Guillaume Dupuytren and finger contractures

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Case report

A 79-year-old man visited his family physician with contracted fingers on both hands. He had undergone operations for these in his early forties, and again 10 years later. The contracture recurred soon after the first operation (limited fasciectomy), and was this time accompanied by numbness and stiffness in the fingers. The little finger on the left hand was amputated during the second operation (figure 1). Disease recurrence was less severe on the right hand. On examination, he had an amputation scar at the metacarpophalangeal (MCP) joint on the little finger on the left hand. The ring finger was turned into the palm with 70–80° flexion contracture at the MCP and proximal interphalangeal (PIP) joints. The third finger on the left hand had 30° and 45° contractures at the MCP and PIP joints, respectively. There were 45° and 70° flexion deformities of the MCP and PIP joints of the little finger of the right hand. There was also a contracture affecting the thumb with a visible and palpable subcutaneous fibrous cord. Operation scars were seen in the palms of both hands. The patient had a single nodule on the sole of his right foot, but no knuckle pads; he did not have Peyronie’s disease. He was otherwise well and took no medication. He had never smoked, but used nasal snuff, and did not drink excessive amounts of alcohol. He had worked for most of his life as an electrician and in manufacturing, but had been a seaman on open boats in the North Atlantic ocean for 8 years in his twenties and thirties. At that time, his main task had been to hunt seabirds by catching them with his bare hands from snares on floating planks and his hands were frequently cold and numb. He was convinced that this activity was the cause of his condition. Nevertheless, there was a strong family history of the disease; his father was affected as were all of his seven siblings (figure 2). He declined further surgery and preferred to treat himself using a traditional remedy—an ointment made from shark liver oil, which he believed to be effective.

Clinical presentation

Dupuytren’s contracture is characterised by shortening of the palmar fascia, resulting in progressive digital flexion deformity. Early clinical signs of the disease are usually thickening of the skin and formation of fibrous nodules in the palm, just distal to the palmar crease. One or more longitudinal fibrous cords can form from the nodule to the finger, and these usually cross the MCP and PIP joints. The diseased part of the palmar aponeurosis starts to contract and the affected fingers bend gradually into the palm (figure 1). Permanent flexion deformity appears over several months or years. The ring finger and little finger are most commonly affected, but it is not unusual for others to be involved. The disease is usually painless. There could be similar fibrotic lesions on the dorsum of the proximal interphalangeal (PIP) joints,1 on the soles of the feet (Ledderhose’s disease), and in the penis (Peyronie’s disease). Dupuytren’s disease can be a serious handicap, causing loss of manual dexterity and the inability to touch, stroke, or shake hands in a normal manner.

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Figure 1: Clinical manifestation of Dupuytren’s disease
Hands with finger contractures. Note amputated little finger on the left hand.

Figure 2: A family affected by Dupuytren’s disease
Shading shows affected individuals. Eight siblings in one family are affected the younger generation has milder symptoms of the disease.
Epidemiology

Dupuytren’s disease is common in white people, especially of northwestern European origin, but is not very prevalent in other ethnic groups. The disease mostly affects men and is associated with advancing age. The highest prevalence has been reported in Scotland, Norway, and Iceland, where about 40% of elderly men are affected. The disease has been related to several medical conditions including diabetes mellitus, liver disease, hyperlipidaemia, and epilepsy. By contrast, rheumatic disorders are seen less frequently in Dupuytren’s disease. Cigarette smoking, excessive alcohol consumption, and manual labour have also been associated with the condition, but many of these associations have been questioned. Increased cancer incidence and mortality in patients with Dupuytren’s disease have been noted in three studies in Scandinavia and Iceland.

Disease cause and pathogenesis

The cause and pathogenesis of Dupuytren’s disease remains unclear. Histological examination of Dupuytren’s nodules shows an increased number of fibroblasts with excess formation of collagen. A high proportion of collagen type III has been reported in the nodules, instead of the usual collagen type I. A specialised cell type, the myofibroblast, has been described in the nodules and is thought to be the source of this altered collagen production. Myofibroblasts are probably transformed fibroblasts with phenotypical characteristics between those of fibroblasts and smooth muscle cells that are capable of producing α-smooth muscle actin isoform and several growth factors. The nodules have some histological features of neoplasia, including a high proportion of mitotic cells. Cells from fibromatous nodules have been reported to show chromosomal aberrations, including trisomies and unbalanced translocations, but the clinical significance is unclear. Some immunological abnormalities have been described in Dupuytren’s disease, suggesting that the immune system has a role in disease pathogenesis. Androgen receptor expression has been noted in the palmar fascia tissue and cultured myofibroblasts, leading to the suggestion that androgen-responsiveness of affected tissue could explain the predominance of the disease in men.

Surgical treatment for Dupuytren’s contracture

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<tr>
<th>Procedure</th>
<th>Description</th>
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<tr>
<td>Fasciectomy</td>
<td>Removal of the affected fascia. The wound is sutured in zigzag manner, which allows it to lengthen. Segments of the wound can be left to heal by secondary intention. Often used as treatment of first choice</td>
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<tr>
<td>Dermofasciectomy</td>
<td>Dermatofasciectomy is used if overlying skin is involved and adherent. Preferred when the condition has recurred after previous surgery, and in patients aged less than 40 years. The removed skin is replaced by skin graft</td>
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<tr>
<td>Fasciotomy</td>
<td>The fibrous cords are divided through small incisions in the palm. This can be done under local anaesthesia, and is often reserved for patients unfit for more extensive surgery or general anaesthesia</td>
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<tr>
<td>Amputation</td>
<td>This is rarely indicated for patients who have not had previous surgery, but may be appropriate after repeated recurrence, or when earlier surgery has been complicated by nerve or vessel damage</td>
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Figure 3: Chromosomal abnormalities from cell cultures from diseased Dupuytren’s tissue

Trisomy 8, one of the most common chromosome aberrations in Dupuytren’s disease can be seen. The Y chromosome is missing in the cell line.

Figure 4: Study for the praying hands of an apostle, Albrecht Dürer (1508)

Dupuytren’s disease might be familial, but the genetic mechanism is still unclear. Few formal studies of inheritance have been undertaken, but an autosomal dominant cause with variable penetrance has been suggested. However, the inheritance might be polygenic, or there could be an interaction between environmental and genetic factors.

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History
The oldest description of permanent finger contractures is probably in the Icelandic sagas, written around 1200, in which bishops miraculously cured finger contractures—possibly by forced fasciotomy. The disease was also well known in Scotland as illustrated by the legendary tale about the curse of the MacCrimmons. The MacCrimmon clan lived in the Western Isles of Scotland. The family ran a bagpipe college that was famous all over Scotland. However, members of the clan frequently developed finger contractures, with the result that they were unable to play the instrument. This became known as the MacCrimmon curse. The model for the famous drawing by the 15th century artist, Albrecht Dürer, might also have been affected by the disease (figure 4).

The oldest reference to Dupuytren’s contracture in the medical literature is believed to be that by Felix Plater in Basel, who described contractures of the ring and little fingers of a master mason in 1614. Henry Cline (1750–1827) described the disorder in England and proposed treatment by palmar fasciotomy as early as 1777. Cline’s work was followed by one of his students, Sir Astley Cooper (1768–1841), who described the condition in lectures he delivered in London. In France, Alexis Boyer (1757–1833) wrote about “crispatura tendinum”—the term used for finger contractures in the early nineteenth century. It was one of Boyer’s students who referred a patient with finger contracture to the surgeon, Guillaume Dupuytren. Dupuytren operated on the patient and reported the case in a lecture given on Dec 5, 1831. The lecture was recorded word for word by his assistants and published in France, before being translated into English and published in The Lancet in 1834 (figure 5).10 Dupuytren emphasised the association between the disease and manual labour—“The greater number of individuals affected by this disease have been obliged to make effort with the palm of the hand or frequently to handle hard bodies”—but his main contribution was to describe the exact anatomical features of the disorder and to treat it successfully by fasciotomy. Around this time, other surgical advances that led to a substantial reduction in postoperative complications and mortality were described including infection control, the use of tourniquets and, in 1842, the use of ether anaesthesia by Crawford Williamson Long in Georgia, USA. In England, William Fergusson (1808–77) was the first to open the whole length of a finger and perform a complete fasciotomy. The standard treatment for Dupuytren’s disease has not changed much since the middle of the nineteenth century.11

Baron Guillaume Dupuytren (1777–1835)
Guillaume Dupuytren (figure 6) was born in Pierre-Buffière, a village near Limoges, France. There had been many surgeons in his family, but he was the son of an advocate. Dupuytren studied at a Jesuit college in Paris through the years of the revolution and the bloody Reign of Terror.12 In 1792, the National Assembly abolished all universities in France, including medical schools. The immediate effects were disastrous, since the French army was at war and became short of surgeons. Medical schools reopened 2 years later. This upheaval did, however, pave the way for a new generation of scientists, and restructuring and modernisation of the medical courses in Paris. Guillaume Dupuytren began his medical studies in 1795, and his university years were far from easy. He almost starved, and had to study in bed because of lack of fuel to heat his room—there are even reports that he used fat from cadavers as oil for his light.13 After receiving his doctorate in 1803, Dupuytren worked at the Hôtel Dieu, the largest and most important hospital in Paris at that time. His appointment there was marred by a bitter conflict with the chief surgeon, Phillippe-Joseph Pellatan, which was to last almost 10 years, until Dupuytren replaced him in 1815.

The next two decades became known as the age of Dupuytren, when he ran the Hôtel Dieu with an iron hand. He was gentle with his patients, but was demanding and authoritarian with his students, who were only allowed to speak when spoken to. The daily routine began at 0600 h with a 3 h ward round, on which he allowed to speak when spoken to. After receiving his doctorate in 1803, Dupuytren worked at the Hôtel Dieu, the largest and most important hospital in Paris at that time. His appointment there was marred by a bitter conflict with the chief surgeon, Phillippe-Joseph Pellatan, which was to last almost 10 years, until Dupuytren replaced him in 1815.

The treatment of Dupuytren’s disease other than surgical correction; steroids, interferons, and radiotherapy are not effective. The primary aim of surgery is to restore hand function if daily activities are impaired, but complications can occur and recurrence or extension of the disease is not uncommon.1 The panel shows the main surgical procedures available. Percutaneous needle fasciotomy, with injection of collagenase, has shown promise as a treatment.10
He returned to the hospital between 6 and 7 o'clock in the evening to see those he had operated on earlier. Dupuytren was an outstanding clinician, and was acknowledged as one of the founders of surgery in France.

Dupuytren made many contributions to the field of medicine. He showed in animal experiments that the spleen could safely be removed. He described the association between right-sided iliac abscess and perforation of the appendix, and showed that chronic enlargement of the testis could be caused by syphilitic gumma and could be treated with mercury tablets (which became known as Dupuytren's pill). He was first to classify burns in stages, and he also described post-traumatic shock, and congenital dislocation of the hip. He received many public honours and was appointed professor of operative medicine at the Faculty of Medicine of Paris in 1812, and was president of the Academy of Medicine in 1824. Louis XVIII and Charles X chose Dupuytren as their surgeon and he was conferred the hereditary title of Baron in 1823. However, his personal relationships were not good. Many of his colleagues regarded him as arrogant, and he was once described as “the greatest of surgeons and the least of men”.

In 1833, Dupuytren's health began to fail, and he had a stroke while giving a lecture. He felt his cheek drooping and could not close his right eye properly. As a clinician, he diagnosed his own stroke but continued lecturing, holding his hand to the cheek to prevent his students from noticing what had happened. 2 years later, he developed empyema, and after considering surgery, decided that death was his destiny. Despite his colleagues' attitude towards him, he was held in great respect by ordinary people, and hundreds of working men in Paris attended his funeral.

References

Figure 6: Guillaume Dupuytren (1777–1835)