Ectopic Dupuytren’s Disease: A Commentary

Ghazi Rayan, MD

From OU Medical Center and Integris Baptist Medical Center, Oklahoma City, OK.

McFarlane et al\(^1\) described the typical presentation of Dupuytren’s disease (DD) as occurring in a white man averaging 57 years of age with bilateral hand and multiple digital involvement. The disease progresses over time, leading to digital contracture but at variable rates. Hueston\(^2\) described DD patients as expressing diathesis, and those with strong diathesis consistently have a family history of DD, severe disease, and ectopic sites of fibromatosis such as the dorsal digital area (Garrod’s nodes), planter fascia (Ledderhose’s disease), and male genitals (Peyronie’s disease). Wrist involvement in DD is extremely rare, but if it transpires it is part of a spectrum and an extension of digital and palmar disease in the typical patient described by McFarlane et al\(^1\) and in the strong diathesis patient described by Hueston.\(^2\)

DD begins by the formation of a small digital or palmar nodule followed by cord development. The nodule may enlarge, but it is extremely unusual for it to exceed 2.5 cm in diameter. Compared with the cord the nodule is more cellular and contains abundant myofibroblasts. Myofibroblasts, however, are not exclusive to or a signature of DD. They are encountered in a myriad of normal, injured, and pathologic tissues including wound healing, various inflammatory arthritic disorders, and neoplastic conditions, whether benign or malignant tumors.

There are a variety of fibrotic soft-tissue tumors that share the same histologic characteristics of DD. A soft-tissue mass that shows “nodular proliferation of spindle-shaped fibroblasts surrounded by dense collagenous tissue” is by no means diagnostic of DD. This is particularly true in the absence of a family history, bilateral involvement, hand affliction, other ectopic manifestations, and the extension of an existing relentless disease from its customary location.

An accurate diagnosis is necessary to gain insight into the prognosis and treatment of this enigmatic disease. In fairness to the authors of the preceding case report it is reasonable to say that accurate diagnosis can be achieved best by gaining knowledge concerning the genetics of DD. Identifying a gene or genes that are linked to the disease will enable us to understand the etiology of this condition better and to substantiate the notion of ectopic DD in the absence of its other signs and symptoms. Until this is possible we have to rely on our collective wisdom and clinical insight. Therefore, in tribute and memory of Hueston\(^2\) and McFarlane et al,\(^1\) let us not brand fibrotic masses in the upper extremity as ectopic Dupuytren’s disease in the absence of its classic manifestations.