

Clinical Study: Diagnosis By Shionoya Criteria and Case Specific Management Of thromboangiitis Obliterans

Sharath Kumar K.L.*, Shashidhar Naik C.*

Registrar, Sri Shankara Cancer Institute and Research Centre, Bangalore.

Abstract

Introduction: Buerger's disease occurs almost exclusively in smokers who are males usually between 20 and 50 years of age. The typical patient is one who is a heavy smoker and had started smoking at an early age. Following is the spectrum of presentation of disease. *Methodology:* The treatment of each patient was individualized with the aim to achieve foot salvage wherever feasible. A record of patient's progress and response to various modalities of treatment was made to evaluate severity of disease, in long term prognosis and also evaluate the efficacy of treatment. *Results:* Ulceration was seen in 9 patients (18%). 11 patients (45.83%) out of total 24 patients with gangrene had a definite line of demarcation and 13 patients (54.17%) did not have definite line of demarcation. *Conclusion:* All the cases were diagnosed with Shionoya criteria, Lower limb medium to small caliber vessels were commonly involved in Buerger's disease

Keywords: Shionoya Criteria; Buerger's Disease; TAO.

Introduction

Buerger's disease or TAO is a distinct clinical and pathological entity. It is defined as a non-atherosclerotic, segmental, inflammatory, occlusive vascular disease of unknown aetiology occurring predominantly in young male smokers and affecting small and medium-sized vessels predominantly Buerger's disease seems to commence peripherally and extend proximally. The disease may pass unnoticed until the occlusive

disease involves the forearm or crural arteries.

Buerger's disease occurs almost exclusively in smokers who are males usually between 20 and 50 years of age. The typical patient is one who is a heavy smoker and had started smoking at an early age. Following is the spectrum of presentation of disease.

Claudication is single most important symptom of arterial occlusive disease in extremity resulting from obstacle to the flow of blood to the muscles. Walking being the exercise invariably the patient will be found to take rest or giving up walking.

The causation of this claudication is due to the accumulation of acid metabolites in the face of ischemia, the chemical nature of metabolite is not known and hence is called as 'substance P'. This stimulates the somatic nerves and produces cramp. Anaemia and arterial spasm exaggerate it.

History taking and physical examination are usually sufficient to make the diagnosis of Buerger's disease, but additional diagnostic procedures are required to evaluate the degree of ischemia objectively and to select the most appropriate therapy. The traditional diagnosis of TAO is based on clinical criteria of Shionoya with all 5 elements required (26).

The only proven strategy to prevent progression of the disease and avoid amputation is the complete discontinuation of cigarette smoking or abuse of tobacco in any form.

The therapeutic goals for Buerger's disease involve

- Improving arterial flow to the limb
- Alleviating ischemic pain
- Treating concomitant infection
- Improving wound healing by treating locally
- Aiming for 'Foot Salvage' which means retention of a functional foot that allows standing and walking without prosthesis

Corresponding Author: Sharath Kumar K.L., Registrar, Sri Shankara cancer Institute and Research centre, Bengaluru, Karnataka 560004.

E-mail: drsharathkumarkl@gmail.com

Methodology

Fifty patients in the age group of 20-50 years who were smokers, presenting with symptoms of ischemia in limbs and were admitted in Medical College Hospital, were taken up for study.

The method of the study consisted of taking a good clinical history in a chronological order as soon as the patient was admitted. A thorough clinical examination was carried out personally to find out and establish clinically first, the presence of vascular obstruction. Detailed vascular system examination was done as per the proforma provided and blood pressure measured to rule out hypertension.

The degree of vascular inadequacy and extent of the spread of the disease was assessed clinically by noting the colour change, extent and spread of gangrene and absence of peripheral pulses in the affected limbs. This together with history of the patient regarding the distribution and type of pain gave in a fairly good number of cases studied, an idea of the state of patient's vascular condition.

Later after clinical scrutiny essential laboratory investigations were done as per the proforma provided to rule out presence of atherosclerotic risk factors, the presence of which formed basis for exclusion of the patient from study. Patients were further evaluated objectively by Doppler scanning whenever feasible to assess the level and degree of obstruction objectively and for confirmation of infra-popliteal segment involvement.

The treatment of each patient was individualized with the aim to achieve foot salvage wherever feasible. A record of patient's progress and response to various modalities of treatment was made to evaluate severity of disease, in long term prognosis and also evaluate the efficacy of treatment.

Patients who returned for follow up were followed up for minimum of six months during each follow up detailed history was taken and progress of the disease was assessed.

Based on history, physical examination and investigation patients were either included or excluded from study.

Results

Blackish discoloration due to hyperpigmentation was seen in only 9 patients (18%).

Gangrene of the limb was seen in 24 patients (48%).

Signs of ischemia were present in 50 patients either in form of thinning of skin, diminished growth of hair, loss of subcutaneous fat, shininess of skin, trophic changes in nail, minor ulceration.

Ulceration was seen in 9 patients (18%).

11 patients (45.83%) out of total 24 patients with gangrene had a definite line of demarcation and 13 patients (54.17%) did not have definite line of demarcation.

Table 1: Cardio vascular system involvement

CVS	Frequency	Percentage
NAD	49	98.0%
Abnormality	01	02.0%
Total	50	100%

Cardiovascular system involvement in form of myocardial ischemia and angina was seen in 1 patient

(2%). No abdominal or skeletal system abnormality was noticed in any of the patients.

Table 2: Investigations profile

Parameters	Investigations		Urea	Creatinine	Cholesterol
	Hb	RBS			
Mean	11.5	96.3	26.6	1.2	172.2
Standard deviation	1.7	1.7	1.4	0.3	2.1
Minimum	8.0	55	10	0.5	146
Maximum	14.5	130	67	12	195
Range	6.5	75	57	11.5	149

Table 3: ECG findings

ECG	Frequency	Percentage
Normal	49	98.0%
Abnormal	01	02.0%
Total	50	100%

Mild to moderate anemia was seen 31 patients(62%). Other investigations were within normal limits.

ECG changes were seen in 1 patient(2%). In other cases ECG was normal. All the patients had features

suggestive of arterial insufficiency of medium and small sized arteries of both lower limbs and all the patients had ABI >0.3

Table 4: Doppler scanning

Doppler scanning	Frequency	Percentage
Abnormal	48	96.0%
Not done	02	04.0%
Total	50	100%

Doppler USG was performed in 48 patients (96%)

Table 5: Modalities of treatment

Treatment	Frequency	Percentage
Conservative management	20	40%
Lumbar sympathectomy	09	18%
Amputation	13	26%
Dis articulation and LS	05	10%
Dis articulation	08	16%

In our study, out of 50 cases 20 patients (40%) were managed conservatively.

09 patients(18%) underwent lumbar sympathectomy. Amputation was done in 13 patients (26%). Disarticulation alone was done in 08 patients(16%).Disarticulation and Lumbar sympathectomy was done in 05 patients (10%).

Discussion

In the majority of my cases the onset and progress of the disease was insidious. My results can be compared with that of Shigehiko Shionoya, MD., who studied 255 patients with Buerger’s disease from 1977 to 1988.

Clinical presentation and progression of Buerger’s disease in 255 patients, comparing it with my results, it would become obvious that paraesthesia, coldness and intermittent claudication were commonest presenting symptoms in Shionoya studies of 255 patients, whereas in my study claudication was the commonest presentation seen in 32 patients(64%). Among them calf claudication is seen in 20 patients(40%) and foot claudication seen in 12 patients(24%).

Findings in contrast to Shionoya study, is rarity of thrombophlebitis migrans in our patients only one patient (2%) gave history of it. Its association with TAO is so much stressed by western authors as to consider it as a specific feature of TAO. But many authors have denied its significance.

Barker (1936), Decamp et al (1952), Ackerman had followed up over years, ranging from 5-10 years the patients who had recurring attacks of thrombophlebitis but they failed to observe or see

their patients develop TAO.

Hence in summary it can be started that it forms a good association with TAO but cannot be recognized as a part of disease process itself. Other features described by Buerger and Shionoya etc., all were not seen in my series

It was hard to find or make out the colour changes of the skin. But in advanced cases some hyperpigmentation was always noticed on the toes or dorsum of the foot and would be of less importance at that stage. Only one patient (2%) showed rubrocy anotic discoloration of dermis over the leg below the knee.

It is well known in TAO that development of pallor on elevation of the leg above horizontal level to 45o or more and delay in return of colour to the leg over 30seconds indicates gross circulatory insufficiency. There will also be corresponding delay in filling of the veins with elevation and depression of the legs. In my study even the postural changes in colour was difficult to make out

Investigations were performed in my 50 cases to rule out of atherosclerotic risk factors and / or diagnosis and quantification of severity of level of block. Doppler USG was performed in 48 patients (96%). Other routine investigations were performed in majority of the cases as per the proforma given and none had atherosclerotic risk factors.

The following investigations were performed.

Non Specific

All cases were subjected to series of screening tests to determine the cause if any, of obliterative vascular

disease and such of those who showed atherosclerotic risk factors were discarded from study.

Hemogram

Hemoglobin estimation was done in all the patients. 31 patients (62%) were found to have mild to moderate anaemia. The lowest Hb% was 8gm% and ranged between 8-14.5gm%. Peripheral smear examination revealed microcytic hypochromic anaemic blood picture. This anemia might exaggerate or even hasten the gangrene formation by aggravating the tissue anoxia present already. Hence the need of iron therapy is stressed in all the cases.

Urine Examination

Urine examination was routinely performed to assess whether patient was diabetic or not and in all cases it was supplemented by RBS or FBS. None of the patients in my study were diabetic.

FBS/PPBS: It was routinely done in all patients to rule out diabetes.

Serum Lipid Profile

This was also routinely carried out in all the patients to rule out atherosclerotic risk factors. The minimum was 146 mg% and maximum 195 mg%.

None of the patients in my series had hyperlipidemia.

Blood Urea and Serum Creatinine

It was routinely done to rule out renal involvement. All the patients in my series had the values within normal limits.

Culture and Sensitivity

It was done in presence of ulceration.

Others

The blood pressure in all my 50 cases was well within normal limits. No evidence of hypertension was present. Other investigations of help were ECG and Chest x-ray to rule out significant cardiac lesion.

Patient No.24 on investigation was found to have following abnormality

- Chest x-ray showed left ventricular type of cardiomegaly
- ECG showed left axis deviation, T wave inversion

in leads V3-V6 and Q wave in leads V1-V4

- Echocardiography showed left ventricular hypokinesia with 50% ejection fraction.

Specific

Doppler Scanning

Doppler scanning was feasible in only 48 patients (96%). All the patients who underwent Doppler scanning had involvement of infrapopliteal arteries and the level of block in most cases was in distal tibial vessels.

Treatment

- *Conservative* In the present series of 50 cases studied by me, majority of the patients were managed conservatively constituting 20 patients (40%). They were treated by vasodilators, exercise therapy and local care of ulcer.
- *Sympathectomy:* In the present series of 50 cases, 4 patients (8%) were managed by LS alone.
- *Amputation:* In my 50 cases, 13 patients (26%) had undergone major amputation during their attendance at the hospital at different times. 10 patients (20%) underwent above knee amputation, 2 patients (4%) underwent below knee amputation and 1 patient (2%) underwent transmetatarsal amputation
- *LS and disarticulation / amputation:* In my 50 cases, 5 patients (10%) were also subjected to disarticulation in addition to sympathectomy. The toe was disarticulated due to previous gangrene.
- *Disarticulation:* Total of 13 patients (26%) were subjected to disarticulation of toe/toes.

Conclusion

Majority of the patients who underwent surgical management were treated with disarticulation alone, or disarticulation with lumbar sympathectomy or above knee amputation.

References

1. Frederick J Shoen. Blood vessels. Robbins and Cotran. Pathologic Basis of disease, 7th edition, Chapter 11, 542.
2. Lie JT. Thromboangiitis obliterans (Buerger's

- disease) revisited. *Pathol Ann.* 1988; 23: 257.
3. Mills JL, Porter JM. Buerger's disease : A review of update. *Semin Vasc Surg.* 1993; 6: 14.
 4. Nishikimi N, Shionoya T, Shionoya S, et al. Microcirculatory characteristics in patients with Buerger's disease. *Angiology.* 1992; 43: 312.
 5. Mills JL, Friedman EI, Taylor LM, et al. Upper extremity ischemia caused by small artery disease. *Ann Surgery.* 1987; 206: 521.
 6. Mills JL, Taylor LM Jr., Porer JM. Buerger's disease in the modern era. *Am J Surg.* 1987; 123: 154.
 7. Dormandy JA, Murray GD. The fate of claudicant - a prospective study of 1969 claudicants. *Eur J Vasc Surg.* 1991; 5: 131-133.
 8. McPherson JR, Jergens JL, Gifford RW Jr. Thromboangiitis obliterans and arteriosclerosis obliterans. Clinical and Prognostic differences. *Ann Intern Med.* 1963; 59: 288.
 9. Shigehiko Shionoya. Buerger's Disease (Thromboangiitis obliterans) : Robert B Rutherford. *Vascular Surgery*, 4th ed, Vol. 1, Chapter 11, W.B. Saunders Company, 235-245.
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