THE HAND-FOOT SYNDROME IN SICKLE-CELL
HAEMOGLOBINOPATHY

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The hand-foot syndrome is a benign self-limiting condition seen in young children with sickle-cell haemoglobinopathy, usually at the time of a crisis. The authors have observed 36 cases among 4920 patients. The features and management of the condition are discussed and the published literature is reviewed.

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Sickle-cell disease is a common type of haemoglobinopathy with a high prevalence around Nagpur in Central India. It is a multisystem disease of genetic origin. Skeletal manifestations are seen in a large number of patients, 10% to 20% of whom experience the hand-foot syndrome or sickle-cell dactylitis. The syndrome occurs in infants and children and is often misdiagnosed. It is more often seen in a specific community of scheduled castes and tribes.

PATIENTS AND METHODS

Between 1972 and 1991 a total of 4920 patients with sickle-cell disease were seen at the Sickle-cell Centre at Sushrut Hospital and the Indira Gandhi Medical College, Nagpur. Skeletal manifestations were found in 496 patients, 36 of whom had the hand-foot syndrome. All were admitted during a sickle-cell crisis. All were children under five years of age and 21 were aged between two and three years. There were 20 boys and 16 girls. In 14 patients this was the first presentation of the disease. All had experienced the acute onset of symmetrical swelling of the hands and feet (Fig. 1). Fever of varying degrees was always present. The children were debilitated, in poor general condition and at times dehydrated. The clinical features resolved after a mean of 21 days.

Severe anaemia and leucocytosis were always present with an average haemoglobin level of 6.2 g/100 ml. The test for sickling was carried out by the method of Daland and Castle (1948). A 2% solution of sodium metabisulphite was used as the reducing agent. The erythrocytes were observed after 15 minutes for an immediate reaction and after 24 hours for delayed sickling. The test was positive in all the patients. Electrophoresis showed an SS pattern in 6 patients (16.7%), an AS pattern in 22 (61.1%) and an ASF pattern in 8 (22.2%).

Fig. 1a
Appearance of the hands and feet in a child with the hand-foot syndrome.

Fig. 1b

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Radiological appearance. The radiological changes were seen on average ten days after the onset of symptoms. The earliest change was the appearance of subperiosteal new bone in one or more of the bones of the hands or feet (Fig. 2). There was cortical thinning, multiple irregular intramedullary deposits, areas of spotty destruction and formation of periosteal new bone, giving a 'moth-eaten' appearance (Fig. 3). The small bones of the hands and feet became rectangular in shape (Fig. 4).

Treatment. All the patients were managed conservatively. Drug treatment included analgesics, antibiotics, folic acid, zinc and sodium bicarbonate. Supportive splints were applied. Patients required correction of fluid imbalance and on rare occasions blood transfusion.

DISCUSSION

In 1941 Danford, Marr and Elsey reported the first case of dactylitis in a child with sickle-cell anaemia. Tori (1954) described destruction and a periosteal reaction in the metacarpals of both hands and Smith (1953) used the term 'hand-foot syndrome' to describe the acute swelling in the hands and feet of children with sickle-cell disease. Watson et al (1963) described the hand-foot syndrome as a common initial manifestation of sickle-cell disease, usually in children.

Since the early reports only a few studies have appeared in the literature (Macht and Roman 1948; Ivy and Howard 1953; Rowe and Haggard 1957; Victor and Imperiale 1957; Moseley 1959; Lambotte 1962; Porter and Thurman 1963; Diggs 1965; Worrall and Butera 1976). The syndrome has been reported to occur in 10% to 20% of patients with sickle-cell disease who have skeletal involvement (Macht and Roman 1948; Porter and Thurman 1963; Watson et al 1963; Worrall and Butera 1976). Most of the patients are under four years of age and the syndrome has not been reported after the age of seven years (Watson et al 1963).

The low incidence in our study may be because our patients were the only ones referred to the orthopaedic department. The age distribution of our patients is comparable with that in other studies.

The clinical symptoms are self-limiting and the duration
may vary from several days to a month (Danford et al 1941; Ivy and Howard 1953; Rowe and Haggard 1957; Watson et al 1963; Worrall and Butera 1976).

Skeletal changes in sickle-cell haemoglobinopathies occur mainly because of hyperplasia of the bone marrow and vascular insufficiency resulting in thrombosis and infarction. Hyperplasia of the erythrocytes increases the viscosity of the blood leading to stasis and thrombosis in the microcirculation and eventually to infarction and secondary infection.

The pathological process responsible for the hand-foot syndrome is not clear. Cockshott (1958) and Diggs (1965) thought that thrombosis due to sickle-cell disease led to obstruction of the microcirculation and cell death due to hypoxia in the metaphyseal area of bones. Ivy and Howard (1953) suggested that the increased density within the medullary area seen in sickle-cell disease was explained by marrow hyperplasia.

Weinberg and Currarino (1972) described the histopathological features observed at post-mortem in a child with sickle-cell dactylosis. These included extensive infarction of the marrow, medullary trabeculae and the inner layers of the cortical bone. This was associated with circumferential periosteal elevation and formation of subperiosteal new bone.

The radiological changes are completely reversible, but may take between six weeks and eight months to resolve (Danford et al 1941; Victor and Imperiale 1957; Cockshott 1958; Moseley 1959; Watson et al 1963).

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REFERENCES