Development of Peyronie's and Dupuytren's diseases in an individual after single episodes of trauma: A case report and review of the literature

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A case is presented in which a patient experienced the development of both Dupuytren's disease and Peyronie's disease after single episodes of sports-related trauma. These disorders and other fibromatoses are linked not only by similar pathologic features but by increased frequency of simultaneous occurrence. Some genetically predisposed individuals experience the development of the disorders after trauma or after some other factor unmasks that predisposition. A review of the literature with emphasis on the relationship between these fibromatoses and the varied nonsurgical attempts at treatment is presented. (J Am Acad Dermatol 1999;41:106-8.)

Peyronie's disease (PD) and Dupuytren's disease (DD) are similar disorders of fascia that are known to occur together in the same patient with a higher frequency than that expected by chance.[1-3] The patient described in this article experienced the development of PD after impact injury to the penis during a snowboarding accident. Five years later, blunt impact trauma to the palm was followed by the development of DD. In this patient, these two similar disorders occurring after single episodes of trauma, along with a family history of DD, support theories that trauma may unmask an inherited diathesis for both disease processes.

CASE REPORT

The penis of a 41-year-old man was impacted against the pubic bone and the hard packed snow during a fall while snowboarding. This resulted in mild pain. No bruising or edema of the penis was noted. That night, mild pain was noted during sexual intercourse. Mild pain continued over the next few days and then disappeared completely. Over the course of 6 weeks, painful nodules developed over the shaft of the penis along with painful, dorsal curvature during erections. A diagnosis of PD was made. Although pain subsided 2 months after appearance of the nodules, dorsal penile curvature of approximately 30 degrees has continued without improvement or worsening for 6 years, despite treatment with empiric remedies such as oral and topical vitamin E and oral potassium aminobenzoic acid and attempted intralesional cortisone injections into the dense, impenetrable fibrous nodules. Sexual function has not been impaired since the remission of painful erections 3 months after the onset of the disorder.

Five years later, the patient used his right palm to hammer a boom end into position during a windsurfing outing. Three weeks later, nonpainful firm nodules of the 4th and 5th rays of the palmar fascia developed just proximal to the distal palmar crease. The nodules measured 0.8×1.2 cm and 1.0×0.9 cm. The overlying skin was bound down or tethered to the nodules. Slight contracture of the ring finger at the metacarpophalangeal joint was noted 2 weeks later. Knuckle pads and evidence of plantar fibromatosis were absent. The patient's father experienced the development of Dupuytren's contracture of the same hand at approximately the same age. The patient's grandparents had immigrated from Sweden and Italy. The patient had neither epilepsy nor diabetes mellitus and did not drink alcohol or smoke cigarettes. A diagnosis of DD was made. The palmar nodules were injected with triamcinolone acetonide suspension, 2.5 mg, on 3 occasions, 2 weeks apart. The nodules subsequently decreased significantly in size but did not completely regress. Ring-finger contracture was not evident 24 months after the nodules first appeared. The patient continues to apply tretinoin cream, 0.1%, and clobetasol propionate ointment, 0.05%, twice daily to the palm without perceptible change in the nodules.

The patient has experienced pain in the right shoulder and forearm, stiffness, and decreased range of motion for the past 5 years. No clear-cut history of acute trauma to the region has been noted. Nuclear magnetic resonance scans and radiographs of the shoulder have not revealed pathologic features.

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DISCUSSION

PD is a localized disorder of the tunica albuginea characterized by fibrous nodules and curvature of the penis most often occurring in middle-aged men.[4,5] The cause is uncertain but certainly complex. Trauma followed by aberrant wound healing, occurring during sexual activity or bluntly to the flaccid penis, is a suspected cause.[1-3] The skewed incidence of the disease in middle age leads to speculation that as the fascia becomes less pliant, it becomes more susceptible to injury during sexual intercourse.[3] Trauma may be an unmasking event that allows expression of the disorder in those men who are genetically predisposed.

A strong diathesis in some families, autosomal dominant inheritance and linkage of histocompatability antigens have all been observed in cases of PD.[6-9]

DD is characterized by the development of palmar fibrous nodules and bands that may eventually cause contracture of the fingers. Most often flexion contracture affects the ring finger.[10] Many patients with this disorder manifest Garrod's nodes, also known as knuckle pads, which are thickenings of skin and fibrous tissue over the finger joints.[11] In addition, the patient with DD is at risk for plantar fibromatosis, also known as Ledderhose's disease. Both Garrod's nodes and Ledderhose's disease are pathologically similar DD and PD. Ledderhose's disease is manifested as nodules affecting the non–weight-bearing midsole area of fascia. The nodules are often discovered after they become large enough to cause pain. For reasons unique to the anatomy of the plantar fascia, contracture of the toes is rare.[12,13]

Other sites of fibromatosis have been found in association with DD. These sites include the shoulder, arm, spine, anterior leg, popliteal and antecubital spaces, volar wrist fascia, deep flexor tendons of the forearm, and neck and around the hip and on the auricular conchae.[14,15]

The reported incidence of DD varies widely with the population group studied. It is most common among middle-aged and older men of northern European extraction, with reported incidence of 20% to 30% in that population.[16]

DD is thought to be an autosomal dominant disorder with variable penetrance.[17] Studies of HLA and autoantibodies lend support to an immunogenetic basis in DD.[18] The degree of penetrance seems to be affected by some additional stimulus, possibly such as diabetes, epilepsy, cigarette smoking, or alcoholic liver disease with which it is seen with increased incidence. Trauma may also be responsible for unmasking the diathesis.[19]

When considering the similarities of the pathologic process of fibromatoses alone, one is tempted to link DD and PD. However, further evidence for such linkage is supported by other findings. DD occurs simultaneously in 10% or more of cases of PD, and strong examples of familial cases manifesting both disorders have been reported.[1,2] Evidence exists for autosomal dominant inheritance for both disorders.[8,17] Both disorders are thought to be diatheses, which may be unmasked by an environmental stimulus such as trauma.[1-3,19]

Most of the literature concerning treatment of these disorders addresses surgical repair to return function. In PD, surgery is considered useful and appropriate only when the patient is unable to penetrate during coitus because of penile deformity.[20] Similarly, corrective surgical procedures are used for DD only when the point of significant functional impairment is reached.[21] Many patients with PD and DD do not reach the point of such loss of function or, if they do, they often elect to forego surgery.

Nonsurgical treatments of PD include intralesional cortisone injections,[22] oral and topical vitamin E,[23] oral potassium aminobenzoic acid,[24,25] oral colchicine,[26] intralesional verapamil,[27] intralesional injection of the anti-inflammatory metalloprotein orgotein,[28] tamoxifen by mouth,[29] radiotherapy,[30] ultrasound,[31] and most recently, intralesional interferon gamma injections[32] and parathyroid hormone.[33]

Reports of nonsurgical treatment in DD are less prolific but include topical cortisone in combination with tretinoin,[34] intralesional cortisone,[11,14] intralesional interferon gamma,[35] radiotherapy,[36] and colchicine.[37] Unfortunately, none of these nonsurgical treatments for PD or DD has been shown to be efficacious with any degree of respectable consistency.

Why should the dermatologist be concerned with these disorders? Knuckle pads, or Garrod's nodes, are often associated with DD and may precede or postdate palmar fibromatosis.[11] The dermatologist discovering Garrod's nodes is in a position to search for early fibromatosis of the palmar and plantar fascia or at least warn the otherwise unaffected patient that he may be susceptible to the development of palmar and plantar fibromatoses and PD. Garrod's nodes are found in many cases of DD. Plantar fibromatosis is also present in 9% to 65% of cases.[38-40] Surely an early warning to avoid palmar trauma is justified in a patient with Garrod's nodes, a family history of Dupuytren's contracture, and northern European ancestry.

The patient with PD, DD, or Ledderhose's disease may be examined, especially early on, by the dermatologist. During early stages of these disorders, the

more cellular fibrous nodules may be more amenable to one of the nonsurgical treatments noted earlier. Discovery of an early palmar nodule before finger contracture has occurred at the very least will allow the uncomplicated, safe but unproven empirical approach of a topical steroid in combination with tretinoin as recommended by Shelley and Shelley.[34]

In conclusion, this case illustrates the link between PD and DD. Although trauma may be the precipitating cause of the development in both disorders, those men afflicted may have an underlying genetically determined diathesis for these fibromatoses. Knowledge of the linkage of these disorders may allow early discovery of those men at risk and the institution of nonsurgical treatment at a point when they may be more amenable to treatment.

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