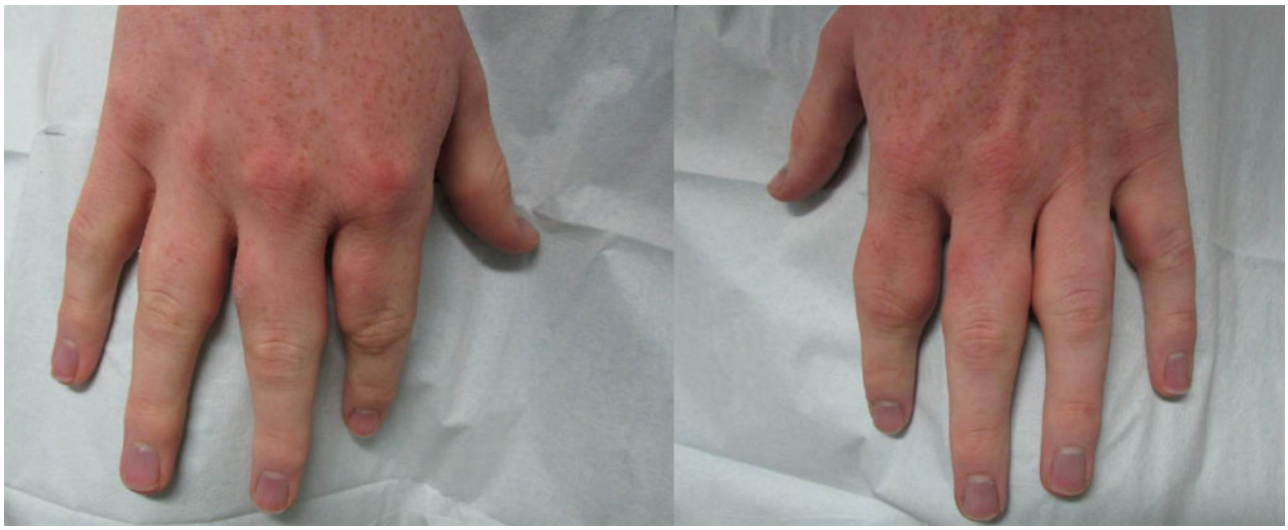
Recognizing the clinical presentation of pachydermodactyly

Pachydermodactyly (PDD) is a unique, benign soft tissue disorder affecting the hands, first reported in 1975. Patients with PDD typically present with painless swelling at the middle joint of the fingers, although in rare cases this swelling has been reported to spread to the knuckles, and even the palm or back of the hand. Patients with PDD are commonly young and male. Both hands are usually affected. Only approximately 90 cases of PDD have been reported worldwide.

Because of how rare the disease is, many hand surgeons and rheumatologists may be unfamiliar with PDD's clinical characteristics. Normally, swelling at the joints raises the possibility of some form or rheumatoid or inflammatory joint disease. This is concerning because an attack by an immune system gone awry can ultimately result in destruction of the body's bony joint surfaces and soft tissue structures, causing severe pain and dysfunction. However, the swelling seen in PDD is unique in that it is limited to the skin and does not spread to the joints. Diagnostic tests such as x-rays and MRI in patients with PDD demonstrate normal joint structures, indicating there is no injury to the bone or cartilage. Blood tests reveal no inflammatory markers and surgical biopsy, if performed, is generally non-diagnostic.



Distinctive bilateral swelling of the skin localized to proximal phalanges II-V.

We reported a single case of PDD in a 19-year old male patient. Swelling first affected his index finger before gradually progressing to affect the middle, ring, and small fingers of both hands over a 4-year period. Previous consultations with hand surgeons and dermatologists had been inconclusive. He used a computer keyboard fairly often and had a habit of manipulating his fingers. No abnormal psychiatric history was noted.

All x-rays, MRI, and blood test results were normal. Although a diagnosis of PDD was under consideration, a biopsy was performed to rule out a more serious underlying systemic disorder. Histological analysis of the specimen revealed increased production in the dermis of short thick collagen fibers and mucin. Immunohistochemical staining positively identified early cell differentiation in endothelial and vascular-associated tissues. However, these pathology findings were not specific to any disease course and only showed that the tissues were fairly new.

We started the patient on a simple therapeutic trial, consisting of application of corticosteroid-impregnated tape to a single finger of his choice for 12 hours every evening. At the same time, he was asked to avoid manipulating all affected fingers. One month later, the swelling in all of his fingers had decreased considerably. No finger seemed better than the others. The taping did not produce superior results.

The cause of PDD is uncertain, but a common trait among affected patients is frequent mechanical manipulation of the fingers such as joint rubbing or knuckle cracking, either due to occupation or habituation. The suggested treatment options range from steroid injection, to anti-allergen administration, to applying steroid-impregnated tape. However, given that our patient improved by simply ceasing to manipulate his hands, we agree with other physicians that behavior modification is sufficient. Modifying the aggravating behavior and, when appropriate, seeking psychological counseling is the recommended treatment for patients with PDD. Recognizing the clinical presentation of the disease will save patients time, stress, and unnecessary treatment modalities not indicated for their condition.

Schneider K Rancy, Scott W Wolfe
Division of Hand and Upper Extremity Surgery, Hospital for Special Surgery, New York, NY, USA

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