The Clinical Course of Buerger’s Disease

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The clinical and social characteristics of 118 patients with Buerger’s disease (thromboangiitis obliterans: TAO) were surveyed. The prognosis for many patients is relatively favorable. Arterial reconstruction plays a role in shortening the healing times of ischemic ulcers despite its poor long-term results. Of 118 patients, 13 underwent major lower limb amputation (11%), 33 underwent foot or toe amputation (28%), one underwent hand amputation (1%), and 12 underwent finger amputations (10%) after the onset of the disease. The progression of symptoms was surely influenced by smoking, but this was not the only deleterious factor as there were patients with stable TAO which was unaffected by their continued smoking. In 66 patients, the new occurrence of ulceration and gangrene was not observed over the age of 60. Ten of 13 patients (77%) who underwent major lower limb amputation lost their jobs, but only 7 of 105 patients without major amputation lost their jobs (7%). In all patients, the progression of symptoms was self-limited and recurrent ulcers occurred less frequently with ageing. To avoid factors that markedly influence the quality of life, early treatment and strict instructions prohibiting smoking should be conducted.

Key words: Buerger’s disease (TAO), prognosis, job loss, smoking, quality of life

INTRODUCTION

TAO is characterized by occlusive segmental and often multiple lesions of medium-sized and small arteries and veins in young male smokers. The etiology of TAO remains obscure, and the prevalence of TAO in Japan appears to be decreasing. The number of new patients who visited our hospital has also been markedly decreasing: 65 patients between the years 1973 and 1979, 66 between 1980 and 1989, 23 between 1990 and 1999, and only 5 between 2000 and 2007. In this context, it is quite important now to reveal the clinical course of this vanishing disease in Japan. In this retrospective study, we surveyed several factors which all influence the quality of life of the patient.

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PATIENTS AND METHODS

Between 1973 and 2007, a diagnosis of TAO was made on 159 patients at the University Hospital, under the following five clinical criteria as described by Shionoya: 1) smoking history, 2) onset before the age of 50 years, 3) infrapopliteal arterial occlusive disease, 4) either upper limb involvement or phlebitis migrans, and 5) absence of atherosclerotic risk factors other than smoking. Infrapopliteal arterial involvement was diagnosed in all cases by means of arteriography. A diagnosis of upper limb involvement was usually made based on the patient’s history and on physical examination by Allen’s maneuver. Arteriography was performed only on patients with symptoms located in the upper extremities. Patients with collagen disease, popliteal entrapment syndrome, cystic adventitial disease, hypercoagulable state, thoracic outlet syndrome, and atrial fibrillation were excluded.

A total of 118 of 159 patients (74%) with TAO for whom complete follow-up data for the period following the onset of the disease were available were included. Follow-up data were derived from records and from
questionnaires. For those patients still alive, all treatments for TAO, the presence or absence of limb amputation, loss of job, divorce, and continued smoking were surveyed by direct interview, written reports, or telephone. For deceased patients, replies were obtained from the patient’s family by written reports or telephone. Graft patency was evaluated by physical examination assessed by ankle blood pressure measurement or arteriography. However, when direct examination was impossible, graft patency was marked as unclear. The graft patency rate (primary, secondary), limb salvage rate, and survival rate were calculated using the life-table method. Associations were tested using the chi square test.

Clinical Course

Before presentation of the results of the clinical course analysis, a description of the typical clinical course of a patient with TAO is helpful in order to summarize most of the characteristics of the disease.

[Case] The patient, a male clerk, began to have clearly visible ulceration on the big toe of his right foot at the age of 24. He smoked a pack of cigarettes a day at that time. TAO was diagnosed and thereafter he underwent several treatments including femoro-tibial bypass, lumbar sympathectomy, a Smithwick operation, intraarterial PGE1 infusion and spinal stimulation due to the frequent occurrence and recurrence of ischemic ulcerations on both his upper and lower extremities. Minor and major amputations of several parts of his upper and lower extremities were performed at the ages of 26, 28, 32, 34, 36, 38, 39, 41, 42, 43, and 44. He was admitted to several hospitals 20 times during a 25-year period, and in total spent a period of over 5 years hospitalized. He continued to smoke, and eventually lost his left hand, all his right fingers, and both lower extremities. He had to change his jobs several times, and he eventually lost all of them due to long hospital stays and limb loss (Fig. 1).

Results

Of 118 patients in our sample, 114 were men (97%) and 4 were women (3%). The age at the initial consultation at our hospital ranged from 20 to 65 years, with an average of 39 years. The age at the initial onset of disease ranged from 17 to 49 years, with an average of 34 years. About 90% of patients had initial ischemic symptoms between the age at 20 years and 44 years (Table 1). The follow-up period ranged from 0.3 to 35 years, with a mean of 19.5 years. All patients were smokers. One hundred and seventeen patients (99%) had arterial involvement of the lower extremities, and 80 (68%) had arterial involvement of the upper extremities. Forty-six (39%) had phlebitis migrans. No patients had atherosclerotic risk factors other than smoking.

Ischemic symptoms were observed in 142 lower extremities and 45 upper extremities. The most severe complaints experienced during follow-up are shown in Table 2.
Before the initial consultation at our hospital, 4 patients had undergone a bypass operation on a lower extremity, 57 extremities of 35 patients lumbar sympathectomy, and 9 extremities of 7 patients thoracic sympathectomy. All previous bypass grafts were occluded. In our hospital, 48 lower extremities of 34 patients underwent a bypass operation, 27 extremities of 27 patients lumbar sympathectomy, and 2 extremities of 2 patients thoracic sympathectomy. In all patients, vasoactive agents, anticoagulants, or antiplatelet agents were prescribed in accordance with the ischemic condition. The kind of agents prescribed and the compliance of patients after discharge were unclear in many cases.

Bypass surgery of the lower extremities was performed 48 times in 41 extremities in our hospital. Bypass was selected for cases of intermittent claudication in 11 extremities of 11 patients and for rest pain or necrotic lesion in 37 extremities of 30 patients. Seven of the 48 arterial reconstructions were redo operations. Proximal anastomotic sites consisted of the external iliac artery in 1 limb, the common femoral artery in 13 extremities, the superficial femoral artery in 30 extremities, and the popliteal artery in 3 extremities. Distal anastomotic sites consisted of the above-knee popliteal artery in 4 extremities, below-knee popliteal artery or the crural arteries in 44 extremities. In 41 extremities, autologous veins were used. In 7 extremities without an available autologous vein, composite grafts, prosthetic grafts, or bovine grafts were used.

Primary graft patency rates of 48 arterial reconstructions were 48%, 34%, and 24%, 1, 5, and 10 years after the operation, respectively. Secondary graft patency rates were 61%, 48%, and 42%, 1, 5, and 10 years after the operation, respectively (Fig. 2).

All bypasses using prosthetic grafts, bovine grafts or composite grafts were occluded within 10 months. The
graft patency rate was not satisfactory; however, only six of 23 extremities with failed grafts (26%) underwent limb amputation. Fifteen of 27 grafts in patients who continued smoking after operation and 4 of 13 grafts in patients who stopped smoking after operation were occluded. There was no significant difference between patients who continued smoking and those who stopped ($p = 0.2571$).

Of 142 lower extremities, 17 underwent major amputation (12%) and 48 underwent minor amputation (34%) after the onset of disease. Of the 17 extremities that underwent major amputation, nine were performed in our hospital. Of 45 upper extremities, one underwent a hand amputation (2%), and 18 underwent finger amputations (40%). Over two thirds of the extremities remained intact during the clinical course.

Of 118 patients, 13 underwent major amputation (11%) and 33 underwent minor amputations (28%) after the onset of the disease. Of the 118 patients, 1 patient underwent a hand amputation (1%), and 12 patients underwent finger amputations (10%).

Age at the onset of disease, age at each amputation, and the range of the follow-up period are shown in Fig. 3. The majority of patients underwent major or minor amputations from 10 to 15 years after the onset of disease, but some patients underwent amputations after a longer interval. No patient underwent amputation over the age of 60.

Of 79 patients who continued smoking, major limb amputation was performed on 13 (16%). Thirty-nine patients who stopped smoking did not undergo a major amputation. There was a correlation between continued smoking and limb amputation ($p = 0.0176$).

Employment conditions after the onset of disease were investigated. Seventy-four patients (63%) were blue-collar workers such as drivers, factory workers, carpenters, and plasterers. Seventeen patients lost their jobs due to frequent hospital admission, and due to limb amputation. All patients who lost their jobs were blue-collar workers. Thirteen patients underwent major amputation. Ten of them (77%) who underwent major lower limb amputation lost their jobs, but only 7 of 105 patients without major amputation lost their jobs (7%). There was a significant association between major amputation and job loss ($p = 0.0017$).

One hundred and three patients were married at onset of disease. Eight of them (8%) divorced, 5 due to frequent hospital admission, limb amputation, and losing their job. The divorce rate due to disease was 8%.

Three of 10 patients who underwent major lower limb amputation divorced, but only 7 of 93 patients without major amputation divorced. There was no association between limb amputation and divorce ($p = 0.0857$).

Twelve patients (10%) died during follow-up. The
causes of death were malignant disease in 4 patients, cerebrovascular accident in 2 patients, heart failure, suicide, and pneumonia in 1 patient, respectively. The cause of death was unclear in three patients. Cumulative survival rates were 98%, 93%, and 79%, 10, 20, and 30 years after the initial onset of disease, respectively.

**Discussion**

In this study, complete follow-up data after onset of disease were available in only 74% of patients. The low follow-up rate is due to a majority of patients being young and of low socio-economic status, thus many of them live in rental-houses and change their addresses frequently. Patients with TAO receive financial assistance for their medical expenses, and every year they must send a medical certificate to the health center of the prefecture where they live. If they do not return to our institution due to a change of address or due to remission of the disease, it is difficult to follow them up.

Although success of the bypass surgery markedly improves ischemic symptoms, it is not often applicable for patients with TAO. Among our sample, it was appropriate for only one quarter of patients, specifically those exhibiting because of widespread arterial occlusions at multiple levels with prevalence in the distal part of the extremities. The results of arterial reconstruction in our hospital were no better than those reported by several other investigators.3, 4)

Problems with grafts occurred frequently within 2 years of surgery. Early graft failures within 30 days were extremely common because of poor technique, low quality of grafts, or poor run-off. Intermediate graft failures within 2 years were also high. They were caused by proliferative changes in the graft itself or in revascularized segments. The progression of the underlying disease occurs at a relatively high incidence despite the administration of antiplatelet agents or anticoagulants.5)

Although the graft patency rate was not satisfactory, major limb amputation was not necessary in only 6 of 23 extremities with failed grafts (26%). Even when arterial reconstruction for Buerger’s disease induces graft occlusion shortly thereafter, limb amputation can be avoided in many patients in whom the healing of ulcers has been achieved during graft patency. Arterial reconstruction plays a role in shortening the interval between surgery and the healing of ulcers.

Although determining how best to treat ischemic symptoms is a real problem, the aim of treatment for Buerger’s disease is to prevent the occurrence and recurrence of ischemic ulcer. Among our sample, it is quite interesting that there was neither occurrence nor recurrence of ischemic ulcer in those over the age of 60. In contrast arteriosclerosis obliterance, it is thought that there is an active phase of disease in the clinical course of TAO. The onset or duration of the active phase may be different in each individual, but the progression of TAO may subside over the age of 60. One of the possible reasons is that developed collateral arteries, which are sufficient to avoid major amputation, are less influenced by atherosclerotic arterial changes with aging.

While the cause of TAO remains unknown, smoking is very closely related to the exacerbation and remission of the disease.2, 6, 7) Thirty-nine patients who quit smoking did not undergo a major amputation. Of 79 patients who continued smoking, 13 underwent major amputation. In other words, all who lost their extremities did not abstain from smoking. There was a correlation between continued smoking and limb amputation (p = 0.0176). It is true that smoking is surely one of the important detrimental factors, however, it is also true that not all patients who use tobacco subsequently go on to require limb amputation. In Japan, an analysis of 850 patients identified in a national study showed that the risk of amputation was 2.73 times higher (95% CI: 1.86−4.01) in patients who continued to smoke.8)

Few studies have reported rehabilitation of patients with TAO. Shionoya et al.9) reported that 17 of 165 patients (10%) lost their jobs, and that 30 patients (18.2%) changed their occupations. In our study, 17 patients lost their jobs due to frequent hospital admission, and due to limb amputation. All of them were blue-collar workers and 13 of 17 underwent major amputation. Physical handicaps especially in blue-collar workers with TAO, was shown to be a major factor causing job loss.

Eight (8%) of 103 married patients divorced. The main reasons were economical failure due to frequent hospital admission over a long period, and job loss in adolescence or in late middle age. The divorce rate for patients with TAO, however, was lower than that for the married Japanese female population as a whole, which ranged from 10% to 20% during the last two decades of the 20th century.10)

The prognosis for most patients with TAO is favorable.2, 7, 11) Among our sample, only 12 patients (10%) died during follow-up. The age at death ranged from 37 to 77 years, with an average of 64 years. The survival rate 30 years after the initial onset of disease was 79%.
REFERENCES


