NON-DUPUYTREN'S DISEASE OF THE PALMAR FASCIA

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The typical Dupuytren's disease patient is of Northern European descent with bilateral progressive multiple digital contractures and is genetically predisposed, with a family history. Palmar fascial proliferations sometimes present as a different entity without the typical Dupuytren's disease characteristics. We identified 39 patients (20 women and 19 men) over a 4-year period with "Non-Dupuytren's palmar fascial disease", with unilateral involvement, without family history or ectopic manifestations. Twenty-three patients presented with unrelated complaints and were discovered, incidentally, to have the condition. In 28 patients, prior ipsilateral hand surgery or trauma precipitated the condition. Other related factors were diabetes mellitus and cardiovascular disease. Ten patients had skin tethering and subcutaneous thickening akin to Dupuytren's nodules and 29 had palmar fascial thickening into ill-defined pretendinous cords. The diseased tissue was in the line of the ring finger in 30 patients. The time from insult to onset of contracture averaged 3.6 months and from onset to follow-up averaged 5.3 years. The condition was non-progressive, or partially regressive, in 33 patients. Seven patients had operations for unrelated conditions and underwent simultaneous fasciectomy without recurrence. Environmental factors, especially trauma, surgery and diabetes, are important in the pathogenesis of Non-Dupuytren's palmar fascial disease, but these patients do not appear to be genetically predisposed for Dupuytren's disease. Typical Dupuytren's disease and Non-Dupuytren's palmar fascial disease are two clinical entities that run different courses and do not share a similar prognosis. This should be taken into account in future epidemiological and outcome studies.

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INTRODUCTION

The earliest reference to Dupuytren's disease was by Felix Platter in 1614. He described a stonemason's hand with palmar digital contracture. Henry Cline, in 1777, was the first to dissect a hand afflicted by progressive fibromatosis of the palmar fascia and identified the location of the disease precisely (Elliot, 1988). His seminal contribution was followed by several descriptions of the condition before that of Dupuytren. It was, however, the detailed description of a coachman with "permanent retraction of the fingers" in 1831 by Guillaume Dupuytren in Paris that earned him the eponym. Over the ensuing 70 years more than 250 articles were published discussing what became known as Dupuytren's disease (Elliot, 1999). In the last century, innumerable manuscripts have considered both the basic science and the clinical description of this enigmatic disease.

McFarlane et al. (1990) described the typical patient with Dupuytren's disease as a white male of Northern European origin, averaging 57 years of age, with bilateral hand and multiple digital involvements by the disease. The condition progresses over time, but at variable rates. Hueston (1991) described Dupuytren's disease patients as expressing disease diathesis with varying degrees of severity. Patients with severe diathesis frequently have an associated family history and exhibit fibromatosis at ectopic sites such as the dorsum of the

proximal interphalangeal joints (Garrod's nodes or knuckle pads), plantar fascia (Ledderhose's disease) and the penis (Peyronie's disease). This form of Dupuytren's disease is believed to be transmitted by an autosomal dominant pattern of inheritance the penetrance of which is mutable and age related (Burge, 1999; Ling, 1963). Risk factors are thought to include diabetes mellitus, alcohol consumption, smoking, treatment with anticonvulsant medication and hypercholesteremia (Burge, 1999; Ross, 1999).

Patients with palmar fascial proliferation are sometimes encountered without the typical disease findings previously mentioned (Rayan, 1999) and, yet, do not fit the mild diathesis variety that share many of the typical characteristics. We believe that this group of patients with atypical disease presents a separate entity from those with typical Dupuytren's disease. We suggest the characterization "Non-Dupuytren's palmar fascial disease" as a non-progressive palmar fascial proliferation that is distinguishable from Dupuytren's disease by the following: the patients have no family history, there is ethnic diversity, with blacks being affected, and there is no gender predilection. The condition is unilateral, confined to the hand and without ectopic manifestations. It involves the palm and may be in line with a single digit, without digital involvement. Although fascial disease is present, joint contracture, especially at the proximal interphalangeal joint, does not occur. Factors other than genetic ones play a role in its

pathogenesis, such as trauma, previous surgery and diabetes. Palmar fascial disease without digital contracture has been observed following hand trauma. This condition was first reported by Anderson (1891).

The purpose of this study was to report a series of patients with Non-Dupuytren's palmar fascial disease and identify characteristics that distinguish them from patients with typical Dupuytren's disease.

MATERIALS AND METHODS

From the records of a Hand Surgery practice over a 4-year period, we identified 49 Non-Dupuytren's palmar fascial disease patients with characteristics as described above. Inclusion criteria were patients with palmar fascial disease of at least 1 year in duration without the typical Dupuytren's disease findings, in particular family history, ectopic manifestations and bilateral hand involvement. Seven of the 49 patients could not be contacted and three patients were found at follow-up to have typical Dupuytren's disease because they developed digital contracture or involvement of the contralateral side during the course of the study, albeit non-progressive. The number of patients examined at follow-up was 39.

The reasons for presentation and seeking medical care were identified. Subjective and objective assessments were carried out.

Subjective evaluation

Each patient completed a questionnaire of his/her demographic data including age, gender, ethnicity, hand dominance and occupation. Additional risk factors, such as prior ipsilateral hand surgery or trauma, hypercholesterolemia, anticonvulsant medication, smoking, alcohol consumption and systemic diseases, in particular diabetes mellitus, were included. These variables were included either because we observed that they were related to Non-Dupuytren's palmar fascial disease or they are known to be risk factors for Dupuytren's disease.

A detailed history was then obtained from each patient with emphasis on the onset, degree and rate of progression of the disease. Patients were questioned about the specifics of any surgery or trauma that had preceded the appearance of the disease. They were asked to detail the time period from injury to onset of the disease, the time period from onset of the disease to the present and whether, or not, the condition seemed to be progressive, stationary or regressive. Any operations performed on the disease were also recorded along with recurrence, if any, after surgery.

Objective evaluation

Each patient was then examined and a record was made of the following: the involved hand, the site and extent of the disease and the presence, or absence, of any metacarpophalangeal or proximal interphalangeal joint flexion deformity. The patients were also examined for any ectopic disease, such as knuckle pads or foot disease. The findings at follow-up time were compared to previous findings in patients' records and notes were made of any progression of the disease.

RESULTS

The number of patients examined at follow-up was 39. Non-Dupuytren's palmar fascial disease was discovered incidentally in 23 patients seen for complaints unrelated to palmar disease. The unrelated problems in order of frequency were hand or wrist pain after injury, compression neuropathy, stenosing tenosynovitis and trapeziometacarpal arthrosis. Ten patients presented with subcutaneous and palmar fascial thickening similar to Dupuytren's disease nodules or cords and six patients had a combination of related and unrelated symptoms. Two of the later were fascial and skin disease following lacerations.

Subjective evaluation

The average age of these patients was 55 (range 25–77) years with 19 patients being younger than 60 years. Thirty-two patients were right handed, five left handed and two ambidextrous. Twenty-seven patients had involvement of the dominant hand. Twelve patients were retired, 18 were engaged in occupations that were not physically demanding and nine had physically demanding occupations. Nineteen patients were men and 20 were women. Thirty-four patients were white-skinned, two were African–Americans, two of Hispanic origin and one was Native American. Only six of the white-skinned patients were of North European origin. The remainder had a mixture of ancestry, including Europeans (from Turkey, Italy, Spain, etc), Asians and Middle Eastern.

The average time period from onset of the condition to follow-up was 5.3 (range 1.5–12) years. Six patients reported initial transient, but minimal, progression of the disease or tissue thickening, after which the condition was stabilized. Thirty-three patients either had partial regression (seven hands) or no changes (26 hands) in the disease after onset. Risk factors were classified into primary and secondary.

Primary risk factors

Twenty-eight patients had one of two primary risk factors, namely a history of surgery or trauma, prior to the onset of Non-Dupuytren's palmar fascial disease (Table 1). Fifteen patients had prior surgery on the affected hand and 13 had antecedent trauma to the ipsilateral hand or wrist. The average time from surgery

Table 1—Primary risk factors in the development of Non-Dupuytren's palmar fascial disease

Factors	Number of patients	Related information	Number of patients
Surgery	15		
		Carpal tunnel release	6
		Trigger finger release	4
		Trapeziometacarpal arthroplasty	2
		De-Quervain's release	1
		Ganglionectomy	1
		Wrist surgery	1
Trauma	13		
		Distal radius fracture	4
		Hand fracture/dislocation	4
		Palmar laceration	3
		Blunt trauma	2

Table 2—Secondary risk factors in the development of Non-Dupuytren's palmar fascial disease

Factors	Number of patients	Related information	Number of patients
Diabetes mellitus	11	Insulin dependent	7
Cardiovascular disease	11	Cardiac surgery	6
Alcohol consumption	10	Occasional	13
Hypercholesterolemia	10	Unknown	22
Smoking	7	< 1 pack daily	4
Seizure disorder	2	On Dilantin	0

or injury to onset of disease was 3.6 (range 1–20) months.

Secondary risk factors

Twenty-five patients had one of many secondary risk factors (Table 2). Fourteen of these patients also had primary risk factors. In 11, only secondary factors predisposed to Non-Dupuytren's palmar fascial disease.

Objective evaluation

All patients had unilateral involvement without ectopic disease. Twenty-nine patients had thickening of the palmar fascia or ill-defined, pretendinous cords localized to the palm and 10 patients had palmar skin tethering and subcutaneous thickening or ill-defined nodules. The diseased tissue was in line with the following fingers: ring in 30 patients, middle in three patients, little in two patients, index in one patient and middle and ring in three patients. Only six patients had metacarpophalangeal joint contractures, manifested as a loss of hyperextension or 5 (range 1–10) degrees of flexion deformity.

Two of these patients had skin contracture following lacerations and one had associated rheumatoid disease. The remaining 33 patients had no metacarpophalangeal joint contractures at follow-up after the onset of Non-Dupuytren's palmar fascial disease. No patients had contractures of the proximal interphalangeal joints.

Only seven patients underwent surgery, but for a variety of unrelated reasons. In addition to the reason for surgery, they underwent fasciectomy. Three patients had Z-plasties for hypertrophic palmar dermal scar after prior lacerations, two had rheumatoid metacarpophalangeal joint arthroplasty to facilitate rehabilitation and two had A-1 pulley releases for trigger fingers. Because of the pre-existing rheumatoid disease, it was difficult to ascertain how much the diseased palmar fascia contributed to the mild metacarpophalangeal joint flexion in the two patients who underwent joint replacement. None of the patients who underwent surgery developed subsequent joint contractures.

Two patients had histopathological examination of surgical specimens from hands that had had prior lacerations. The diseased tissue deep to the skin showed collagen compatible with scar tissue without any myofibroblasts.

DISCUSSION

Dupuytren considered the disease named after him to be occupational and due to local trauma Elliot (1988). Goyrand contested the relationship with manual work (Elliot, 1988) and cited his hospital manager with bilateral Dupuytren's disease who had never done one day of hard work. This debate continues today with proponents (Meagher, 1990) and opponents to manual work being a factor in the development of Dupuytren's disease. McFarlane (1991) found insufficient epidemiologic evidence to state that manual work either hastens the onset or progression of Dupuytren's disease, but pointed out that, occasionally, a single acute injury can precipitate the condition. The prevalence of trauma, acute and repetitive, as an aetiological factor in Dupuytren's disease has been reported to vary from 9% to 27% (McFarlane et al., 1990) and even 54% (Sladicka et al., 1996) in the black population.

Anderson (1891) considered Dupuytren's disease after acute trauma to be a different entity and called it "False Dupuytren's contraction, traumatic form". He described seven such patients. Gordon (1961) reported a similar condition following soft tissue trauma. A mild form of Dupuytren's contracture was reported to follow distal radius fractures (Cooney et al., 1980; Stern and Derr, 1993). In a series of 235 such patients, Stewart et al. (1985) reported an 11% incidence of "Dupuytren's Disease" which developed within 6 months after displaced distal radius fractures; all were mild, in the form of palmar nodules or skin pits in the palm, in line with the ring or middle finger, except one with a digital

nodule. Of the 23 patients reported, only one was male, only two patients had mild joint contracture and none required surgery. Kelly et al. (1992) observed 12 of these patients at a later date. In 11 patients, the disease was static. In one, the disease had progressed in the same hand. In seven cases of unilateral fractures, disease had transfer to the other hand. Lanzetta and Morrison (1996) reported three further cases of "Dupuytren's disease" triggered by minor hand surgery, viz. carpal tunnel decompression, trigger finger releases and foreign body removal. All patients had a mild, unilateral, nonprogressive condition without ectopic disease. The contracture developed 3 weeks to 3 months after surgery. Elliot and Ragoowansi (2005), in a historical review, collected 385 cases of "Dupuytren's disease" secondary to acute injury, operation or infection on the ipsilateral limb, including 52 cases of their own. Only six of their 52 patients required surgical treatment and one patient had bilateral contracture. These cases, most probably, all fall into the category of "Non-Dupuytren's palmar fascial disease". It seems that acute, rather than chronic, repetitive trauma is a factor in development of Non-Dupuytren's palmar fascial disease.

We have reported a series of patients with Non-Dupuytren's palmar fascial disease. We found the condition to be asymptomatic, without compromise to hand function and often discovered incidentally. Although encountered among white skinned people of Northern European stock, any ethnic group can be affected. The patient's ethnicity and gender are less important factors in Non-Dupuytren's palmar fascial disease and family history is absent. We found the primary predisposing factors to be antecedent trauma or surgery to the affected hand and these were present in 28 of 39 patients. Most of these patients develop Non-Dupuytren's palmar fascial disease 3.5 months after hand surgery or injury. The main secondary factors were diabetes, cardiovascular disease, hypercholesterolemia and alcohol consumption. Fourteen patients had a combination of primary and secondary factors. Ectopic manifestations are lacking in Non-Dupuytren's palmar fascial disease and the condition is unilateral, isolated to the palm, most frequently in line with the ring finger, without digital involvement or flexion deformity of the proximal interphalangeal joint. Metacarpophalangeal joint contracture is very rare and, if present, averages 5° and is often non-progressive or, sometimes, partially regressive. Surgery is usually not indicated but, if carried out, recurrence is probably rare. Long-term follow-up to confirm non-progression is important for diagnosing Non-Dupuytren's palmar fascial disease. We feel that it is important to distinguish between Non-Dupuytren's palmar fascial disease and Dupuytren's disease. Table 3 is a comparison between Non-Dupuytren's palmar fascial disease and Dupuytren's disease.

Diabetes mellitus and cardiovascular disease were the most frequently encountered secondary risk factors among our Non-Dupuytren's palmar fascial disease patients. Arkkila et al. (1997) have shown that Dupuytren's disease is prevalent among diabetic women and one third of diabetic patients have a mild, nonprogressive form of Dupuytren's disease (Chammas et al., 1995). The duration of diabetes increases the incidence of Dupuytren's disease, which can be as high as 67% at 20 years (Ross, 1999). Hurst and Badalamante (1990) have observed that the severity and location of Dupuytren's disease in diabetic patients are different from the typical patient. Nodules and skin tethering occurred without digital contracture. They speculated that diabetic Dupuytren's disease may be a different condition from the non-diabetic disease. McFarlane et al. (1990) recognized that patients with Dupuytren's contracture and diabetes do not seek surgical intervention. Mild Dupuytren's disease was also reported in patients with rheumatoid arthritis and diabetes (Rayan, 1988). Berry (1963) observed an association of palmar fascial thickening "similar to Dupuytren's contracture", shoulder-hand syndrome and syringomyelia patients. He also described thickening of the palmar fascia among patients with myocardial infarction. Plewes (1956) reported the development of de novo palmar fascial disease, mostly without progression, in 33 of 34 patients who had developed reflex sympathetic dystrophy following upper limb injuries.

Table 3—Characteristics of Dupuytren's disease and Non-Dupuytren's palmar fascial disease

Characteristic	Dupuy tren's disease	Non-Dupuytren's disease
Family history	Present	Absent
Gender distribution	Male predominance	No gender predilection
Ethnicity	Caucasian/Northern European	Random
Primary predisposing factors	Genetic	Environmental: surgery or trauma
Diseased tissue	Palmar or digital cord	Palmar subcutaneous/fascial thickening, no digital involvement
Digital contracture	Present	Usually absent
Digital involvement	Multiple	Single
Side affected	Bilateral	Usually unilateral
Ectopic disease	Present	Absent
Course	Progressive	Non-progressive

In a review of Dupuytren's disease epidemiology, Ross (1999) found that the reported prevalence rates varied from 2% to 42%. Similarly, recurrence rates after surgery for Dupuytren's disease vary from 0% to 66% (Hueston, 1969; Tubiana and Leclercq, 1985). The disparity among epidemiologic and outcome studies of Dupuytren's disease is probably multifactorial including difficulty in accurate early diagnosis of the affected population. However, it may also include lack of distinction between these two types of palmar fascial disease, namely Dupuytren's disease and Non-Dupuytren's palmar fascial disease. Excluding Non-Dupuytren's palmar fascial disease patients from Dupuytren's disease population studies may yield more accurate prevalence data. Burge (1999) stated that, although there is strong evidence for an inherited susceptibility for Dupuytren's disease and pedigrees exhibiting the autosomal dominant model, there are many sporadic cases. Many of these, probably, represent the Non-Dupuytren's palmar fascial disease variety.

Although Dupuytren's disease is most prevalent among white-skinned individuals from Northern Europe, it is not rare among Orientals (McFarlane et al., 1990) and has been reported among Blacks (Mennen, 1986; Mitra and Goldstein, 1994; Muguti and Appelt, 1993; Sladicka et al., 1996), Indians (Srivastava et al., 1989), Japanese (Egawa, 1985), Chinese (Liu and Chen, 1991) and other Orientals races (Maes, 1979).

Patients reported with "Dupuytren's disease" from the non-Caucasian races often lack family history (Mitra and Goldstein, 1994) and their conditions are mild, confined to the hand, usually unilateral (Maes, 1979) and, when treated surgically, have no recurrence (Srivastava et al., 1989). Many reported cases of palmar fascial contracture among non-Caucasians are probably the Non-Dupuytren's palmar fascial disease variety. Only 6 of 34 white-skinned patients in our study were of Northern European origin. While palmar fascial disease among Northern European, white-skinned people is often, but not always, of the typical variety, these patients can develop Non-Dupuytren's palmar fascial disease. Future epidemiologic and outcome studies should take into consideration these two clinical entities and they should be investigated separately.

Non-Dupuytren's palmar fascial disease is probably a local response of the palmar tissue to environmental (mechanical or chemical) factors, whereas in true Dupuytren's disease, most probably, genetic factors play a primary role in its pathogenesis. Histopathological examination of the surgical specimens was done in two, but not all, seven patients, because these patients had surgeries for a variety of conditions other than Non-Dupuytren's palmar fascial disease. Although these findings were compatible with scar tissue rather than those of typical Dupuytren's disease, histological assessment of specimens from all seven patients would have been more helpful. Histologically, the affected tissues of Dupuytren's disease and Non-Dupuytren's

palmar fascial disease may not be different, but the pathogenetic mechanisms that trigger the process at the molecular level are, probably, dissimilar.

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