Dupuytren's disease

Sir,

Dupuytren's disease is a fibro-proliferative disorder of unknown etiology, often leading to disabling contractures of the hand. We report a case of Dupuytren's contracture with review of etio-pathology, clinical presentation and management.

A 34-year-old Asian manual laborer presented with progressively worsening contracture of seven years duration involving right ring finger, lately affecting his job. There was no history suggestive of trauma, burns and collagen vascular disease. There was no familial history, no history of diabetes, epilepsy, alcohol consumption and smoking. Physical examination revealed thickening of right palmar aponeurosis with contracture of the finger. There was no detectable joint swelling, tenderness or knuckle pads. Routine laboratory investigations showed no abnormality. The X-ray of the hand was suggestive of flexion deformity of the ring finger with well preserved joint space. In view of significant morbidity; surgical intervention with limited regional fasciectomy technique was done with satisfactory result [Figures 1-4].

Dupuytren's disease is a disorder of the connective tissue primarily affecting the palmar and digital fascia often leading to disabling contractures of the metacarpophalangeal (MCP) and proximal interphangeal (PIP) joints of the hand.

The condition was first introduced in 1777 by Henry Cline into the medical literature, however, Dupuytren's contracture is named after a French anatomist and

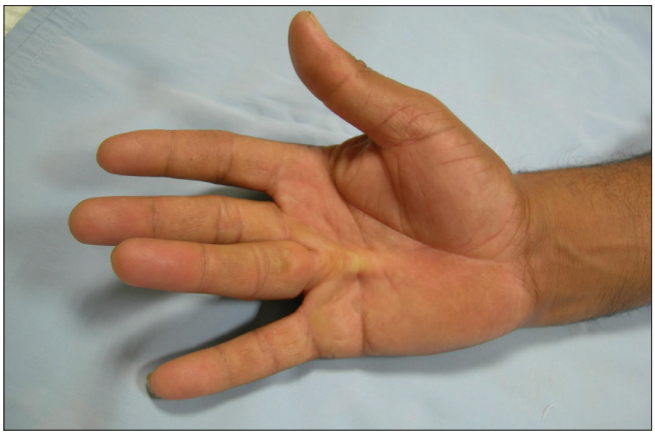

Figure 1: Dupuytren's contracture involving right ring finger

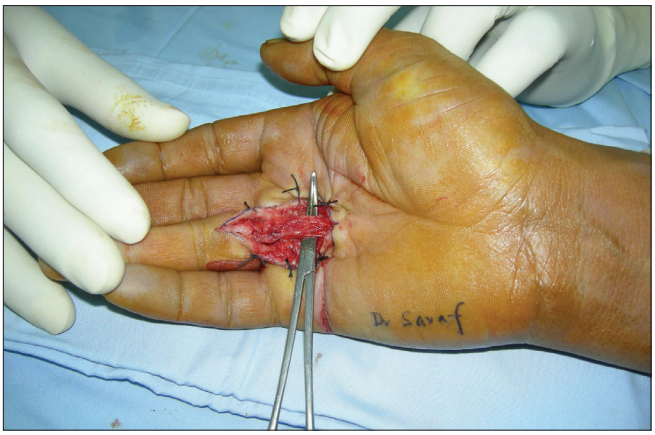

Figure 2: Intra-operative demonstration of Thickening and contracture of fibrous band

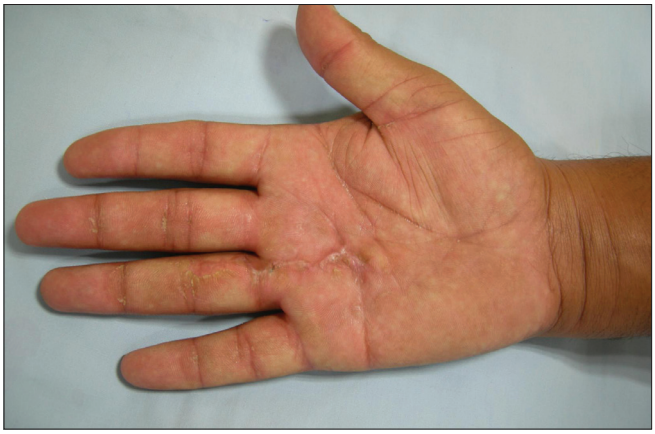

Figure 3: Postoperative complete extension

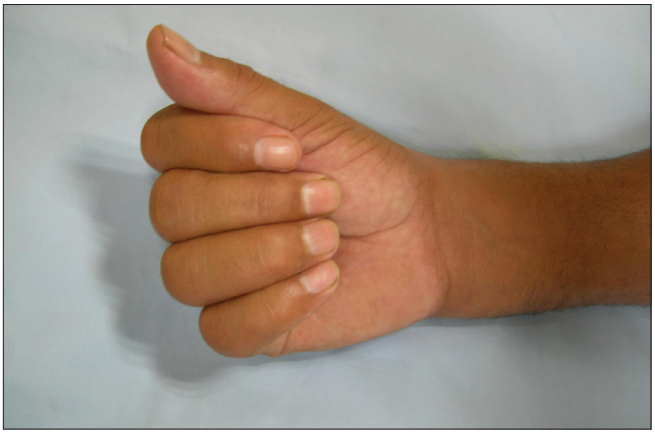

Figure 4: Postoperative complete flexion

surgeon Baron Guillaume Dupuytren who described the condition in detail in 1834.[1,2]

The etiology of Dupuytren's disease is still unknown. Genetics probably plays an important role and alcoholism, diabetes mellitus, epilepsy and HIV are considered to be significant risk factors for development of this disabling condition. There is a reported positive family history, which provides strong evidence for a genetic influence with many studies suggesting an autosomal dominant pattern of inheritance with variable penetrance.[3,4] Studies have shown that persons of any race can be affected.[5]

Dupuytren's disease usually has bilateral involvement, however, in unilateral cases, the right side is affected more often than the left. The ring finger is usually affected first, followed by the little finger, and then the middle finger. The disease is six to 10 times more frequent in males than in females.

The progression of the disease occurs in stages. Initially in proliferative stage, nodules develop in the palmar fascia which are known to be the pathognomonic lesion of Dupuytren's contracture and are composed of fibroblasts and type III collagen. With further progression of disease the nodules become a thickened fibrous cord underneath the skin which later thickens and shortens in the active contractile, or involutional stage leading to the development of contractures.[6] The basic pathophysiology of Dupuytren's contracture remains myofibroblast proliferation with increased water levels, type III collagen, chondroitin sulfate, reducible cross-linkages fibroblast proliferation and collagen deposition. The exact cause of uncontrolled proliferation remains unknown, however, many studies have implicated increased expression of growth factors, including basic fibroblast growth factor, platelet-derived growth factor, and transforming growth factor-beta for these changes.[7,8]

The management of Dupuytren's remains challenging and needs to be individualized. Various medical and surgical modalities have been described depending upon the severity of the symptoms. Usually, asymptomatic cases are better managed with conservative management, while symptomatic disabling cases are indications for surgery.

The non-surgical approach includes splinting, skeletal traction, steroids, calcium channel blockers, dimethylsulfoxide, vitamin E, allopurinol, ultrasound, laser therapy, radiation, Vitamin E and collagenolytic fasciotomy. However, they have not been proven to be of long-term value unless combined with surgical treatment.[9] Steroids do not stop progression of the disease but are often effective in reducing symptoms. A study showed that injections with triamcinolone acetonide (Kenalog), monthly, for up to five months, or every six weeks for up to three injections, followed by a six-month respite lead to significant disease regression.[10]

The role of IFN (interferon) alpha and gamma is also being evaluated and some preliminary studies have found encouraging results.[11,12] Needle fasciotomy is a quick minimally invasive procedure with no down time. However the chances of recurrence are high and are not effective in treating severe Dupuytren's contracture.

Surgically, excision or incision of the diseased fascia remains the cornerstone of the treatment. The presence of palmar nodule and cord in early stage are not necessarily indications for surgery. A MCP joint contracture and PIP joint contracture of 30 degrees or more is considered to be an indication for surgery.[13] Fasciectomy is the most preferred procedure. The various types of fasciectomy operations described are open fasciectomy (radical), dermofasciectomy and regional fasciectomy.[14] Regional (Selective/ Limited) fasciectomy involves excising only grossly involved fascia and is one of the most commonly employed surgical technique with low morbidity and low recurrence rates. However, none of the treatment modalities cure the disease. They only help in preventing further progression of the disease. In near future, further medical advances in genetic engineering might help in stopping or reversing the progression of the disease at cellular level and the ongoing research will form the foundation for future clinical interventions.

Sanjay Saraf

Department of Plastic Surgery, NMC Specialty Hospital, Dubai, UAE

Address for correspondence: Dr. Sanjay Saraf,
Department of Plastic Surgery, NMC Specialty Hospital,
Dubai, U.A.E.
E-mail: drsaraf@hotmail.com

DOI: 10.4103/0378-6323.62985 -

REFERENCES

1. Dupuytren BG. Permanent retraction of the fingers, produced by an affection of the palmar fascia. Lancet 1834;2:222-5.
2. Townley WA, Baker R, Sheppard N, Grobbelaar AO. Dupuytren's contracture unfolded. BMJ 2006;332:397-400.
3. Saar JD, Grothaus PC. Dupuytren's disease: an overview. Plast Reconstr Surg 2000;106:125-34.
4. Burge P. Genetics of Dupuytren's disease. Hand Clin 1999;15: 63-71.
5. Gonzalez MH, Sobeski J, Grindel S, Chunprapaph B, Weinzweig N. Dupuytren's disease in African-Americans. J Hand Surg Br 1998;23:306-7.
6. Seyhan H, Kopp J, Schultze-Mosgau S, Horch RE. Increased metabolic activity of fibroblasts derived from cords compared with nodule fibroblasts sampling from patients with Dupuytren's contracture. Plast Reconstr Surg 2006;117:1248-52.
7. Tomasek JJ, Vaughan MB, Haaksma CJ. Cellular structure and biology of Dupuytren's disease. Hand Clin 1999;15:21-34.
8. James WD, Odom RB. The role of the myofibroblast in Dupuytren's contracture. Arch Dermatol 1980;116:807-11.
9. Hurst LC, Badalamente MA. Nonoperative treatment of Dupuytren's disease. Hand Clin 1999;15:97-107.
10. Ketchum LD, Donahue TK. The injection of nodules of Dupuytren's disease with triamcinolone acetonide. J Hand Surg 2000;25:1157-62.
11. Sanders JL, Dodd C, Ghahary A, Scott PG, Tredget EE. The effect of interferon-alpha2b on an *in vitro* model Dupuytren's contracture. J Hand Surg Am 1999;3:578-85.
12. Nedelec B, Shen YJ, Ghahary A, Scott PG, Tredget EE. The effect of interferon alpha 2b on the expression of cytoskeletal proteins in an *in vitro* model of wound contraction. J Lab Clin Med 1995;126:474-84.
13. McFarlane RM. Dupuytren's disease. In: McCarthy JG May JW, Littler JW, eds. Plastic Surgery. Vol 8, 1st ed. Philadelphia: WB Saunders; 1990. p. 5053-8.
14. McGrouther DA: Dupuytren's contracture. In Green DP, Hotchkiss RN, Pederson WC, eds: Operative Hand Surgery, vol 1, 4th ed. New York: Churchill Livingstone; 1998. p. 563-91.