Dupuytren's Contracture (Palmar Fibromatosis) Extending over the Arm

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A rare case of Dupuytren's contracture, with associated fibrosis extending over the arm, is reported.

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CASE REPORT

A 65-year-old man was referred to the Dermatology Department with a 1-month history of fibrosis of the right palmar skin. He suffered concurrently from insidious progression of hardening of the anterior surface of the right arm. He had not experienced any trauma; nor was there any notable past history except for pulmonary tuberculosis. His family history was negative for similar lesions. At his first visit to the Dermatology Clinic, he presented with several indurated subcutaneous nodules measuring not more than 1 cm in diameter and partially forming cords on the ulnar side of the right palm. This was accompanied by flexion contracture of the right ring finger and little finger. The right arm also exhibited flexion deformity and restricted movement at the right elbow joint (Fig. 1). The indurated plaque covered almost the entire anterior surface of the same arm. The biopsy specimens from the lesions of the palm and the proximal portion of the forearm were stained with hematoxylineosin (HE), Masson trichrome and Verhoeff-van Gieson. The palmar section stained with HE showed subcutaneous nodular thickening of the fascia (Fig. 2). This tissue was more fibrotic than cellular. Elongated and compressed cells were separated by thick wavy bundles of collagen. The findings for the HE-specimen of the thickened fascia from the forearm lesion were consistent with those for the palmar lesion, while fibrous thickening of the fascia was also evident. With Masson trichrome, both the tissues stained blue throughout. With Verhoeff-van Gieson's stain, nodules from both locations stained predominantly red-purple with black sparsely scattered throughout. These findings implied that proliferation of elastic fibers was minimal in the fibrous tissue. Laboratory data were unremarkable, including those for serum LDH and CPK activities, serum anti-nuclear antibody, C₃ and antinative DNA antibody levels. The case was assumed to have developed from Dupuytren's contracture and to have manifested over the palm and arm. The patient did not wish to be operated on.

DISCUSSION

Dupuytren's contracture, also known as palmar fibromatosis, is a fibromatous hyperplasia of the palmar fascia, associated with dimpling of the skin covering the fascia, and usually forms cords extending axially to each of the flexed fingers (1). The condition may provoke insidious progression of flexion contracture at one or more digits. The typical hand deformity was named after Baron G. Dupuytren and shows an incidence of around 2% among the general population (2), while the age of onset is generally between 30 and 50 years (3). The condition

occurs more commonly in patients with alcoholic cirrhosis, epilepsy and diabetes mellitus (4, 5). However, the basic cause is obscure. Histopathologically, the palmar fascia, especially on the ulnar side, is thickened with a combination of fibrous tissue and focal nodules (6, 7). The nodules are composed of a dense collection of fibroblasts in various states of maturation and form a heavy reticulum network. As the fibroblasts mature, rather thick acellular collagen bundles develop, although the fibrosis does not involve tendon sheaths. Furthermore, no new formation of elastic fibers takes place. Treatment often consists of partial fasciectomy, although the more nodular lesions may respond to intralesional corticosteroid injections in their early stage (8, 9).

In the present case, the clinical and histopathological findings of the right hand corresponded to the features of Dupuytren's contracture. The ipsilateral arm clinically manifested hardening of the anterior surface, and the histopathological findings for its fascia were highly compatible with those for Dupuytren's contracture.

In some patients with this disease, there is an association with plantar fibrosis, usually present over the medial side of the soles, while one or more nodules may become painful (5, 10). Finally, a similar fibrosing condition involving the dorsum of the penis, known as Peyronie's disease, is occasionally associated with fibrosis of the palmar fascia (1). In a series of 159 cases with Dupuytren's contracture, Hueston (11) reported plantar fibromatosis in 12%, knuckle pads in 42%, and bilateral palmar fibromatosis in 89%. In a separate series of 22 patients with Dupuytren's contracture, he found that 27% had Peyronie's disease. However, associated fibrosis at other sites has so far been, to the author's knowledge, quite unknown in the literature.

It is therefore assumed that this represents the first case of Dupuytren's contracture associated with fibrosis of fascia extending over the arm, or the disease developing over not only the hand but also the arm.

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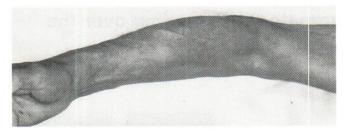
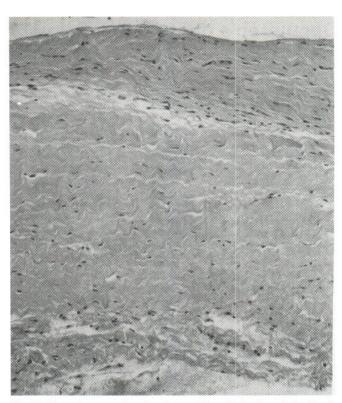


Fig. 1. A flexion deformity of the right arm.

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 $\it Fig.~2$. The palmar section showing nodular thickening of the fascia (HE).