Dupuytren’s disease

Dupuytren’s contracture of the palmar fascia, better called Dupuytren’s disease, remains one of the big mysteries of surgery. In fact, in a recent collection of essays on this disease (Maladie du Dupuytren, edited by R. Tubiana and J. T. Hueston, l’Expansion, Paris, 1972), the famous hand surgeon, Marc Iselin, contributes a chapter on “les aspects mystérieux.”

In 1831, the French surgical genius, the Baron Dupuytren, made several incisions into the thickened contracted palmar aponeurosis of an elderly man and demonstrated that the disability of the ring and little fingers was due to a lesion of the aponeurosis and not the skin or tendons, thus earning himself a place in history, although others such as Astley Cooper had described the disease earlier. The lesion was at first thought to be due to local trauma, but this theory is no longer tenable. The lesions are only slightly commoner in manual workers and affect both hands in 40% of cases, while handedness has nothing to do with the choice of affected side. Moreover, when surgeons stopped looking at the hands alone and looked at the patients, they discovered that Dupuytren’s contracture was often accompanied by pads over the knuckles, where there is no aponeurosis, by nodules in the plantar fascia, and by induration of the penis known commonly as Peyronie’s disease.

It looks therefore as if the cause of the disease is generalized, and there is now good evidence that it is inherited by a dominant gene mechanism. For some unknown reason it affects only people of European stock, with a preference for Scots and Scandinavians, and there is a feeling around that the disease appears when some added insult is inflicted on an individual with the Dupuytren tendency. The insult may be epilepsy, alcohol abuse, liver disease or just trauma.

One of the fairly certain relationships is with epilepsy, also a genetically determined disease. Lund drew attention to this in 1941 and Skoog (Acta Chir Scand 96: Suppl. 139, 1948) noted that 42% of epileptics had Dupuytren’s disease also, a much higher proportion than would have been expected. Skoog thought that the lesions might be caused by the barbiturate medication given to the epileptics, but in an Edinburgh study James (Hand 1: 47, 1969) did not confirm this association.

Recently Pojer et al. (Arch Intern Med 129: 561, 1972) as a result of a study at Rochester, New York, have carried the story a little further. They note an incidence of 44 to 66% Dupuytren’s disease among alcoholics with liver disease, as well as relationships with epilepsy, pulmonary tuberculosis, and chronic invalidism, and think it likely that the disease represents a hyperplastic growth of connective tissue in attempts to repair some unknown lesion.

In spite of James’s findings, they do not exclude barbiturates as a cause, noting that in the pre-barbiturate era Féré and Françillon (Revue Méd 22: 539, 1902) claimed that it was very rare in epileptics. That barbiturates could be involved is also supported by the occasional finding of connective tissue disease (such as rheumatoid arthritis or systemic lupus erythematosus) in chronic barbiturate overuse.

The main link emphasized by Pojer and his colleagues is between Dupuytren’s disease and abnormal liver function. Their own investigations suggest that liver enzyme levels are often abnormal in Dupuytren’s disease. In their series of alcoholics, those with the disease tended to have high levels of glutamyl transpeptidase and evidence of chronic liver disease. In another series of epileptics, those with the disease had more elevated alkaline phosphatase (78% as against 45% among those without Dupuytren’s disease) and leucine aminopeptidase levels (25% and 4% respectively). In male epileptics the serum glutamic pyruvic transaminase and creatine phosphokinase levels also tended to be abnormal.

It may well be that epileptics on barbiturates have disorders of liver function in spite of the absence of clinical signs or of a raised serum bilirubin level, and that it is the liver disorder that is the common link with Dupuytren’s disease. The authors also raise the question whether muscles are also metabolically abnormal in the disease.

We have come a long way from the Parisian coachman (operated on by Dupuytren) whose contracture was believed to be due to his clutching his whip too tightly, and are now apparently dealing with yet another metabolic disease.