Buerger’s disease: a multidisciplinary diagnostic and therapeutic challenge

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Buerger’s disease, also known as thromboangitis obliterans, is a rare condition that is characterized by vascular thrombosis, and which in the past almost exclusively affected men.

Introduction

We describe a case of Buerger’s in a 43-year-old female smoker who initially presented with painful lumps on her limbs misdiagnosed as panniculitis. The case highlights the diagnostic and therapeutic challenges faced by the multidisciplinary team involved in her care, and how a holistic approach has allowed her to maintain her independence.

Case report

Diagnostic challenge

Our patient is a right-handed 43-year-old unemployed English woman who was seen for the first time in the vascular clinic 14 years ago. She initially presented to her local hospital in 1992 with a number of painful lumps on her feet and arms which were diagnosed as panniculitis. She had a 22-pack-year history having started smoking at the age of 14. She had no other risk factors for arterial disease.

She was then referred to a tertiary centre for rheumatology in 1996 where Buerger’s disease was clinically suspected. At this time, Buerger’s was uncommon in women, and to confirm the diagnosis she underwent a series of further investigations, to exclude other possible causes, namely the classical vasculitides.

In August 1997, while undergoing investigations, she complained of a painful right hallux, which was thought to be due to vasculitis or vasospasm. She was commenced on an iloprost infusion, but this was unsuccessful in controlling her symptoms. Over a four-week period, she developed gangrene in two of her right toes that necessitated amputation. However, the tissue loss and pain worsened, and she underwent a right-sided below-knee amputation. These amputation specimens histologically confirmed her diagnosis.

Therapeutic challenge

To date, having undergone a total of 45 operations, our patient has bilateral above-knee amputations, a right mid-forearm amputation, and a non-dominant left hand with two amputated digits. A timeline of her operations is shown in Figure 1.

Despite her significant physical impairment, she lives alone in a ground floor flat which has been modified. Our patient has three children, who are very supportive, helping with her shopping. She is otherwise independent with her activities of daily living and has no formal care package. Our patient mobilizes using a wheelchair and drives an adapted car. She is able to perform her everyday tasks through the provision of prostheses by limb-fitting services, rehabilitation provided by physiotherapists, and the use of special equipment at home provided for her by occupational therapy. The involvement of the different members of the multidisciplinary team is summarized in Table 1.

Our patient has insight into smoking as a significant contributing factor to the progression of her Buerger’s disease; however, she has tried to quit smoking on numerous occasions. The longest period she has not smoked for has been eight months in the past 14 years. She has tried support classes, oral and topical nicotine replacement, and alternative therapies to no avail.
Figure 1
Summary of the operations which our patient has had performed. Other operations include a left femoral endarterectomy, upper and lower limb nerve blocks and sympathectomies, and nail avulsions

| Operation date | 1981 | 1995 | 1997 | 1999 | 2001 | 2002 | 2003 | 2004 | 2008 | 2009 | 2010 | Total |
|---------------|-----|-----|-----|-----|-----|-----|-----|-----|-----|-----|-----|-----|
| Major amputation |    |     |     |     |     | 6  |     |     |     |     |     | 6    |
| Minor amputation |    |     |     |     |     | 10 |     |     |     |     |     | 10   |
| Other |     |     |     |     |     | 29 |     |     |     |     |     | 29   |
| Total |     |     |     |     |     | 45 |     |     |     |     |     | 45   |

Table 1
The different members of the multidisciplinary team involved in the care of our patient, in chronological order, and commonly used pharmacological interventions employed in her management. The professions and pharmacotherapeutics highlighted in italics have been involved with her care and utilized, respectively, on a number of occasions

| Profession                              | Involvement in our patient’s care                                                                 |
|-----------------------------------------|-------------------------------------------------------------------------------------------------|
| Rheumatology                            | Initially for the investigation of a vasculitis. Subsequent referrals for treatment and to re-confirm the diagnosis of Buerger’s disease in 2002 and 2008 |
| Vascular surgery                        | For the assessment of her vasculature and to plan and implement surgical and non-surgical re-vascularization strategies; also to manage tissue loss |
| Anaesthetics                            | Preoperative assessment and administration of anaesthesia for operations                        |
| Gastroenterology                        | Perform investigations for gastrointestinal symptoms and to exclude inflammatory bowel disease |
| Vascular scientists                     | Regular duplex ultrasonographic assessment of both arterial and venous systems                   |
| Radiology                               | Performing and interpreting angiography and tomographic imaging                                 |
| Pain team                               | Regular assessment of her pain control postoperatively and during her rehabilitation; this includes the management of phantom limb syndrome |
| Limb-fitting services                   | Fittings with prostheses                                                                         |
| Physiotherapy and rehabilitation        | Provided her with rehabilitation and exercise plans following her operations; referred her to occupational therapy for household modifications |
| Dermatology                             | Diagnosis of cutaneous manifestations of her Buerger’s disease                                   |
| Neurology                               | Assessment and management of chronic lumbar back pain                                            |
| Spinal surgery                          | Review of chronic lumbar pain; advised that no surgical intervention appropriate                 |
| Dieticians                              | Regular reviews of her nutritional status, especially during her prolonged admissions complicated by depression |
| Psychiatry                              | Assessment of our patient’s low mood                                                               |
| District nurses                         | Help with her postoperative care at home, particularly wound care including removal of sutures   |
| Occupational therapy                    | Assessed her for and provided her with an electric bath seat, level access shower (uses a shower wheelchair), kitchen trolley and ramps; also assessed and lowered cooking surfaces |
| Social services                         | Provided her with a wheelchair and assisted with mobility issues, including an adapted car        |

Pharmacotherapeutics

| Pharmacotherapeutics | Indications                                                                 |
|----------------------|-----------------------------------------------------------------------------|
| Iloprost infusions   | Vasodilating prostacyclin analogue                                           |
| Aspirin              | Antiplatelet, prior to warfarisation                                         |
| Clopidogrel          | Antiplatelet, prior to warfarisation                                         |
| Warfarin             | Anticoagulant for the prevention of thrombosis and embolization              |
| Cyanocobalamin       | Vitamin B12 vitamer used to decrease blood homocysteine levels, a risk factor for peripheral arterial disease |
| Simvastatin          | For the lowering of cholesterol and also for its pleotropic effects         |
Discussion

Buerger’s disease, also known as thromboangiitis obliterans, was first described over 130 years ago and later detailed by Leo Buerger through pathological examination of amputation specimens.\(^1\) It is a rare disease with differing worldwide prevalence; as low as 0.5% in Western Europe and as high as 80% in Ashkenazi Jews.\(^2\) Buerger’s disease, once considered a disease which almost exclusively affected men, is now increasing in prevalence among women owing to changing patterns in smoking.

It commonly affects small-to-medium-sized arteries and veins. The exact pathogenesis is unknown, however, it has been described to involve both inherited and prothrombotic factors leading to intravascular thrombosis.\(^3\) Combined with the effects of smoking, inflammatory infiltration of the vessel wall results.

Diagnosis is made using a number of diagnostic criteria, for example Olin’s criteria, which includes a patient less than 45 years of age with present or recent tobacco use.\(^4\) These patients usually have distal ischaemia, where other causes such as hypercoagulability have been excluded.

Long-term outcome with Buerger’s disease holds an amputation risk of 25% at five years, 38% at 10 years, and 46% at 20 years.\(^5\) However, amputation may be avoided, or deferred, by smoking cessation.\(^6\)

This case illustrates the difficulties faced in making certain diagnoses and conveys the vitality of engaging a multidisciplinary team to better the care we offer our patients.

The difficulty in initial diagnosis arose from the atypical patient demographic; a young female smoker, which highlights the changing presentation of Buerger’s disease. Our case seems to reflect current population trends with a higher proportion of smokers being women. It has been reported that despite the higher prevalence of smoking in men, the difference in proportions of smokers is less than previously recorded; 3% more men (22%) smoked than women (19%) in 2007 compared with 6% more in the 1980s.\(^7\)

The prevention of initiation and progression of Buerger’s disease is key. It is important to engage and educate patients about smoking cessation, including the conservative and pharmacological options. The risk of amputation in patients with Buerger’s can be reduced from 27% to 5% after stopping smoking.\(^1\)

Amputation is an emotive subject and our patients require both physical and psychological preparation and support. For a young woman facing amputation, the implications extend further and include aspects of body image. Several studies have looked at the effects of amputations and have demonstrated the need for early and long-term multidisciplinary support using a bio-psycho-social model.\(^8\)

The role of medical management in Buerger’s disease is to try to prevent tissue loss. However, these patients tend not to respond well to the typical treatments, due to the narrowing and thrombosis of their vessels. Endovascular repair is not commonly used, and bypass operations where diseased arteries are avoided as targets for reconstruction can be performed. The outcome of these have generally been reported as poor with 41% primary graft patency at 1 year, 32% at 5 years, and 30% at 10 years postoperatively.\(^9\)

The diagnosis of Buerger’s disease remains difficult due to the lack of specific clinical, radiological and pathological findings, as well as lack of internationally validated diagnostic criteria.

Recent advances in the treatment seem promising. The use of granulocyte colony-stimulating factor-mobilized autologous mononuclear cells resulted in a 79% one-year amputation-free rate, and may be considered in patients unsuitable for traditional revascularization.\(^10\)

This case has demonstrated the challenges in diagnosis and treatment of Buerger’s disease in a patient who has undergone a number of amputation operations. Through the application of a holistic and individualized multidisciplinary team approach, her independence has been maintained.

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