of the body. Thrombi (blood clots) ultimately cause destruction and damage to the skin tissues, leading to infectious gangrene, which starts from the tips of the toes and eventually spreads to the legs and arms.

Risk Factors of Buerger’s Disease

Buerger’s disease is thought to be autoimmune in nature. The attributed cause is undefined, but the consumption of tobacco is the main culprit. The pathophysiological factors thought to be responsible are platelets, leukocytes, endothelial cells, and sensory neurons. The disease is caused by inflammation and swelling of small blood vessels.

Additionally, possible triggers for the disease include genetic factors, male gender, mental stress, and infectious agents. Cardiovascular disease and glucose intolerance in diabetic patients may also be triggers.
Pathophysiology of Buerger’s Disease

Although thromboangiitis obliterans is considered to be idiopathic, some causes may stem from genetic alterations.

Inflammatory endarteritis is a severe state of thromboangiitis obliterans. It causes vasculopathy inflammation, in which a prothrombotic state is generated that eventually leads to a vaso-occlusive state. Initiation of the inflammation process occurs within the tunica intima. This condition is closely linked with heavy tobacco use, and its permanence leads to expansion of the peripheral blood vessel structures.

Mild to severe claudication patients often experience critical limb ischemia, attributing to tissue loss and rest pain. Symptoms of sensitive limb ischemia include paresthesia, poikilothermia, paresis, pallor, mottling, and lack of pulse.

The chief pathophysiology of these Buerger’s disease symptoms appears when vessels become inflamed and form clots. This may result in obstructed blood flow throughout the tissues of the body, leading to the death of tissues due to lack of oxygen and nutrients. Ultimately, this produces vasculitis as well as ischemic changes in the extremities.

Buerger’s disease affects people who smoke 1–2 packs of cigarettes daily and who are hypersensitive to tobacco extracts when injected under the skin. As well, they have an amplified cellular predilection toward peripheral endothelium-dependent vasorelaxation and collagen types I and III. Patients also display a complex frequency of leukocyte antigen (HLA)—A9, HLA-B5, and HLA-A54, thus signifying a genetic element to the disease.

Clinical Features of Buerger’s Disease

Buerger’s disease commonly initiates with lethargy and pain in the affected areas. Symptoms may include:

- Pain in the hands, legs, and feet that occurs sporadically, along with open sores on the hands and feet
- Pale fingers or toes at low temperatures
- Swollen veins
Further signs and symptoms may appear as a result of decreased blood supply:

- Alteration in the skin color of the lower and upper limbs, with shininess or thickness
- Weak pulse in the leg and foot
- Lack of blood flow, causing the death of tissues (gangrene)
- Non-healing ulcers and wounds appearing on the leg and foot
- Reduction of hair growth on the lower limbs
- Bluish toes
- Severe burning sensation and pain in the toes
- Pain and cramps in the leg while the patient is lying down
- Feeling of numbness and heaviness in legs and toes
- Reddish-blue color of the lower limbs
- Thick and opaque toenails

If a patient experiences any signs and symptoms of Buerger’s disease it is essential to seek medical attention. It should not be assumed that these signs are merely due to the aging process.

**Investigations and Diagnosis of Buerger’s Disease**

The majority of patients affected with Buerger’s disease are between the ages of 20 and 45. This ailment rarely affects either elderly or pediatric patients. Men are three times more likely to have Buerger’s disease than women. However, with the increasing number of female smokers, the disease rate in women is assumed to rise.

Because the signs and symptoms of this disease may resemble those of many other diseases, a detailed assessment by a physician is required to make a definitive diagnosis. There is no clear test to assess if a patient is suffering from arterial disease, but there are certain tests a physician can carry out to exclude other diseases, including a simple blood sample test.

An additional assessment that a physician may use is the Allen test. This is a test in which the patient is asked to tighten their fist while pressure is applied to an artery of the hand. When the fist is opened, the physician will release the increased pressure from the artery and the hand should look paler than before it was tightened. If the paleness disperses very slowly, this may be a sign of Buerger’s disease.

**Physicians should look for five key signs when making a diagnosis of Buerger’s disease:**

- Regular use of tobacco
- Pain during rest
- Ulceration of the feet and hands, especially before the age of 50
- Blockage of arteries
- Blockage of artery or clot formation with an unknown cause

**An analysis based on certain criteria is recommended:**

- Age is less than 50 years
- Recent or present history of tobacco
- Existence of infrapopliteal arterial occlusive ailment signified by pain while resting, claudication, gangrenes, or ischaemic ulcers (assessed through a noninvasive vascular test)
- Either phlebitis migrans or upper-limb participation
- No atherosclerotic risk issues (except for smoking)
- Hypercoagulable states
- Autoimmune diseases
- Proximal source of emboli by echocardiography or arteriography
- Diabetes mellitus

There is no recommended route of investigation, but the following tests should be used to diagnose the root cause.

**Angiogram**

This is considered the most common test for arterial blockage along with X-rays, which are taken after the inoculation of dyes.

**Serological**

There are no particular serological indicators to identify Buerger’s disease. Suggested tests to uncover other reasons for vasculitis include liver function tests, full blood count, fasting glucose, renal function tests, c-reactive protein, erythrocyte sedimentation rate, and autoantibodies.

**Angiography**

Some angiographic characteristics are typical of Buerger’s disease (though not pathognomonic), including “corkscrew” arteries resulting from vascular damage, mainly the arteries in the ankles and wrists. Angiograms can also highlight stenoses or occlusions in different areas of both the legs and arms. Sometimes it may be necessary to conduct angiograms on other parts of the patient’s body, such as a mesenteric angiogram, to exclude other types of vasculitis that include vascular areas commonly affected in Buerger’s disease.
Doppler ultrasound

This test can be used to exclude Buerger’s disease and other reasons for secondary Raynaud’s color.

Echocardiography

This test can eliminate the source of recurring emboli.

Others

Due to the risk of cancerous elements, sometimes a biopsy in non-healing areas is required. Skin biopsies are very rarely performed, however.

Therapy of Buerger’s Disease

Treatment of Buerger’s disease

Pharmacological therapy is usually ineffective; quitting smoking is the only known way to prevent disease progression. In the medium- and small-sized arterial vessels, the chances of endovascular revascularization or surgery are very low due to the lack of a distal objective for revascularization. As the disease progresses, the only possible option is amputation.

There is no satisfying treatment for Buerger’s disease yet. However, the signs and symptoms of this disease can be controlled and treated by increasing blood circulation, quitting smoking, moving to a warmer climate, and, in certain situations, performing surgery on affected nerves. The nerves are surgically severed to stop the pain via a procedure called sympathectomy. Circulation is increased by taking an adequate amount of liquids and getting regular exercise.

Vascular surgery

Vascular surgery is a process in which a bypass of the blocked vein is implanted. A vein is taken from a different part of the patient’s body or an artificial grafting is used, which is then connected to the affected vein, thus allowing the blood to bypass the constricted area.

Patients should avoid smoking and should also stay away from secondhand smoke.

Review Questions

The correct answers can be found below the references.

1. What is the underlying pathophysiological process in Buerger’s disease?
   
   A. Inflammation process
   B. Allergic reaction
   C. Bacterial invasion
   D. Fungal infection

2. A 30-year-old smoker complains of soreness in the fingertips. On examination, ulceration and gangrene are identified. What would be the
possible diagnosis?

A. Diabetes
B. Cardiovascular Disease
C. Buerger’s Disease
D. Raynaud’s Disease

3. What should be the best possible diagnostic criteria for Buerger’s Disease?

A. Existence of atherosclerotic
B. Proximal Emboli
C. Non-hypercoagulable state
D. Diabetes insipidus

References


Correct answers: 1A, 2C, 3B

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