DUPUYTREN'S DIATHESIS

A Case Report

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Since Baron Dupuytren's famous account in 1832 of the characteristic appearance of the hand associated with thickening and contracture of the palmar aponeurosis the condition has been known by his name. Dupuytren himself mentioned similar changes in the sole of the foot, and in 1904 Garrod described firm plaques of tissue adherent to the skin overlying the dorsum of the proximal interphalangeal joints of the fingers in association with Dupuytren's contracture of the palm. It was not until 1941, however, that Lund drew attention to the association of the palmar changes not only with knuckle pads and nodules in the sole, but also with stiffness of the shoulder joint and with Peyronie's disease in epileptics treated with barbiturates. This association has since been confirmed (Skoog 1948, Early 1962, Hueston 1963) and has led to the idea of a diathesis or constitutional predisposition to the production of Dupuytren's tissue (Hueston 1963).

Although each lesion has been recorded in association with Dupuytren's contracture on several occasions, there appears to be no record of all the lesions occurring simultaneously in the same individual. The female epileptic patient discussed below developed several of the lesions, in particular large fibrous nodules in both soles and prominent knuckle pads.

CASE REPORT

Early in 1959 a married woman aged twenty-seven, who had suffered from idiopathic epilepsy since the age of fourteen, was referred to the Royal National Orthopaedic Hospital with painful swellings in the soles of both feet. In 1956 small lumps had appeared, first in the right foot and then in the left, and these had gradually increased in size and number until they produced discomfort on walking. The lumps were situated towards the medial side of the soles and were attached deeply, but only on the right side was there attachment to the skin (Fig. 1).

In 1958 the first knuckle pad appeared in the right ring finger, and within a few months similar swellings had developed over the proximal interphalangeal joints of all the fingers (Fig. 2) and had become painful. Small firm nodules were discovered in the palm of each hand at the base of each little finger and of the ring finger on the right, but there was no suggestion of contracture or puckering of the skin.

The upper gums had a granular, pitted appearance and were hypertrophied, especially around the left upper lateral incisor and canine teeth (Fig. 3).

Operations—In December 1959 the greatly thickened plantar fascia bearing the painful nodules was completely excised from the right foot (Figs. 4 and 5) and six months later a similar operation was carried out on the left foot. Histological examination of the specimens revealed the characteristic picture of “Dupuytren's tissue” (Figs. 6 and 7). Convalescence after each operation was complicated by the development of a wound haematoma and deep vein thrombosis. The right foot became painless, but the left, although considerably improved, remained uncomfortable when weight bearing long after discharge from hospital.
Progress and further treatment—In August 1960 the left shoulder became painful, with limitation of abduction and rotation. There were no abnormal neurological signs in the limb, and radiographs of the cervical spine and shoulder were normal. Heat and exercises produced no immediate improvement, but gradually the symptoms disappeared.

The clinical picture was further complicated by an emergency admission to another hospital in November 1961 for an acute exacerbation of low back pain which had been troublesome on and off since a pregnancy nine years earlier. After conservative treatment had failed to relieve the symptoms, operation was resorted to and a moderate protrusion of the lumbo-sacral disc was curetted.

In the meantime the small nodules in the palm of the right hand had become more obvious, and a definite band had appeared in the palm at the base of the ring finger. Further nodules had appeared in the soles of both feet, and walking had become painful again despite surgical shoes with sponge rubber insoles. The situation was aggravated by an increase of 28 pounds (or 13 kilograms) in body weight in two years. Because of the disappointing results of previous surgery, it was decided to treat the recurrent nodules, and also the right hand, by radiotherapy. Treatment was begun for the palmar lesion, but the hand and fingers became painful and swollen, and after a total dose of 2,700 rads the treatment was stopped. The right shoulder also became stiff and painful, and in May 1962 further admission to hospital was arranged for physiotherapy.
The right foot at operation in December 1959 showing the widespread nodular thickening of the plantar fascia (Fig. 4), and the extent of the excision, with the excised fascia (Fig. 5).

Figure 6—Low power view of a section from the excised plantar fascia showing parts of three of the cellular nodules which are found scattered through the aponeurosis, with normal fascial tissue between. (× 26.)

Figure 7—High power view of the margin of a nodule showing the whorled arrangement of the fibroblasts, merging with the adjacent aponeurosis. (× 125.)
The feet were now so painful that walking was almost impossible, and the patient requested further operation. In July the left foot was again explored and the scar tissue which had replaced the plantar fascia and the associated nodules were excised. With intensive physiotherapy the right shoulder and hand improved, and the patient was discharged from hospital walking with crutches.

Recent condition—At the time of review (January 1964) both feet were still painful and sticks were used. There was a tender lump one centimetre across in the sole of the left foot and there were two smaller nodules on the right side. The scar on the right sole was thickened and tender, and both second toes showed a hammer deformity not present before (Fig. 8). In the hands there were nodules at the base of the middle, ring and little fingers of the right hand, as well as in the palm and the web of the thumb. Bands had developed in the palm in line with the middle and ring fingers. The knuckle pads had completely disappeared from the right hand after radiotherapy; those on the left side remained, and an additional pad was noted on the thumb. There were persistent pain and stiffness in the right shoulder, but the left shoulder was symptomless. The appearance of the gums was unchanged.

Investigations—Laboratory investigations were normal throughout. The haemoglobin was never below 80 per cent and the erythrocyte sedimentation rate did not exceed 12 millimetres in the first hour. Rose's test was negative, and plasma proteins and electrophoresis, serum uric acid, blood urea and cholesterol, the total white cell count and the differential count were all within normal limits.

DISCUSSION

The incidence of Dupuytren's contracture in the general population increases with age and is higher in men than in women. In young women the disease is rare (Early 1962). Among epileptics the incidence is not only increased but the condition develops much earlier in life and the sex difference is less apparent (Lund 1941, Skoog 1948, Early 1962, Hueston 1963).

Plantar fibromatosis undoubtedly occurs as a solitary finding, but the true incidence is difficult to assess because symptoms are usually absent. Lund found only one case in over a thousand brewery workers, but among 361 epileptics there were twenty-five with plantar
lesions. The association between the lesions in the palm and in the sole has repeatedly been confirmed and is unlikely to be fortuitous. The unusual features in this case were the widespread involvement of the plantar fascia (the lesions usually being limited to the medial side of the sole), the skin involvement and the deformity of the second toes.

Although excision is seldom necessary, recurrence after operation is not unusual. Pickren, Smith, Stevenson and Stout (1951) suggested that this was due to incomplete excision of the plantar fascia, but this explanation seems unlikely in this case, particularly in view of the recurrence after a second operation with free surgical exposure.

Involvement of the palmar fascia was inconspicuous here, but became more obvious with time. Whereas marked involvement of the foot is unusual without similar lesions in the palm, well developed knuckle pads often precede the development of Dupuytren's contracture (McInndoe and Beare 1958). It is noteworthy that the knuckle pads disappeared from the right hand after irradiation. The swelling of the right hand in itself is not without interest, for it is an unusual complication of radiotherapy for this condition. Radiotherapy may perhaps be contra-indicated in the presence of the widespread fibrous tissue abnormality which must have been present in this case, and this reaction suggests some degree of hypersensitivity of connective tissue.

A stiff, painful shoulder is a common complaint in the orthopaedic out-patient department, but not at the age of thirty. It may have been fortuitous, like the low back pain, or perhaps related to the immobilisation after radiotherapy. Nevertheless, among Lund's 361 epileptics there were twelve with "periarthrosis humeri," and Early found a similar association between Dupuytren's contracture and periarthrosis of the shoulder in 7 per cent of non-epileptic patients and in 10 per cent of those with epilepsy.

Hyperplasia of the gums is well recognised in epileptics treated with Epanutin and has been reported with methoin (Mesontoin), but it had not been seen with methoin by two experienced neurologists and a professor in dental surgery who were consulted (Hulbert 1959, Sandifer 1963, Prophet 1964). This patient had never had Epanutin, and had received methoin for only three years, between 1951 and 1954. A possible mechanical cause was slight malocclusion of the teeth (Prophet 1964), but the hyperplasia could also be a part of the generalised fibrous tissue abnormality associated with Dupuytren's contracture and one hitherto undescribed.

The histological appearances of plantar nodules, knuckle pads and palmar lesions are similar (Lund 1941, Skoog 1948 and Hueston 1963), and the typical features of Dupuytren's tissue can be seen in the sections from the plantar nodules removed from the left foot. These nodules consist of hyperplastic cellular foci, scattered irregularly through the aponeurosis, the fibre bundles of which seem to split to enclose them (Fig. 6). The cellular areas are made up of numerous well differentiated fibroblasts with oval nuclei, which become more elongated towards the periphery and mingle with increasing amounts of collagen; they merge imperceptibly with the adjoining aponeurosis (Fig. 7). This lack of a clear-cut margin in association with cellular proliferation and mitotic figures has occasionally led to the erroneous diagnosis of fibrosarcoma of the sole of the foot and to amputation (Pickren et al. 1951). The recurrent lesions in this case showed rather more scar tissue with few cellular areas but the typical appearance was still apparent.

The cause of Dupuytren's contracture remains unknown, but most authors now agree that trauma plays no part. Ling (1963) found that thirty-four of fifty patients had affected relatives and came to the conclusion that a single dominant gene was responsible for this inherited predisposition. In epileptics a familial association has been more difficult to trace; our patient had an uncle with palmar contractures. This case throws little light on the cause of the condition, but any theory must take into account not only the majority of cases with isolated lesions in the hands but also such cases as this one. It seems that there is an inherited predisposition or diathesis to the formation of "Dupuytren's tissue" (Hueston 1963).
SUMMARY

1. An epileptic patient with Dupuytren's contracture of the palms, severe plantar fibromatosis, prominent knuckle pads, periartitis of the shoulders and hyperplasia of the gums is described.
2. The plantar lesions recurred despite radical excision. The knuckle pads on one hand disappeared after radiotherapy.
3. The association of each of these conditions with epilepsy and with Dupuytren's contracture is reviewed, and it is suggested that the hyperplasia of the gums may be a hitherto unrecognised feature.
4. The characteristic histological appearances of the plantar lesions are described and the risk of confusion with fibrosarcoma is emphasised.

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REFERENCES

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