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Buerger's Disease Problems and Strategies Way Forward

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Abstract

Back Ground

Buerger's disease, the aetiology of which is still largely unknown, is a peripheral vascular disease, which occurs predominantly in young men who smoke. The severity of disease lies in loss of work days and the need for amputation in more than a quarter of patients.

Aim

The purpose of this study was to evaluate the progression of Buerger's disease and response to various modalities of treatment

Methods: A retrospective study was carried out for a period of five years (January 2005 to January 2010) on 143 patients at GMC Jammu to evaluate the progression of Buerger's disease. 89% (128) in our study were smokers. Buerger's disease was diagnosed according to Olin's diagnostic criteria.

Results

Total number of patients admitted during past 5 years was 143; 56% (79) patients who were on medical treatment were benefitted with improvement in claudication distance. 26% (37) patients had to be put on Prostacycline analogues infusion to manage critical limb ischemia out of which 21 (54%) responded, however 10 (31%) patients required lumbar sympathectomy for pain relief. 6 (15%) were subjected to amputation.

Conclusion

Buerger's disease management is challenging considering the age of the patient, poor socioeconomic status, type of pathology with unavailability of definite surgical procedure, except for discontinuation of tobacco use, there is no other definite therapy.

Keywords: Buerger's Disease, Thromboangitis Obliterans, Critical Limb Ischemia.

Introduction

Thromboangitis obliterans (Buerger's Disease) is a type of vasculitis in young mostly male subjects and is strangely linked to smoking. It affects the small and medium sized arteries and veins of the limbs.¹ The prevalence of the disease among all patients with peripheral arterial disease varies from as low as 0.5 to 5.6 percent in Western Europe to as high as 45 to 63 percent in India, 16 to 66 percent in Korea and Japan, and 80 percent in Israel among Jews of Ashkenazi ancestry.² The clinical criteria include: age under 45 years; current or recent history of tobacco use; presence of distal-extremity ischemia indicated by claudication, pain at rest, ischemic ulcers or gangrenes and documented by non-invasive vascular testing; exclusion of autoimmune diseases, hypercoagulable states and diabetes mellitus; exclusion of a proximal source of emboli by echocardiography or arteriography; consistent arteriographic findings in the clinically involved and non-involved limbs.³ The severity of disease lies in loss of work days and the need for amputation in more than a quarter of patients. Complete cessation of smoking remains the cornerstone of therapy.⁴ Rheology regulating drugs cilostazol and pentoxifylline are integral part of medical treatment. Prostacycline analogues can be used to help the patient through critical ischemia.⁵ Lumbar sympathectomy is very helpful in controlling rest pain, ulcerations and impending gangrene.⁶ Salvage procedures like omental flap transposition and Ilizarov's procedure are required to prevent amputation.^{7,8} The use of vascular endothelial growth factor gene or cell based therapy to induce therapeutic angiogenesis have opened up new possibilities for treatment.⁹

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Material and Methods

Of 143 patients diagnosed to have Buerger's disease at Govt Medical College Jammu from 2005 - 2010, 98% were males. Maximum number of patients 82% (118) presented in 3rd decade.

Table 1

Presenting symptom	%
Claudication	41%
Foot	(25%)
Calf	(16%)
Critical Limb Ischemia (Parasthesia, Coldness, Cyanosis)	31%
Rest Pain	10%
Gangrene or ulcer	15%
Thrombophlebitis	3%

Diagnosis was based upon Olin's criteria (2000)¹⁰. Ankle Brachial Index and duplex scans were done in all patients. Tests to rule out other causes of vasculitis which were done included a complete blood cell count; liver function tests; determination of serum creatinine concentrations, fasting blood sugar levels and sedimentation rate; tests for antinuclear antibody, rheumatoid factor, serologic markers for CREST (calcinosis cutis, Raynaud phenomenon, sclerodactyly and telangiectasia) syndrome and scleroderma, and screening for hypercoagulability. However, arteriography was done in 45% of patients, dorsalis pedis artery biopsy in 14 (10%) of patients for pathological confirmation of diagnosis.

Management protocol

Patients presenting with claudication were managed with medical treatment in the form of cilostazol/Pentoxifylline along with pain relief (tramadol) and antiplatelet drugs. Patients presenting with critical limb ischemia were managed with medical treatment along with prostacyclin analogue infusion. Patients with rest pain and impending gangrene not responding to medical management were managed with lumbar sympathectomy. Patients who could not be benefited from above mentioned management regimes were progressing to gangrene were subjected to salvage procedures in the form of omentopexy or Ilizarov's procedure. Our unit is in the process of starting vascular endothelial growth factor stem cell therapy; hence 3 patients were referred to other centres for stem cell therapy.

Results

On continuous follow-up 72% (102) patients developed ulcer/gangrene, 42% (60) developed phlebitis migrans and about 10% patients had upper extremity involvement and 36% (51) patients had bilateral lower limb disease. 56% (79) patients who were on medical treatment were benefited with improvement in claudication distance. 26% (37) patients had to be put on prostacyclin analogue infusion to manage critical limb ischemia, out of which 21 (54%) responded, however 10 (31%) patients required lumbar sympathectomy for pain relief. 6 (15%) were subjected to some sort of amputation. 25% (36) patients had to be subjected to lumbar sympathectomy either unilateral or bilateral to tackle rest pain, impending gangrene or to prevent

progression of symptoms. 10 (29%) patients still required either toe or forefoot amputations, 4 (11%) required salvage procedures. 3% (4) patients were subjected to salvage procedures in the form of omental flap transposition/Ilizarov procedure to save the limb and in 3 out of 4 amputations was prevented. 2% (3) patients referred for vascular endothelial growth factor stem cell therapy and results have been encouraging in the form of pain relief and ulcer healing. 20% (28) patients were subjected to some sort of amputations (toe n=14) (fore quarter amputation n=12) and (syme's amputation n=2). 9% (12) patients were lost in follow-up.

Discussion

Buerger's disease was first defined 130 years ago and the details were defined by Leo Buerger by means of histopathologically examining the amputation specimens¹¹. Amputation risk of the long-term results of Buerger's disease are 25% per 5 years, 38% per 10 years and 46% per 20 years.¹² Tissue loss is tried to be prevented by applying medical treatment. Buerger's disease is a type of vasculopathy, which involves peripheral vessels, mostly the lower limbs among young tobacco smokers. Due to multiple remissions and repeated hospitalization, this disease costs a huge economic burden. As the patients of Buerger's disease are young, the earning age is disturbed. This causes socioeconomic morbidity for the family and society too. So, early diagnosis, prevention and management of Buerger's disease has prime importance. Usually, males suffer Buerger's disease. In our study 2% were female. The Japanese study showed 4% female cases¹³. Different studies have suggested 8% to 20% female patients among Buerger's disease cases. Increasing incidence of Buerger's disease among females might be due to increasing smoking habits in females.

Mean age of Buerger's disease patients at the time of onset in the Japanese study¹³ was 36.8 years. Similarly, 82% of cases in our study belonged to the age group 31 to 45 years. Another study from Japan published in 1999 including 266 male and 21 female, currently suffering from clinical symptoms showed a mean age of 60 years¹⁴. 89% of Buerger's disease in this study was a tobacco smoker and most of the patients smoked 5 to 25 cigarettes daily. Similar study from Hong Kong reported 9 to 23 cigarettes per day smoked by their patients¹⁵. The same study on 89 patients in Hong Kong reported 62% lower limb involvement; Japanese reported 85% while our study has shown 90% lower limb involvement.

Upper limb involvement in our case was 10% in comparison to 15% in Japanese patients¹³. Turkish surgeons have studied the hospital records of 216 patients (214 men and 2 women) with Buerger's disease retrospectively. Of these patients 21 had had an arterial revascularization; 183 lumbar and 20 thoracic sympathectomies had been performed¹⁶. 25% of patients underwent lumbar sympathectomy resulting in a 29% later amputation rate in our patients was much more successful than lumbar sympathectomy leading to 70% later amputation is

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Japanese study¹³. German workers have shown 35.8% amputation rate mostly in old age group¹⁷. The higher incidence of amputation may be due to continuing cigarette smoking by our patients. Recent Japanese study has also shown the higher amputation rate among current smokers¹⁴

Conclusion

Buerger's disease management is challenging considering the age of the patient, poor socioeconomic status, type of pathology with unavailability of definite surgical procedure, except for discontinuation of tobacco use, there is no other definite therapy, prostacycline analogues may help patients with critical limb ischemia, lumbar sympathectomy provides short term pain relief, promotes ulcer healing and delays amputation. Recent innovative genetic and cell based therapeutic approaches have been proposed to induce angiogenesis but they require evaluation in randomized controlled trials to confirm their beneficial effects in patients with Buerger's disease

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Fig 1 Clinical Presentation of a patient with Buerger's Disease



Fig 2 Angiography picture of a patient of Buerger's disease

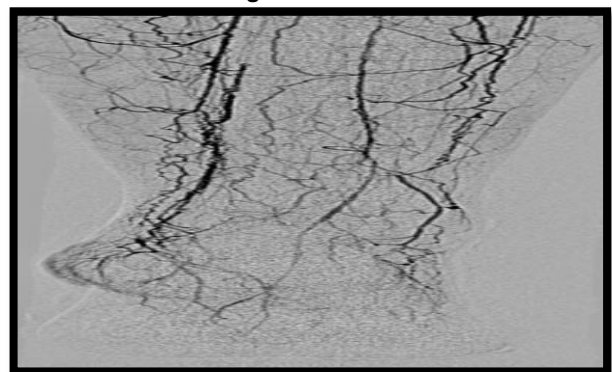


Fig3 Omental flap transposition in one of the patients

