Buerger's Disease

What is Buerger's Disease?
Buerger's (also known as Thromboangiitis obliterans) is a disease of the small and medium arteries and veins that restricts blood flow to the hands and feet. Clots (thrombus) develop inside the blood vessels. This in turn leads to skin ulcers and gangrene in the fingers and toes and numbness and tingling if the nerves are affected. It is not uncommon for toes, fingers and limbs to need to be amputated if the gangrene progresses. It is not clear whether this is a true vasculitis.

Who are affected?
The disease is seen almost exclusively in smokers, mainly in young men aged 20-40 years. Recently, however, a higher percentage of women and patients over 50 have been diagnosed.

What are the symptoms?
The initial symptoms include claudication (pain induced by insufficient blood flow during exercise) in the feet and or the hands. The pain usually begins in the extremities but may radiate to other parts of the body. Patients may experience numbness and tingling in the limbs and also Raynaud's Phenomenon. Raynaud's is a condition where the extremities of the hands and feet turn white when exposed to cold. A common sign in Buerger's is skin ulceration and gangrene of the fingers and toes.

What is the aetiology (cause)?
Use of tobacco, particularly cigarette smoking, is the overwhelming factor predisposing to a diagnosis of Buerger's Disease. The majority of patients are heavy smokers, although some cases have been reported in moderate smokers and even in patients using chewing tobacco or other forms of tobacco that are not smoked. It is thought that this disease is triggered by some constituent of tobacco.

Diagnosis
Angiograms of the limbs can be helpful in making a diagnosis of the disease. Biopsies are not usually recommended as a diagnostic tool for Buerger's Disease because of the possibility that the biopsy area may not heal well. Typically blood tests are normal and do not show any evidence of inflammation.
Treatment

It is essential that people affected with Buerger's Disease stop smoking completely and immediately. Continued smoking or use of other forms of tobacco, even small amounts, increases the risk of losing fingers, toes or limbs due to necessary amputation.

Although there is evidence of inflammation inside the vessels and within the blood clots this differs from other types of vasculitis were the inflammation is in the blood vessel wall. Anti-inflammatory and immunosuppressant treatments have not been shown to be effective in Buerger's Disease. Other treatments such as iloprost or prostacyclin that help to open up the blood vessels may be helpful.

Side effects

For information on drugs used in the treatment of vasculitis and their side effects see Glossary of drugs and side effects.

Prognosis

Good with correct treatment and complete cessation of smoking and tobacco use.

Key points

- Buerger's disease is associated with the use of tobacco.
- The most important treatment is stopping smoking.

Pharmacological treatment for Buerger's disease

Background

Buerger's disease is characterized by recurring progressive inflammation and clotting in small and medium arteries and veins of the hands and feet. Its cause is unknown, but it is most common in men with a history of tobacco use. It is responsible for ulcers and extreme pain in the limbs of young smokers. In many cases, mainly in patients with the most severe form, there is no possibility of improving the condition with surgery, and therefore, drugs (pharmacological agents) are used. These can be pharmacological agents, such as cilostazol, clopidogrel, and pentoxifylline, or medicine derivatives of prostacyclin and prostaglandin, which redirect blood flow and improve the circulation in affected areas, and theoretically, help to heal ulcers and relieve rest pain. This review assessed the effectiveness of pharmacological agents in the treatment of patients with Buerger's disease.

Key results

Our search identified five randomised controlled trials, with a total of 602 participants and a treatment period of around four weeks (evidence current until April 2015). The comparisons included prostacyclin analogue versus placebo, aspirin, and a prostaglandin analogue, and folic acid versus placebo. We did not identify studies that assessed pharmacological agents such as cilostazol, clopidogrel and pentoxifylline, or studies that compared oral prostanoid versus intravenous prostanoid. The included studies assessed derivatives of prostacyclin and prostaglandin, which have the ability to redirect blood flow and improve the circulation in affected areas.

Moderate quality evidence from one study suggested that intravenous iloprost was
effective in healing ulcers and relieving rest pain after 28 days of treatment when compared with oral aspirin, but no differences were found in the rates of amputation. Evidence from two studies suggested that prostacyclin was as effective as prostaglandin analogues in healing ulcers (very low quality evidence) and eradicating pain at rest (low quality evidence), but rates of amputation were not assessed. Moderate quality evidence from one study suggested that there was no difference between placebo and the oral prostacyclin analogue iloprost (200 mcg and 400 mcg) in healing ischaemic ulcers or eradicating pain at rest after eight weeks and six months, and rates of amputation after six months. Very-low quality evidence from one study showed no difference between placebo and folic acid, in patients with Buerger’s disease and hyperhomocysteinaemia (a medical condition characterized by abnormally high level of homocysteine in the blood), in rates of amputation and pain scores. Ulcer healing was not measured. Treatment side effects, such as headaches or nausea, did not result in treatment interruptions or more serious consequences. Outcomes such as amputation-free survival, walking distance or pain-free walking distance, and ankle brachial index were not assessed by any study.

Quality of the evidence

Overall, the quality of the evidence was very low to moderate, with few studies, small numbers of participants, variation in severity of disease of participants between studies and missing information regarding for example baseline tobacco exposure. High quality trials assessing the effectiveness of pharmacological agents (intravenous or oral) in people with Buerger’s disease are needed.

Related Vasculitis Articles

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Related Vasculitis Articles

- Fertility and Vasculitis - Dr David Jayne

Further reading

- Buerger’s disease/Thrombosis obliterans - Gregory Piazza, Mark A. Creager