EDITORIAL

Orthopaedic Manifestations in Sickle Cell Disease

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Sickle cell disease (SCD) is the most commonly inherited haematological disorder affecting man. It encompasses a group of genetic disorders characterized by presence of haemoglobin S (Hb-S) which results from substitution of valine for glutamic acid at codon 6 of the beta globin chain gene on chromosome 11. The disease is characterized by repeated vaso-occlusive crises (VOC) with complications affecting various systems with an early mortality as a result of pulmonary complications and sepsis. The musculo-skeletal system is commonly affected in SCD, manifesting itself as bone infarction, femoral head osteonecrosis, osteomyelitis and myonecrosis, myofibrosis and fasciitis.

Diffuse infarction affects long bones of the extremities, the metacarpals and phalanges[1] but spares the metaphyses due to extensive collateral circulation in the region of the growth plate. Dactylitis (hand-foot syndrome) appearing more often at 6-18 months; may be the first ischaemic bony manifestation. Bone infarcts may also affect the spine, the skull and facial bones[2]. Secondary infection of infarcted areas with Staphylococcus aureus and Salmonella is not uncommon.

Osteonecrosis of the femoral head is another important complication that causes considerable morbidity as a result of persistent pain and limitation of movements. It affects 3% of patients under the age of 15 years and bilateral disease occurs in more than 50% of cases[3]. The pathophysiology is believed to be due to sludging of sickled cells in the marrow sinusoids leading to bone marrow necrosis. Repair process may produce a rise in intramedullary pressure with subsequent resorption and collapse of the femoral head. The treatment is arthroplasty, but the prognosis is not as favourable[4] due to the presence of hard sclerotic bone making placement of the femoral component more difficult and due to loosening of the prostheses and high incidence of infection[5]. However, recent advances in surgical techniques has led to marked improvement in life expectancy of hip prostheses[3] but the chance of requiring revision within a year and 3 years is 8% and 20%, respectively[3].

Osteomyelitis is another major skeletal manifestation[6,7]. It mimics acute bone infarction but positive blood culture favour the diagnosis of osteomyelitis. Plain radiographs may reveal normal findings or merely soft tissue swelling in the early stages. Repeat radiographs after 10-15 days usually show bone destruction and periosteal reaction, which may also be seen after acute bone infarction. Technetium-gallium radioisotope scan is useful in distinguishing osteomyelitis from bone infarction[8], but routine use is not justified. *Salmonella* is the main causative organism of osteomyelitis in SCD in many world series and antibiotics specific to salmonella should be administered even before microbiological confirmation[6,7,9,11]. The recommended treatment is exchange transfusion, adequate hydration and oxygenation. Aspiration of infected bones and culture of bone aspirates is mandatory and prompt decompression of all bone abscesses must be performed. Parenteral antibiotics are administered and should be continued for 6-8 weeks.

There has been small number of reports regarding muscular complications of SCD. Myositis and fasciitis are observed during VOC in 4% of patient[12]. Pain and swelling of bilateral proximal groups of muscles of upper and lower limbs are common presenting complaints. Necrotizing myonecrosis and fasciitis may affect deltoids and thigh muscles symmetrically after prolonged sickling crises[12]. Myofibrosis has been noticed at the sites of repeated injections of various drugs such as pentazocine, morphine and insulin, and to lesser extent after penicillin and chloramphenicol in SCD patients as well as non-sicklers[12]. However, the myonecrosis and fibrosis reported by Valeriano-Marcet and Kerr were in the muscles, which were not routinely used for intramuscular therapy[13]. The condition gradually resolves within 2-4 weeks with the standard therapy for sickle crisis and application of warm soaks to the affected muscles for pain relief without the need for surgical intervention[13]. In some cases, surgical fasciotomy or incision and drainage may be needed to relieve areas of localized, tight swelling[12]. Chronic sequelae consists of muscular atrophy and contractures resulting in functional impairment.

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