Buerger's Disease in Tehran University of Medical Sciences Hospitals: A Fifteen Years Study

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Abstract- Buerger's disease is an occlusive inflammatory disease of the small and medium-sized arteries and accounts for a variable proportion of patients with peripheral vascular disease throughout the world. The aim of this study was to review the records of Buerger's disease patients admitted to surgery wards of our university hospitals. 277 patients with Buerger's disease were surgically treated between 1987 and 2002, in affiliated hospitals of Tehran University of Medical Sciences, in Iran. Two hundred and seventy three (98.6%) of the patients were male, aged 41.5 ± 11 years (mean ± SD); 99.6% of which were smokers with an average of 22.9 pack/years tobacco use. The major complaints included: ischemic ulcers in 203 (73.3%, CI 95%: 0.68-0.77) patients, rest pain in 201 (72.6%, CI 95%: 0.64-0.73), paresthesia in 143 (51.3%, CI 95%: 0.48- 0.58). Vascular bypass, sympathectomy and amputation were performed in 9.7% (CI 95%: 0.08-0.14) and 69.3% (CI 95%: 0.51-0.60) and 59.6% (CI 95%: 0.65-0.73) of the patients, respectively. Lumbar sympathectomy was carried out in 177 (63.9%) patients, while 15 (5.4%) patients underwent thoracic sympathectomy. In our study, afflicted patients were mostly young males, inveterate tobacco smokers. Patients presented frequently with ischemic ulcers or severe rest pain; thrombophlebitis and Raynaud's phenomenon were infrequent. Vascular reconstruction was rarely possible due to distal and segmental involvement; therefore sympathectomy and amputation were inevitable in a large group of patients in this study.

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Key words: Buerger's disease, thromboangiitis obliterans, smoking, vasculitis

Introduction

Buerger's disease is a non atherosclerotic, segmental, inflammatory, non supurative, progressive, obstructive panarteritis in 25-35 year old aged male smokers. It commonly affects the small and medium sized arteries, veins and nerves of arm and legs (1). Buerger's disease accounts for a variable proportion of patients throughout the world: 0.75% in North America, 0.5% to 5.6% in Western Europe, 16% to 66% in Korea and Japan, 45% to 63% in India and 80% in Israel among Jews of Ashkenazi descent (1). The reported incidence of this disease is 8 to 12.6/100,000 populations in North America, where the prevalence had declined substantially in the last 25 to 30 years (1, 2). Despite its relatively high prevalence in Middle East, Buerger's disease has been seldom reported from the countries in the region (3-6). However, Iran is accepted as one of the countries in which Buerger's disease frequently occurs, the incidence of this disease in Iran cannot be exactly determined as a nationwide epidemiological study has not been performed in this country. In an effort to determine the clinical course and surgical treatment methods of the disease in our university hospitals, we reviewed the records of the Buerger's disease patients during a fifteen years period.

Patients and Methods

We retrospectively reviewed hospital records and operative reports of all the patients diagnosed as Buerger's disease in a 15 years period (1987-2002) in Sina and Imam Khomeini Hospital. Vascular surgery wards of these hospitals are referral centers in Iran, with wide experiences in surgically treating patients with Buerger's disease. The definitive diagnosis of Buerger's disease was based on Shionoya's clinical criteria: (1) smoking history; (2) onset before the age of 50 years; (3) infrapopliteal arterial occlusive lesions; (4) either upper limb involvement or phlebitis migrans; and (5)

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absence of atherosclerotic risk factors other than smoking (7).

The patients with critical ischemic complaints such as rest pain, non-healing ulcer and gangrene without popliteal artery pulse underwent arteriography to find a suitable segment for arterial revascularization. Vascular reconstruction was done if there were suitable inflow and outflow artery. We performed sympathectomy to the patients who non-suitable for revascularization when there were limited ischemia complains such as rest pain, non-healing ulcer without infection.

Data were collected using a specified form and included age, sex, cigarette smoking status, clinical presentation, the affected limb (right or left, upper or lower extremities), and the performed therapeutic procedures such as angiography of limb arteries, amputation, sympathectomy and vascular bypass surgery. All analyses were performed using SPSS 13.0 software package (SPSS Inc., Chicago, IL). Data are presented as mean ± standard deviation (SD).

Results

During a 15-years period (1987 -2002), 277 patients with the diagnosis of Buerger's disease were treated surgically in vascular surgery wards of Sina and Imam Khomeini Hospitals, Tehran-Iran; two hundred and seventy three (98.6%) of which were male, with a mean age of 41.5 ± 11 years. Their age ranged from 16 to 75 years; most of the patients were in their 30s (n=90, 32.5%) and 40s (n = 88, 31.8%), respectively. Elderly patients aged 60 or older comprised 7.9% of the patients. Figure 1 shows the distribution of the patients by age.

The majority of the patients (99.6%) were heavy smokers. The average cumulative tobacco use was 22.9 pack-years. Figure 2 summarizes the clinical manifestations of the patients enrolled in this study; sensory complaints included rest pain (72.6%, CI 95%: 0.64-0.73) and paresthesia (51.3%, CI 95%: 0.48-0.58). The positive past medical history of ischemic ulcer was present in 203 patients (73.3%, CI 95%: 0.68-0.77) frequently reported in toes. The migratory thrombophlebitis was present in seven patients (2.5%, CI 95%: 0.02-0.05).

Lower and upper extremity involvement was observed in 295, 68 cases respectively. There were 16 cases (9.5%) with vascular disease in both upper and lower extremities. Angiography was performed in 29.2% of the patients.

Table 1. Therapeutic procedures performed in the Buerger's disease patients

Procedure	Frequency
Vascular bypass surgery	**27 (9.7%)**
Femoro-Popliteal	11 (4%)
Femoro-Peroneal	6 (2.1%)
Femoro-Tibialis Posterior	5 (1.8%)
Femoro-Tibialis Anterior	5 (1.8%)
Amputation	**165 (59.6%)**
Toe(s)	138 (49.8%)
Transmetatarsal	5 (1.8%)
Below knee	21 (7.6%)
Above knee	1 (0.4%)
Sympathectomy	**192 (69.3%)**
Lumbar	177 (63.9%)
Thoracic	15 (5.4%)

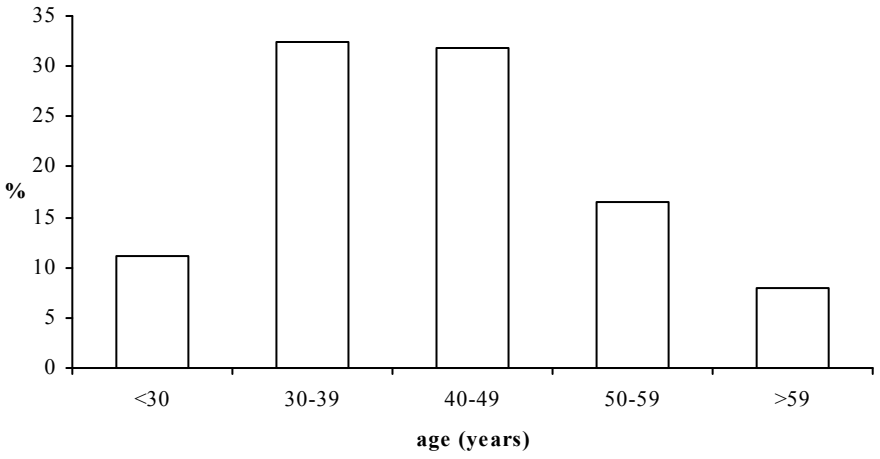

Figure 1. Distribution of the patients according to age

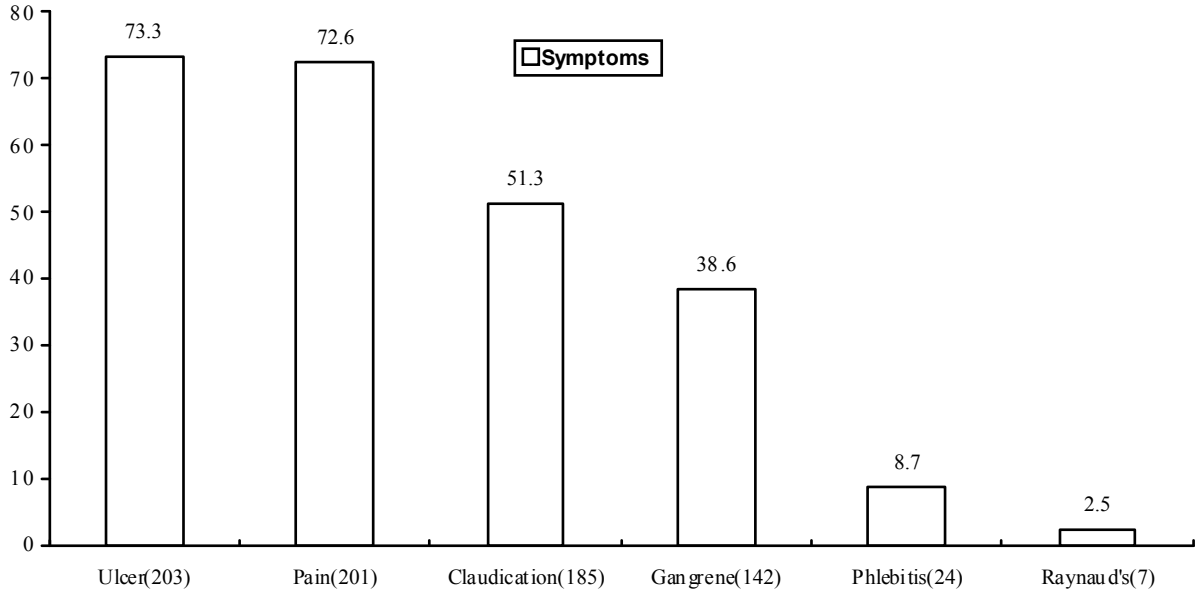

Figure 2. Clinical presentation of the patients with Buerger's disease

The majority of the patients underwent an operation so that an amputation, sympathectomy, or reconstructive arterial surgery would be performed. Sympathectomy was carried out in 192 patients (69.3%, CI 95%: 0.51-0.60); one hundred and ninety- two patients (64%) of which had lumbar sympathectomy and the other 5.4% had thoracic sympathectomy. Bypass grafts construction via an autologous vein was performed in 27 patients (9.7%, CI95%: 0.08-0.14). Table 1 outlines the different therapeutic procedures done during hospitalization period.

Discussion

Thromboangiitis obliterans or Buerger's disease is a relentless and devastating vasculitis causing significant loss of digits and limbs. It is a disease more seen in males of age 35 to 50 years (1). In a recent report from Japan and Bangladesh, mean age at admission of these patients was 42.5 and 38.6 years, respectively (4, 8). In our study approximately, all patients were young smoker males, which is in agreement with previous studies. It should be noted that patients aged 60 or older, at the time of survey, comprised 7.9% of our patients.

Until the middle of 1980s, female or elderly patients were thought to comprise less than 2% of the patients with Buerger's disease. However, the prevalence of Buerger's disease in women is increasing, nowadays; which is attributable to the increased number of female smokers in the past few decades. In the studies carried out by Lie (1987), Olin *et al*. (1990), Mills *et al.* (1987), females consisted 11, 19 and 23 percent of Buerger's disease patients, respectively (1, 9, 10). In our study, the prevalence of Buerger's disease among females was found to be less than the reported prevalence. It may be due to cultural beliefs and as a result low incidence of smoking among Iranian women. Furthermore, our sample was not large enough to cover all the rarely encountered women with Buerger's disease. Although the cause of this disease is unknown, but tobacco use seems to be a trigger for what appears to be an autoimmune mechanism in a group of patients and there was a strong seasonal variation for admission, with winter admissions being significantly the most common (11, 12). In addition to abnormal peripheral microcirculation, some systemic factors (immunological or hormonal) and an interaction between the coagulation system, its control proteins, and the vascular endothelium may be involved in the development of Buerger's disease (13, 14). Therefore TAO development in the absence of a past history of smoking, observed in one (0.4%) of our patients, would not be an extraordinary finding. Same cases have also been reported in other publications: 6.8% of the Buerger's disease patients studied by Sasaki and colleagues were non-smokers (11). The common complaints in our patients included ulcer of the foot (73.3%), Raynaud's phenomenon (8.7%) and thrombophlebitis (2.5%), (Figure 2) which differs from the results achieved in Cleveland

Clinic Foundation. In the latter study, 76% suffered from ischemic ulcers, 44% revealed to have Raynaud's phenomenon and 38 % had thrombophlebitis (9). In fact, Raynaud's phenomenon and thrombophlebitis had a lower prevalence in the present survey compared with the findings of Olin *et al.* (1990) at Cleveland Clinic Foundation (9). However, thrombophlebitis was reported in 6% of cases in another study (15). Intermittent claudication and gangrene of the extremities were present in 66.8 and 38.6% of our patients whilst a Colombian study reported these complaints in 62 and 70% of their cases, respectively (16).

The upper extremity involvement was seen in 24.5% of our patients in opposition to 28-50% in other studies (1, 9).

Although the exact reason is unknown, the fact that Allen test and upper extremity angiogram are not routinely performed in our centers may account for the resulted lower incidence. In other words, more patients with upper extremity affection may be identified if these tests would be performed routinely in those with ischemic symptoms in lower extremity (9).

Complete discontinuation of cigarette smoking or other use of tobacco is the only proven strategy in order to prevent the progression of the disease and further amputation (13, 17). As a rule, ischemic lesions are healed with complete tobacco asceticism and new ischemic lesions reoccur rarely, without tobacco re-exposure. Smoking one or two cigarettes a day, using smokeless tobacco (chewing tobacco or snuff), or using nicotine replacement may keep the disease active. In other words, medical and surgical therapy can only palliate accrued damage; complete abstinence from tobacco use is the sole which allows stabilization of the process (7).

The majority of patients in the present survey were surgically treated because our hospitals are referral centers admitting patients with complaints intractable to routine medical therapy, requiring operations.

Vascular bypass and sympathectomy are the most commonly performed surgical treatments now a days, furthermore any effort should be taken to avoid a major amputation (18).Vascular reconstruction is usually not possible in patients with Buerger's disease because of the diffuse segmental involvement and distal nature of the disease (1). However, bypass surgery via an autologous vein should be considered if the patient has severe ischemia in a distal target vessel. An arterial revascularization was performed in 21 out of 216 patients (9.7%) in Sayin et al. study (19). Bypass grafts were performed in 27 patients (9.7%) of our study.

Sympathectomy seems to be helpful in the healing of superficial ischemic ulcerations, yet its role in preventing amputation or treating pain remains unclear (9). Present study revealed a higher rate of sympathectomy compared with other sources (69% versus 51%). It should also be noted that sympathectomy has also an important role in treating smoking abuse.

Amputation was performed in 27 and 25.2% of the patients studied by Olin and Sasaki, respectively (9, 11). In comparison to our study (table 1), more patients lost their affected limbs in the present survey. This is perhaps a result of delay in referring patients to our centers, which leads to severe symptoms (almost all patients had ischemic ulcers) at admission necessitating more aggressive treatments. Toes compromised half of the limb loss cases in our series, consisted one-third of all of amputations in Cleveland Clinic (9).

In conclusion, the present study allows an understanding of the clinical characteristics of Buerger's disease in two referral centers in Iran. Iran is a country in the Middle East region, where the Buerger's disease is relatively common. The majority of our cases were young smoker males and thrombophlebitis and Raynaud's phenomenon was rather infrequent as well as the upper-extremity involvement. It could be concluded that the majority of our cases had severe symptoms of limb ischemia and as a result amputation was inevitable in a large group. According to the fact that Buerger's disease has a close relationship to smoking habits; and remission is usually achieved merely if the patient stops smoking, patient education may be the most important means in treating this disease.

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