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Dr Millar E; Mr Knight R

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Dupuytren's Contracture - a Review of Pathology and Treatment

Dr Millar E; Mr Knight R

Institution

West Midlands Deanery,
Birmingham Hand Unit, Queen
Elizabeth Hospital

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Abstract

This review outlines the condition called Dupuytren's disease and aims to discuss the epidemiology, pathophysiology, clinical presentation and treatment options of the disorder. Dupuytren's disease is characterized by the presence of flexion contractures of the fingers and the hand. It belongs to a group of fibroproliferative disorders where the myofibroblasts of the subcutaneous palmar tissue proliferate in the form of nodules and cords and may result in progressive and flexion contractures of the finger joints. Most cases are idiopathic in nature but the trait can be inherited and there are recognised risk factors.^{1,2}

Whilst there have not been many advances in the understanding of the aetiology of this condition in recent years, there has been some progress in treatment options with the development of injectable collagenases as well as more traditional surgical techniques. However it should be recognised that at present there is no cure for Dupuytren's disease. Treatment consists of managing patients' functional impairments that result from the disease.

Key Words

Dupuytren's; Hand; Collagenase; Anatomy; Surgery.

Corresponding Author:

Dr Millar E: E-mail: e.millar1@nhs.net

Introduction

This review outlines the condition called Dupuytren's contracture and aims to discuss the epidemiology, pathophysiology, the clinical presentation and treatment options.

Dupuytren's disease is characterized by the presence of flexion contractures of the fingers and the hand. It belongs to a group of fibroproliferative disorders where the myofibroblasts of the subcutaneous palmar tissue proliferate in the form of nodules and cords and may result in progressive and flexion contractures of the finger joints. Most cases are idiopathic in nature but the trait can be inherited and some studies suggest trauma and certain types of manual labour may be contributing factors^{1,2}.

Whilst there have been not many advances in the understanding of the aetiology of this condition in the recent years, there has been some progress in the treatment options with the development of injectable collagenase.

Presentation

Dupuytren's contracture is a common condition in the western world and prevalence ranges from 3-5%

worldwide. It occurs more frequently in men than women and commonly in adults aged between 40-60 yrs. Men typically present earlier than females and have more severe disease³. When compare to the general population siblings of affected patients are three times more likely to have the disease⁴.

History Key Points:

- Handedness/Occupation
- Family History
- Ledderhose's/Peyronie's Diseases
- Diabetes Mellitus
- Epilepsy
- Alcohol Excess
- Smoking
- Functional Limitations

Although it is most prevalent in Scandinavian and Celtic populations a small number have been recorded in Afro-Caribbean and Asian populations³.

Patients may present at different stages and most commonly this is a disease which progresses over the course of years at differing rates. Occasionally a

finger may be come markedly flexed over the course of weeks-months. This more severe form of Dupuytren's is often referred to as Dupuytren's diathesis I and has a higher association with patients of a younger age at onset, diabetes mellitus, epilepsy, smoking and alcoholism⁵.

There is an association with the other fibroproliferative disorders such as Ledderhose's disease where similar nodules can be seen on the soles of the feet and Peyronie's disease which is fibrosis of the corpus cavernosum of the penis⁶. Patients with these associations are more prone to progressive and recurrent disease.

Pathogenesis

The disease process of Dupuytren's essentially involves fibroblast proliferation and collagen deposition and can be classified in three stages⁷.

Stage 1 – proliferative stage. This is in the initial phase of the disease where the nodule develops secondary to fibroblast proliferation. It can be painful and some patients report an itching sensation. The digital nerve can be or enveloped by the nodule. It is at this stage that the collagen fibrils are arranged randomly⁸.

Stage 2- involutonal stage. This is where the cord develops as the disease progresses along the fascia and into the fingers. Myofibroblasts are most active

during this stage and aggregate towards the collagen fibrils which are now aligned along lines of tension.

Stage 3- residual stage. This is where the cord continues to develop and thicken forming a contracture. The myofibroblasts are inactive and leave behind acellular tissue with thick strands of collagen. Type I collagen is surrounded by type III collagen^{9,10,11}.

There is an increase in the amount of type I and type III collagen in Dupuytren's.

Anatomy

When discussing Dupuytren's disease it is important to have a basic understanding of the underling anatomy. It is important to understand the terminology used to describe different structures. . Bands make up the normal anatomy and the cords formed are as a result of a pathological disease process. In the hand the palmar aponeurosis is the central portion of the deep palmar fascia and it runs longitudinally towards the base of the fingers. It crosses over the superficial transverse palmar ligament and splits into pretendinous bands. Running along each finger are the lateral digital sheath and the Grayson and Cleland's ligaments. In Dupuytren's disease infiltration of these bands with fibroblasts form form cords. The cords can displace or envelop the neurovascular bundle. Pretendinous cords result in MCPJ deformity, central cords and lateral cords

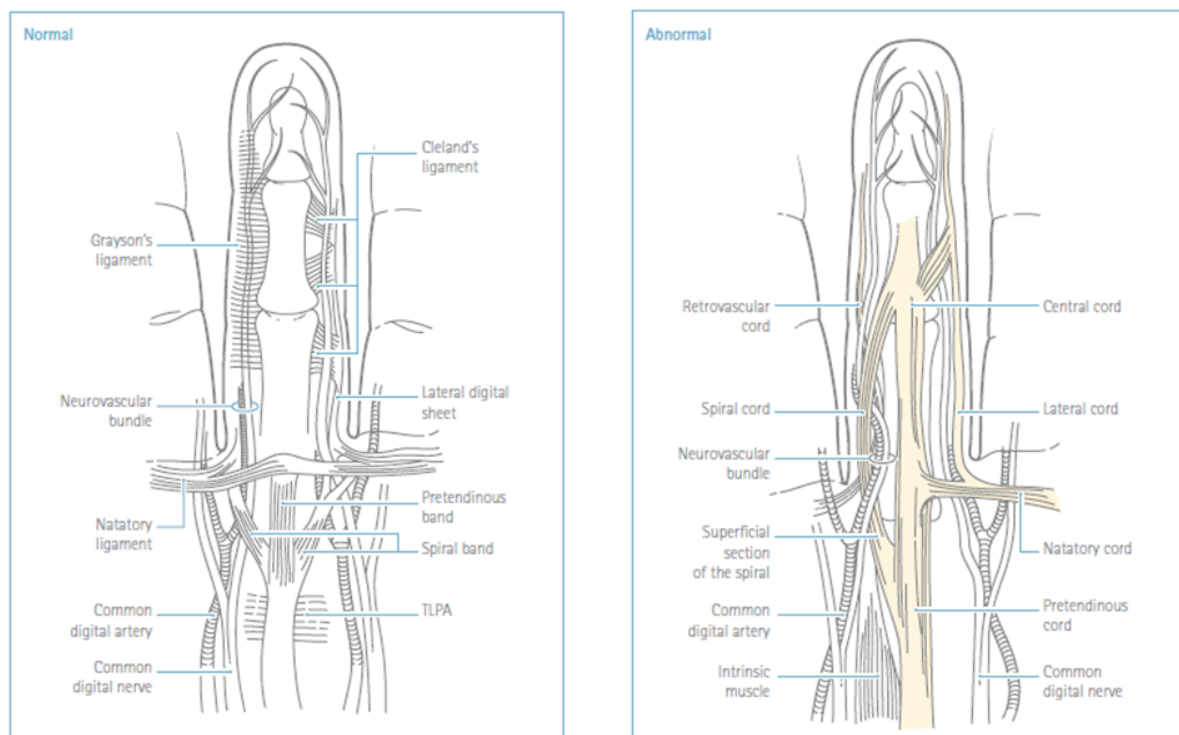


Figure 1: Anatomy of pathological cords (Courtesy of Swedish Orphan Biovitrum AB)

result in PIPJ deformity, Natatory cords limit digital abduction and spiral cords can displace the neurovascular bundle superficially resulting in an increased risk of damage in surgery.

Examining the patient

History taking from the patient includes basic questions such as age, handedness, occupation and hobbies and questions about the onset, progression and disabilities from the disease. It is important to note that despite severe contracture many patients are able to manage their activities of daily living well due to the fact that the radial side of the hand is less affected and the ulnar side grip is preserved. The common complaints are about difficulty in washing the face or shaking hands. History also should include questions about association's as such plantar fibromatosis and penile involvement as well as family history. As many of the patients are elderly it is important to ask about their medical history, drug history, allergies and social circumstances.

Examination of the upper limb starts with the shoulder as many patients with Dupuytren's also have a frozen shoulder and it is difficult to perform a Dupuytren's correction without abducting the shoulder during the operation. When examining the hand, the presence of contractures are noted. It is important to record the degree of contractures using a goniometer. Ring and little finger are commonly affected. Cords can be palpated in the palm and fingers. It is important to palpate for nodules which may or may not be tender. It is also important to look for common skin changes such as blanching of the palmar skin on finger extension, skin puckering, Garrod's pads (which is the thickening of the dorsal knuckle pads), thinning of the overlying subcutaneous fat and even pitting. Heuston table top test can be performed whereby a positive result indicated the patient cannot lay their hand flat on the surface. Dupuytren's can be unilateral and more commonly bilateral but is rarely symmetrical.

Classification

Tubiana and Michon's classification system can be used when describing Dupuytren's contractures. It is based upon the total of the angles of contractures at the MCPJ and the PIPJ¹². Stage N refers to the presence of nodules alone. Stage 1 is a total flexion deformity of 0-45°, Stage 2- 45-90°, Stage 3- 90-135° and stage 4- 135-180°. PIPJ contractures alone of more than 60° have been shown to be an independent risk factor for disease recurrence following surgery¹³.

Investigations

Dupuytren's is a clinical diagnosis and rarely needs further investigations.

Management

No treatment can fully eradicate Dupuytren's disease, and there are varying rates of recurrence depending on the treatment offered. Patients should be made fully aware of this.

Traditionally surgical treatments were used ranging from simple fasciotomy through to total or radical dermofasciectomy¹⁴. Each surgical procedure carries its own variations, risks and complications.

Percutaneous needle fasciotomy can be a cost effective procedure and a procedure of choice particularly for the elderly patient with multiple co morbidities. It can be done under local anaesthetic and therefore as an outpatient. However the rate of recurrence following needle fasciotomy has been reported to be as high as 90% in patients at three to five years¹⁵. As neurovascular complication rates are higher when used in the fingers it is preferred that needle fasciotomy is used in the palm.

In more severe cases of Dupuytren's contracture especially when a more definitive release is required then a limited fasciectomy may be performed. Fasciectomies involve removing the thickened cord and vary from partial, segmental or dermofasciectomy. When compared to percutaneous needle fasciotomy literature suggests fasciectomy is a more effective long term treatment and has lower recurrence rates of around 20-25%¹⁶. However as it is more invasive it has an increased rate of complications such as haematoma, neurovascular damage and infection.

Treatment Options:

- Observation/Monitor progression
- Needle Fasciotomy
- Collagenase Treatments
- Fasciotomy
- Fasciectomy
- Dermofasciectomy
- Amputation

Amputation is often reserved for the severest of cases and is most commonly used in the presence of recurrent disease of the little finger. Complications include neuropathic pain, reduced functionality particularly grip strength and psychological issues.

The mainstay of treatment for patients with Dupuytren's is largely surgical intervention as outlined above. Recently however management has changed and developments in non-surgical techniques mean they are increasingly becoming a more successful option.

In light of current literature including the CORD I and CORD II studies the use of collagenase clostridium histolyticum (CCH) is one of the most common and arguably more successful non-surgical methods alongside that of radiation therapy. It is important to bear in mind that will all non-surgical methods a better outcome is achieved when used prior to advanced disease progression.

Prior to the release of collagenase clostridium histolyticum (CCH) a wide variety of enzyme combinations were used in injections. Containing mixtures of anti-inflammatory and proteolytic enzymes these would aim to disorder the formation and deposition of collagen with varying results¹⁷. CCH however contains two specific enzymes that work in a complimentary and synergistic way to specifically target the collagen present in the Dupuytren's cords, type I and III. The two distinctive enzymes are AUX-I and AUX-II. AUX I works by targeting the terminal ends of the collagen chain whilst AUX II targets the interior segment¹⁸. This specific and localised therapeutic action therefore does not require systemic use in order to be effective. It is also important to note that type IV collagen, which is the primary supporting structure of arteries, veins and nerves, is resistant to the action of CCH and as a result such structures are protected.

The result of the CORD I study showed that CCH had reduced the degree of contracture to that of five degrees or less in 64% of patients compared to 6.8% of placebo controls when measured approximately 30 days after the last injection. This improvement was also demonstrated in the CORD II study (44.4% vs 4.8% of placebo controls)¹⁹.

A limitation of the CORD I and CORD II studies is that CCH use on contractures of the thumb were excluded. CCH also has its own side effect profile including pain, swelling, lymphadenopathy, itching, ecchymosis and more significantly flexor tendon rupture. Flexor tendon rupture is most likely to

occur in the little finger and therefore current guidance recommends that injections cannot be placed >4mm distal to the proximal digital flexion crease. Patients also need to be warned about the possibility of disease recurrence²⁰.

As this is a fairly new treatment method the long term side effect profile is yet to be defined. Radiation therapy is another non-surgical method used with NICE approval and guidance²¹. It aims to prevent progression of disease by disabling the development and growth of fibroblasts and is therefore recommended for use earlier in the disease process at the time of nodule or early cord formation. In one study it was found the symptoms of Dupuytren's contracture had improved in >50% of patients. Patients opting for this treatment however face the possibility of thinning of the skin, ulceration and long term risk of developing malignant skin lesions.

Patients with painless, minimal deformity and slowly progressive disease may benefit from observation and physiotherapy and occupational therapy input. Patients with large or painful nodules may benefit from corticosteroid injections. However these carry their own side effect profile and give an increased risk of skin discoloration, fat atrophy and possible tendon rupture.

Conclusion

In summary Dupuytren's contracture is common. It is a benign fibroproliferative genetic disorder caused by abnormal collagen deposition and formation. No curative management exists and disease recurrence is noted with all management options. Treatment is based on a symptom management approach. In light of the recent developments of non-surgical interventions treatment may be offered at an earlier stage and the need for surgical interventions and their associated complications may be reduced. However it appears to be that there will always be a place for surgical management particularly when treating severe disease.

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