

Long-term clinical outcomes in patients diagnosed with severe digital ischemia

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Summary

QUESTION UNDER STUDY: To investigate the aetiology and long-term clinical outcomes of patients diagnosed with digital ischemia.

METHOD: Data of 36 consecutive patients presenting with digital ischemia were collected in July 2000 to June 2001 from a vascular referral centre. Demographic data, aetiology, medication and treatment were abstracted from the medical records. Clinical outcomes were assessed at 5 year follow-up including ulcer healing, digital amputation and mortality.

RESULTS: Of the 36 patients, 69.4% were male and the mean age was 55 ± 14 years. In 15 patients (41.7%) a systemic disease was present and of those 53.3% was due to connective tissue disease. Twelve patients (33.3%) had hypothenar hammer syndrome and in 8 patients (22.2%) no apparent cause was found. Whereas 13 patients (36.1%) presented with rest pain or trophic lesions at baseline, no patients presented with these symptoms at follow-up. At follow-up, 18 (62.1%) patients had symptoms on provocation and 5 patients (4 patients with systemic disease and 1 with no apparent cause) had died. Digital amputation was performed in one patient at initial presentation and no digital amputation was performed at follow-up. No ulcer re-occurred and no workers' insurance compensation was applied. Of those with hypothenar hammer syndrome, 80.0% had symptoms on provocation at follow-up.

CONCLUSIONS: Among patients with digital ischemia, systemic disease and hypothenar hammer syndrome were the most frequent aetiologies. In patients with hypothenar hammer syndrome the clinical outcome was remarkably benign, although symptoms may persist with provocation, whereas patients with systemic disease have a high mortality rate.

Key words: epidemiology; upper extremity; ischemia; digital artery occlusion; occupational exposure.

Introduction

Symptomatic ischemia of the upper extremity is less frequently encountered than lower extremity vascular insufficiency and thus, poses challenging problems for diagnosis and management [1, 2]. While 12% of all peripheral arterial obstructions are located in the upper extremity, 70% of those affect the small arteries distal to the wrist [3]. The clinical manifestations of digital ischemia are various and range from sensitivity to cold and Raynaud's attack to rest pain, digital ulceration or gangrene [4]. Despite many publications concerning digital ischemia, it remains a real challenge to diagnose, to evaluate the aetiology and to determine therapeutic strategies. Secondly, most studies have focused on the prevalence and incidence of connective tissue disease in patients initially diagnosed with digital ischemia and many other aetiologies and long-term clinical outcomes are thus rarely discussed [5–7]. The differential diagnosis of digital ischemia is broad and includes cardiac or arterio-arterial embolism, local thrombosis, systemic disease, especially scleroderma, diabetes, chronic renal failure and coagulation disorders, and traumatic injury of the wrist arteries. Injuries to the wrist arteries may be caused by repetitive use of the palm in manual activities such as pushing, pounding or twisting, leading to hypothenar hammer syndrome [8]. The discovery of occupational hazards associated with hypothenar hammer syndrome may have implications in obtaining workers' insurance compensation. Thus, a thorough history and diagnostic work-up is essential.

From the last 20 years, we are not aware of any contemporary large series on the natural history and long-term clinical outcomes in patient diagnosed with digital ischemia, the development of ischemic ulcers, and the need of amputation. Thus, the purpose of this study was to evaluate the aetiology, to describe the therapeutic strategy, and to assess long-term clinical outcomes of patients diagnosed with digital ischemia.

Methods

Study population

From July 2000 to June 2001, 36 consecutive patients with digital ischemia were seen in a tertiary vascular referral centre and data on demographic and baseline clinical characteristics were collected and entered into a database. To ensure representative inclusion, patients were recruited consecutively and the weekly consultation lists were checked for missing cases. Patients with hand ischemia due to thrombo-embolic brachial artery occlusions or catheter-related ischemia were not included in the study. In all patients, the diagnosis of digital ischemia due to palmar and digital artery occlusion was made by abnormal pulse volume recordings at the tip of fingers performed by photoplethysmography (Parks 1059 CTM, Parks Medical Electronics Inc. USA). Brachial and wrist artery pressure was measured using a Doppler probe (Parks 1059 CTM, Parks Medical Electronics Inc. USA). Abnormal pulse volume recording was defined as flattened waveform with loss of diastolic notch. The medical records of all patients were reviewed, abstracted and entered into a database. Data collection included patient's history at initial presentation with particular attention to work related injuries. The severity of symptoms, presence of ulcers or gangrene, and tobacco use was reviewed and it was considered whether the diagnosis of connective tissue disorder had been established. Digital ischemia symptoms were classified as asymptomatic, symptoms on provocation (e.g. cold), rest pain and ulcer or gangrene. Initial management was abstracted from the medical records.

In 2006, all 36 patients were invited to return to our vascular outpatient clinic for a follow-up visit. We were able to contact 29 patients, or their next of kin, out of the 36 patients, of which 24 of them came for a clinical follow-up visit. Five patients had died in the meantime and their medical records were used to abstract follow-up data. Of the seven patients who were lost to follow up, two had moved abroad, three refused the follow-up visit and two had moved within the country and could not be contacted. Assessment at follow-up visit included a careful review of the patient's history, focusing on the course of symptoms over the previous five years including finger ulcers, digital amputation, severity of symptoms, smoking habits, changes of occupation, workers' insurance compensation and physical examination. Non-invasive vascular work-up included blood pressure and pulses of both arms, an Allen test and auscultation for vascular bruits. Brachial and wrist artery pressures were measured using a Doppler probe. Descriptive statistics were used to analyse the data as a total and separated by aetiology.

In this study, the anonymity of the patients' data was retained and informed consent was received according to the requirements of the institutional ethical review board. The study was performed according to The Declaration of Helsinki.

Results

Between July 2000 and June 2001, 36 patients (25 men, 11 women) were diagnosed with digital ischemia due to digit-

al artery occlusions. Baseline characteristics and cardiovascular risk factors are given in table 1. The clinical severity of digital ischemia at baseline are given in table 2 and are separated by aetiology.

In 15 of the 36 patients (41.7%), a systemic disease was the underlying cause. Of those, 5 (33.3%) patients were found to have scleroderma, 4 (26.7%) patients had generalised atherosclerosis due to longstanding diabetes and/or renal failure, 2 (13.3%) had unclassified connective tissue disease, 1 (6.7%) patient had rheumatoid arthritis, 1 (6.7%) patient had antiphospholipid antibody syndrome, 1 (6.7%) patient had arterio-arterial embolisation from the brachio-cephalic trunk and 1 (6.7%) patient had newly diagnosed atrial fibrillation. One of the 36 (2.8%) patients with a history of intravenous drug abuse had an accidental intra-arterial injection. A total of 12 (33.3%) patients had a history of work-related hand and wrist strain, causing hypothenar hammer syndrome. Their occupations were: stonemason (n = 1), farmer (n = 2), scaffolder (n = 1), physical education teacher (n = 1), carpenter (n = 1), butcher (n = 1), mechanical engineer (n = 1), mechanic (n = 1), joiner (n = 1), lathe operator (n = 1) and car mechanic (n = 1). Two patients remembered a causative event at the onset of symptoms: The car mechanic experienced coldness and hypoesthesia in the right middle finger after hammering out a piece of metal; The mechanical engineer, who routinely used his hand as a hammer at work, noticed that his index finger was colder than the others after laying tiles in his garden with his bare hands, during which he again used his hand as a hammer. In 8 (22.2%) patients, no aetiology could be found.

The initial treatment of digital ischemia was based on the severity of the symptoms and the underlying disease. The treatment options applied to our patients are given in table 3 in total, and are separated into groups of underlying disease. Angiographic evaluation was performed in 9 of the 12 patients with hypothenar hammer syndrome. Digital artery occlusions were confirmed in all cases.

The severity of digital ischemia at 5 year follow up are given in Table 4 and are separated by aetiology. At the 5-year follow-up, the patient with digital ischemia due to arterial self-injection was asymptomatic but had had three fingers amputated. None of the patients with systemic disease had rest pain or ulcers or gangrene but four had died. None of the patients needed to change their profession and none of them claimed insurance compensation. Of the 11 initial patients with ulceration, 10 had healed without the need for an amputation. No reoccurrence of ulceration or gangrene was reported in any patient at follow-up. Of the 17 (47.2%) patients who smoked, only one patient with hypothenar hammer syndrome had stopped at follow-up. Eight patients with undetermined aetiology at initial presentation remained undetermined at follow-up.

Discussion

Digital ischemia has not been a focus of vascular research as the available literature on the natural history and clinical outcomes are limited by small sample sizes and frequently focused on the prevalence of connective tissue disease [5–7]. We are not aware of prior publications specifically addressing the clinical outcomes of patients with sympto-

matic digital artery occlusion in the last 20 years. Thus, we present herein long-term clinical outcomes of patients with digital ischemia diagnosed by abnormal photoplethysmography recordings, which has been shown to be as accurate as angiography in diagnosing digital ischemia [4].

Our results demonstrate that 41.7% of digital ischemia was caused by systemic disease and of those 53.3% were due to connective tissue disease. The second most frequent aetiology of digital ischemia was hypothenar hammer syndrome with a total of 33.3%. In 2.8%, self-inflicted occlusion of the palmar and digital arteries was the cause and in 22.2% the aetiology was undetermined despite thorough history taking, physical exam, laboratory tests and imaging. Occluded digital arteries are intimidating events for patients as they can limit their dexterity and ability to work. In the 1970s, two retrospectively analysed case series were published. In the first series, out of 65 patients diagnosed with digital artery occlusions over a 10 year period, 59% had systemic diseases, 19% had a traumatic aetiology, 19% had an iatrogenic aetiology, and 2% an unknown aetiology [9]. Another case series with 91 patients, excluding scleroderma, showed that 33% patients had an underlying disease and one (1%) patient had a traumatic digital artery occlusion [6]. A decade later, a retrospective analysis of 44 pa-

tients showed 50% of patients had systemic diseases, 9% had a traumatic aetiology and 41% had an unknown aetiology [4]. Thus, our results represent similar findings, which highlights the validity of the current study.

Severity of symptoms at initial presentation as well as digital amputation, severity of symptoms at long-term follow up and mortality differed, depending on the aetiology of digital ischemia in our dataset. Patients with systemic disease had a more severe clinical stage at presentation compared to the other groups. At follow-up, mortality in the systemic disease group was 40.0%, whereas none of the patients with hypothenar hammer syndrome had died. Digital amputation occurred in one patient who was a IV drug addict. A case series of 44 patients, where patients with digital artery occlusions with and without connective tissue disease were retrospectively compared, showed that patients with connective tissue disease had a more severe clinical stage and a worse clinical course of local symptoms at follow up [4].

In contrast to our expectations, most patients with hypothenar hammer syndrome had persisting symptoms, although with lessened frequency and severity. The prevalence of hypothenar hammer syndrome depends on the prevalence of predisposing occupations in the area. A study

Table 1: Baseline characteristics of 36 patients with hand and digital ischemia.

	Total (n = 36)	Systemic disease (n = 15)	Hypothenar hammer syndrome (n = 12)	No apparent cause (n = 8)	Arterial self-injection (n = 1)
	Number (%)*				
Age, mean ± SD, y**	55 ± 14	62 ± 13	45 ± 10	59 ± 13	34
Male	25 (69.4)	10 (27.8)	12 (33.3)	2 (5.6)	1 (100)
Current smoking	17 (47.2)	4 (26.7)	10 (83.3)	2 (25.0)	1 (100)
Dyslipidemia	3 (8.3)	1 (6.7)	2 (16.7)	0	0
Hypertension	6 (16.7)	4 (26.7)	0	2 (25.0)	0

*Number and percentage if not otherwise indicated
**SD: standard deviation; y: years

Table 2: Severity of symptoms at baseline in all 36 patients and separated by etiology.

Severity of symptoms	Total (n = 36)	Systemic disease (n = 15)	Hypothenar hammer syndrome (n = 12)	No apparent cause (n = 8)	Arterial self-injection (n = 1)
	Number (%)				
Asymptomatic	0	0	0	0	0
Symptoms on provocation	23 (63.9)	6 (40.0)	9 (75.0)	8 (100.0)	0
Rest pain	2 (5.6)	0	2 (16.7)	0	0
Ulceration or gangrene	11 (30.6)	9 (60.0)	1 (8.3)	0	1 (100.0)

Table 3: Initial management of patients with hand and digital ischemia.

	Total (n = 36)	Systemic disease (n = 15)	Hypothenar hammer syndrome (n = 12)	No apparent cause (n = 8)	Arterial self-injection (n = 1)
	Number (%)				
No treatment	16 (44.4)	6 (40.0)	4 (33.3)	6 (75.0)	0
Antithrombotic agents	14 (38.9)	7 (46.7)	6 (50.0)	1 (12.5)	0
Bier' block	3 (8.3)	0	2 (16.7)	1 (12.5)	0
IV prostaglandine	3 (8.3)	2 (13.3)	0	0	1 (100.0)

Table 4: Severity of symptoms and signs at 5-years follow-up.

	Total (n = 29)	Systemic disease (n = 10)	Hypothenar hammer syndrome (n = 10)	No apparent cause (n = 8)	Arterial self-injection (n = 1)
	Number (%)				
Asymptomatic	6 (20.7)	0	2 (20.0)	3 (37.5)	1 (100.0)
Symptoms on provocation	18 (62.1)	6 (60.0)	8 (80.0)	4 (50.0)	0
Rest pain	0	0	0	0	0
Ulceration or gangrene	0	0	0	0	0
Death	5 (17.2)	4 (40.0)	0	1 (12.5)	0

in the early 1970's found a 14% prevalence of hypothenar hammer syndrome in habitual hammerers, defined as persons who use the hand as a hammer at least once daily [10]. The Swiss National Accident Insurance (SUVA) company reported, in the year 2000, only 185 of 721'873 cases (0.03%) with work-related injuries to have digital arteries including open injuries of digital arteries [11]. In our series, the second most prevalent aetiology apart from systemic disease was hypothenar hammer syndrome. Those patients were exclusively men, usually craftsmen, younger than patients with typical atherosclerotic disease, which is in accordance with other studies [12, 13]. Thus, strenuous manual work has to be considered as a risk factor for digital artery occlusions. Remarkably, 83.3% of the patients with hypothenar hammer syndrome were identified as current smokers. Smoking may predispose vessels to thrombotic complications, hyperviscosity and endothelial dysfunction. It has also been suggested that smoking delays the recovery of hypothenar hammer syndrome [13]. As only one patient of the group stopped smoking, no conclusion regarding natural history of symptoms in current and former smokers can be made. Our study has some limitations which are worth mentioning. This group of patients was heterogeneous and thus diagnostic work-up had to be individualised. Although the aetiology of digital ischemia due to systemic disease or work-related strains seems high, it reflects data from a tertiary vascular referral centre and may not reflect the true prevalence in the population. We are also concerned about recall bias at follow-up. However, if patients had had a digital amputation, this would be obvious at the physical exam.

In conclusion, this is the first large series in the last 20 years assessing the long-term clinical follow-up of patients with digital ischemia demonstrating that systemic disease and hypothenar hammer syndrome were the most frequent aetiologies. Digital artery occlusions in patients without systemic disease are remarkably benign, although symptoms may persist with provocation, whereas patients with digital artery occlusions due to systemic disease have a high mortality rate.

Study funding / potential competing interests

All authors declare no conflicts of interest.

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