Dupuytren's Palmar Contracture in Women

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Key words: Dupuytren's disease, palmar contracture, women, ethnicity, fasciectomy

Abstract

Background: Dupuytren's disease is a fibroproliferative disorder of the palmar fascia that can cause disabling digital contractures. The pathogensis of the disease is still unclear, and it afflicts predominantly white males of northern European origin. Gender-related differences of Dupuytren's disease and the distinctive characteristics of the disease in females are not yet well defined.

Objectives: To evaluate and illustrate the distinctive characteristics of Dupuytren's disease in females.

Methods: A retrospective study was performed of all female patients with Dupuytren's disease seen and followed at our Hand Surgery Unit over a 20 year period. The study group consisted of 48 women (56 hands). The collected data included clinical and epidemiological features on admission, and outcome of surgical intervention.

Results: Of the 48 women (56 hands) with Dupuytren's disease, 23 (26 hands) underwent limited fasciectomy. The average age at presentation was 60.1 years. A few of the patients originated from Asia and Africa. Manifestations and pattern of the disease were nearly comparable to those observed in the male group, except for a slightly higher incidence of proximal interphalangeal joint contracture in female patients. Generally, females expressed less severe contractures on presentation and a slower progression thereafter. A favorable functional postoperative outcome was observed. Seven patients had minor complications including local hematoma and painful scars. Two patients developed moderate signs of complex regional pain syndrome.

Conclusions: Further investigations are needed to assess the potential role of androgens in the pathogenesis of Dupuytren's disease, and a possible protective role of estrogenic hormones, rendering Dupuytren's contracture a postmenopausal affliction.

IMAJ 2008;10:445–447

Dupuytren's disease is a benign fibroproliferative disorder of the palmar fascia that progresses slowly but can result in disabling contractures of the digits [1]. Both genetic and environmental etiologies have been postulated. In many reported pedigrees, the inheritance appears to follow an autosomal dominant model. Features such as age at presentation and severity of disease may be related to variable penetrance of gene expression. The genetic expression is partially limited to males (partial sex limitation). However, other modes of inheritance are consistent with the published data [2]. The association of this disease with alcoholism, smoking, diabetes, liver failure, seizure disorders, and trauma supports an environmental etiology [1].

The majority of patients reported with Dupuytren's disease are native to or descendants of ancestors from northern Europe [3]. While other epidemiological aspects of Dupuytren's contracture, especially relating to prevalence and incidence in different populations, have been repeatedly addressed in previous studies, only a few reports in the literature address gender-related differences in terms of incidence, presentation, natural history, and outcome of surgical intervention. The distinctive characteristics of the disease in females are not yet well defined.

PIP = proximal interphalangeal

Patients and Methods

The medical charts of all female patients with Dupuytren's disease who were seen and followed for at least 1.5 years at our unit between the years 1986 and 2005 were reviewed. For each subject, the data included age at presentation, reasons for seeking medical evaluation, time lapse before seeking medical advice, dexterity, affected side, family history of the disease, systemic diseases and pertinent associated conditions, location and pattern of contracture, modality of treatment, postoperative results, functional outcome, complications, extension and recurrence. In addition, demographic data were collected looking at the geographic region of origin and self-reported racial descent. All demographic details relied on the information disclosed by patients and related entirely to the self-determined identity.

Diagnosis of Dupuytren's disease was based on history and physical examination. Surgical intervention was recommended for patients with more than 30º flexion contracture of the metacarapophalangeal joint, or less than 30º flexion contracture associated with any flexion, contracture of the proximal interphalangeal joint; or a progressing PIP joint flexion contracture of even less than 30º [Figure 1].

Surgical intervention

The approach is though a zigzag digitopalmar incision extending from the palm to the distal interphalangeal joint of the affected ray. Z-plasty incisions were applied when necessary. Skin flaps are developed and elevated. Wide exposure is achieved to permit dissection and identification of the neurovascular bundles; tracing, protecting, and keeping them under constant supervision, while enabling a complete excision of the diseased tissue. In cases of severe PIP joint contracture, digital fasciectomy was followed as needed by gentle passive joint manipulation and sequential capsuloligamentous surgical release, including the checkrein ligaments.

Results

Of the 317 patients (371 hands) with Dupuytren's disease seen at our unit between 1986 and 2005, 269 were men (315 hands) and 48 (56 hands) were women. Male to female ratio was 5.6:1.

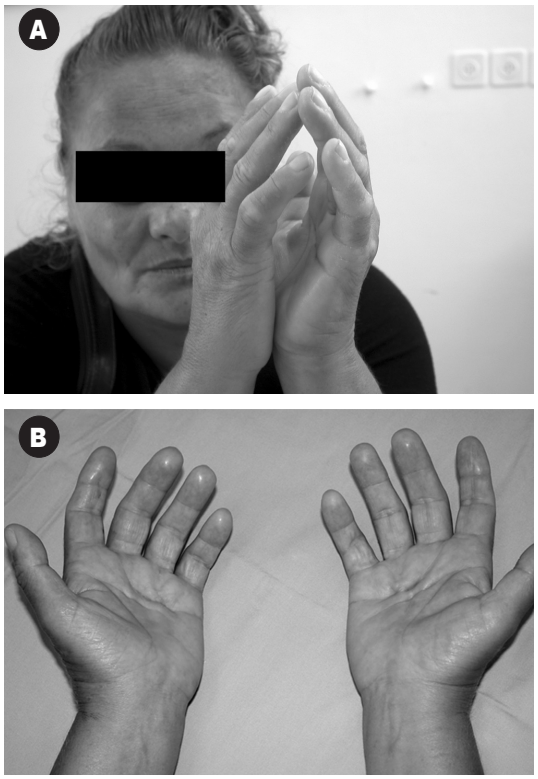

Figure 1. Patient with bilateral contracture of both the ring and fifth rays, **[A]** mainly involving the PIP joints in the small fingers. **[B]** Note the thickened palmar fascia with cords ending in firm nodules, characteristic of Dupuytren's disease. The patient subsequently underwent partial fasciectomy of the right hand due to progression of flexion contracture.

With regard to the female group, mean follow-up was 2.5 years (range 1.5–19 years). Average age at presentation was 60.1 years (range 42–76 years). The patients presented on average 2 years after the disease was first noticed (range 10 months to 5 years). All patients complained about some functional disability of the affected hand. Seven patients were concerned about the unsightly appearance and the cosmetic blemish posed by the disease. Twelve were unable to wear a ring. Six were concerned about a potential neoplastic nature of the disease.

Thirty-five patients were Ashkenazi Jews, originating from North America, Poland, the former USSR, Romania, England, and Australia. Ten were Sephardic Jews, originating from Asia and North Africa. Three were Arabs (two Christians and one Moslem). The right hand was dominant in 41 patients and the left in 7. The disease involved 45 right hands and 11 left hands. Eight patients had the disease in both hands. The small finger was involved in 25 hands, the ring finger in 21, and both small and ring fingers in 10 hands. Four women had PIP joint contracture with no evidence of the disease elsewhere. One patient had knuckle pads. Four patients reported a family history of the disease. Three had diabetes. None of the patients reported a history of alcohol consumption or having done manual labor.

Limited fasciectomy was performed in 26 hands of 23 patients who fulfilled the criteria for surgical intervention. The other 25 patients (30 hands) were observed and examined periodically. On follow-up examination of the operated women, PIP joint and MCP joint extension improved considerably [Table 1], with favorable functional outcome reported by all patients. Patients with long-term PIP joint contracture or preoperative PIP joint contracture over 60º failed to regain full extension on follow-up. Three patients had a postoperative local hematoma that resolved uneventfully. A "flare reaction" manifesting itself as tender swelling and redness was observed in two patients starting in the third postoperative week. None of the patients had an injury of the digital nerve. Four patients had a painful scar. On follow-up, three patients had late local extension of the disease that did not necessitate additional intervention. Five had recurrent disease; one subsequently underwent dermatofasciectomy and skin grafting.

Discussion

The male to female ratio as indicated in previous reports in the literature ranges from 5.9:1 to 15:1 [5-7], with the frequency in women catching up to that in men later in life [8]. Hueston reported an equal incidence in both genders after the age of 40 [7]. A total of 269 male patients (315 hands) with Dupuytren's disease were seen at our unit within the same period. Male to female ratio was 5.6:1. This relatively high representation of females in our series may be attributed to the higher age profile of our patients.

In an attempt to determine the role of androgens in the male predominance of the disease, Pagnotta et al. [9] showed that the expression of androgen receptors in Dupuytren's contracture is considerably higher than in the normal palmar fascia. Later on, Pagnotta and team [10] showed also that the palmar fascia is a target tissue for androgen action via androgen receptors.

The average age at presentation was 60.1 years and 62 years in female and male groups, respectively. Ling [11] reported a peak incidence of the disease in men around the age of 50. It has previously been suggested that women tolerate the contractures better than men and are less likely to seek medical treatment [7]. This may explain the relative delay in presentation averaging 2 years in the female group compared to 1.2 years (range 5–42 months) in the male group of our series. Concern about an unsightly appearance posed by the disease and about a potential neoplastic nature of the disease as a main complaint was rare among male patients in our series, but was a chief complaint in 27% of our female patients.

MCP = metacarpophalangeal

Table 1. Improvement of flexion contracture

Joint	Mean preoperative contracture	Mean postoperative contracture	Mean improvement of extension
MCP	40º (range 10–60º)	1º (range 0–15º)	97.5%
PIP	35º (range 5–70º)	12º (range 0–35º)	66%
DIP	10º (range 0–15º)	0º	100%

MCP = metacarpophalangeal, PIP = proximal interphalangeal, DIP = distal interphalangeal

The epidemiology of Dupuytren's contracture has attracted the attention of surgeons almost from the first description of the disease [3]. Certain populations show a particularly high prevalence of disease [12]. The majority of patients with Dupuytren's disease are native to or descendants of ancestors from northern Europe [3]. A striking geographic variation in population prevalence was reported [2] with exceptionally high rates of prevalence in Scandinavian and Celtic populations. The Middle East, Greece and the Orient are considered areas in which the condition is virtually unknown [12,13]. Rambam Medical Center in Haifa is the largest hospital in the north of Israel and serves a large population that offers a representative model of the Israeli demographic landscape. In contrast to the previously reported epidemiological data, our series (a total of 317 males and females) included patients from virtually all communities of the Israeli demographic landscape. Overall, 229 patients (72.2%) were Ashkenazi Jews, 63 (21.5%) were Sephardic Jews, and 20 (6.3%) were Arabs.

Manifestations of the disease, location, and type of contractures were nearly comparable to those observed in the male group, except for a slightly higher incidence of proximal interphalangeal joint contracture in female patients. Generally, females had fewer severe contractures on presentation as measured by joint contractures in comparison to males; the pattern of contracture however was similar. The disease in women followed a slower course in comparison to men. Sixteen female patients followed for at least 5 years showed hardly any progression in their clinical signs. Only one patient had an ectopic lesion manifesting as a knuckle pad. The disease was bilateral in 8 patients (17%), while the previously reported incidence of bilateral involvement is between 38% and 52% [6,14]. In concordance with previously reported series [6], the small finger was most frequently involved, and a predilection for the right hand was observed also in our series.

Risk factors, which indicate an environmental input to the etiology of the disease, including alcohol consumption and a history of manual labor, were absent in our female patients, and diabetes as a concomitant disease was less prominent than in males.

The favorable functional outcome reported by all our patients concurs with the report of Tonkin et al. [15], who believed that women treated by fasciectomy did as well or better than men, and contradicts Wallace's report [14] that 10 of 16 women (59%) between age 39 and 59 who were treated surgically for Dupuytren's disease had a "bad" result.

Seven patients had minor complications including local hematoma and painful scars. Two patients developed moderate signs of complex regional pain syndrome including diffuse swelling, hyperemia, dysesthesias, and pain out of proportion to that expected. No significant residual finger stiffness was observed. This postoperative complication, also referred to as a "flare reaction," has been reported to occur in 5–10% of patients [5-6]. Zemel and collaborators [6] postulated that female patients are twice as likely as men to have this complication, and reported that patients who had a carpal tunnel release at the time of operation for treatment of Dupuytren's disease, or those who had an extensive fasciectomy, were more apt to have a flare reaction. The term flare reaction was popularized by Howard in the late 1950s [16], was adopted thereafter by other authors, and actually expresses the complication currently termed "complex regional pain syndrome."

Only one patient developed a significant recurrent disease, which necessitated a second surgical intervention, compared to the male group where a higher rate of recurrence was observed.

Further comparative clinical studies are required to better refine the gender-related differences in the prevalence of the disease, clinical presentation, natural history, and surgical outcome of Dupuytren's disease. The questions whether control over androgenic action may alter the evolution of the disease, and whether estrogen hormones play a protective role, making females less liable to develop the disease, and rendering Dupuytren's contracture a postmenopausal affliction are yet to be investigated.

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