Dupuytren's disease and the sickle-cell trait in a female black patient

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Summary—Dupuytren's contracture is not often seen in the black-skinned patient. The combination of a Dupuytren's contracture in a black female patient who also carried the sickle-cell trait is described. This association of two diseases in a dark skinned patient has important surgical and anaesthetic implications which must not be forgotten in those countries with a large number of immigrants from the "sickle-cell" areas of the world.

Sickle-cell anaemia was originally an African hereditary disease brought to the Americas and West-Indies by the slave-traders, whereas Dupuytren's contracture is predominantly a "European" disease (Hueston, 1974). However due to increased immigration in the last 10-15 years many European doctors will have encountered patients with sickle-cell anaemia or the sickle-cell trait in their own countries (Fig. 1). In sickle-cell anaemia the globin part of the haemoglobin molecule is abnormal, a glutamic acid group being replaced by a valine group. True sickle-cell anaemia appears only in homozygotes: the erythrocytes contain a high percentage of sickle-cell haemoglobin (HbS), a relatively small percentage of foetal haemoglobin (HbF) and no normal adult haemoglobin (HbA). In the blood of the heterozygotic sickle cell trait carriers the erythrocytes contain a mixture of HbS (in a much lower concentration) and HbA. Under normal physiological conditions the sickle-cell trait carrier (HbAS) lives a normal life, in sharp contrast to the patient with the homozygous, symptomatic form (HbSS) of the disease.

The only beneficial effect of possession of the sickling gene is that it affords protection against plasmodium falciparum malaria (Gilles, 1971). This may explain the high incidence of this gene in some areas of Africa. However the sickle cell trait is not altogether harmless: under certain conditions, such as anaemia, hypoxia, hypothermia, hypotension and infection, sickling can occur with serious results (Davey, 1968). The clinical effects of sickling are well known to the tropical doctor but less readily recognised by his western counterpart.

In black patients Dupuytren's disease is extremely rare and very few cases have been reported (Larsen, 1966; Mennen and Grabe, 1979; Zaworski and Mann, 1979) and most of the patients are male. Ideas on the sex distribution of Dupuytren's disease have changed in the light of experience. Originally the disease was believed to occur almost exclusively in men (Klapp, 1923). Later a male to female ratio of 7:1 was reported (Wallace, 1965) and as high as 2.5:1 in a recent paper by Mikkelsen (1978) whose figures are amongst the most reliable as they were based on a population study.

In the past, operation statistics gave the wrong impression that the disease affected men much more frequently (Hueston, 1973, 1977). Nowadays Dupuytren's disease is believed to be
an autosomal dominant trait in which the penetrance is diminished in females (Matthews, 1979).

Case report
A 79-year-old black female patient, born in Surinam (Southern America) presented with a typical Dupuytren's contracture in the palm of her dominant right hand (Fig. 2).

The disease had been progressing slowly over the previous year and there was a slight, but annoying contracture of the little finger. She had also noticed a tiny band of scar in the thumb of her opposite hand. The patient who had been a washer-woman all her life, was still very active and had emigrated to The Netherlands 9 years previously.

There was no history of any other disease, pulmonary tuberculosis or epilepsy and she did not drink alcohol. As far as she knew there had been no inter-racial marriages in her family and no relations had any signs of Dupuytren's disease.

Laboratory investigations showed no evidence of anaemia: a positive sickling test, urobitin in the urine and a CPK reading of 72mU/ml (Normal 5-30mU/ml).

At operation a limited fasciectomy was carried out through a longitudinal zig-zag skin incision (Orlando et al., 1974). Two cords of thickened palmar fascia were removed.

Intravenous regional anaesthesia (Bier’s block) was used with the second wrap technique (Haas and Londeen, 1978). Oxygen was given by nasal catheter during the operation and for 24 hours post-operatively and the temperature in the operating theatre was kept a little higher than normal.

Discussion
Although sickle-cell anaemia is only one of approximately one hundred different kinds of haemoglobinopathies (Muntinghe, 1971) sickle-cell gene penetration is rising in our community as the result of immigration.

It is important to diagnose both the homozygous and heterozygous forms because there are implications in wound healing and hazards related to anaesthesia. Complications may also be encountered in patients with the relatively asymptomatic sickle-cell trait (DeHaan and Saunders, 1963).

In the USA it is estimated that 8% of the negro population are affected by the sickle-cell phenomenon, 97% of them in the heterozygous form (HbAS). In Surinam the incidence of the sickle-cell trait gene is surprisingly much higher and is estimated to be present in 15.5% of the negro population (Zuidema, 1972). In tropical Africa sickle-cell anaemia is mainly a disease of children who rarely reach adult age due to serious secondary complications such as malnutrition and infection. These serious sequelae are not usually seen in the Americas and Europe, though leg ulcers are common and the recurrence rate is high (Wolfort and Kriek, 1969).

Anoxia during general anaesthesia is a well recognised risk (Davey, 1968; Zuidema, 1972) but even local anaesthesia is dangerous and the use of a pneumatic tourniquet can be especially dangerous (Manson-Bahr, 1968; Davey, 1968; Allan, 1971). Under a tourniquet, circulation in a limb is reduced to less than 1% of that in the control limb (Klenerman, 1980) and a fall in oxygen tension to as low as 10 mm Hg is reached in approximately twenty minutes. This oxygen tension remains constant during the rest of the tourniquet time (Jorza et al., 1980) and can produce intravascular sickling in patients with sickle-cell anaemia and/or the sickle-cell trait.

The erythrocytes in the AS heterozygous patient show the Hbs tactoid crystalline form when the partial pressure of oxygen falls to 20 mm Hg but this is reversible if the hypoxia-time is short lived (Jonxis, 1977). In the homozygous state sickling will occur at an oxygen pressure of between 40 and 60 mm Hg (Manson-Bahr, 1968).

Sickling of the cells increases the blood viscosity in the capillaries and obstructs the small vessels to produce infarction, thrombosis and necrosis. These hazardous side effects can be avoided if the limbs are efficiently exsanguinated.
the operation is short and no hypoxic episode is produced by general or local anaesthesia.

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References

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