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Surgical treatment of thoraco-lumbar fractures in sickle cell disease: a case report

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Abstract. – Sickle cell disease (or drepanocytosis) is a hemoglobinopathy characterized by an increase in viscosity and adhesivity of the typically sickle-shaped erythrocytes. The pathological osteo-articular involvement in the course of drepanocytosis is secondary to the avascular necrosis of the bone marrow, caused by vaso-occlusive episodes in the microcirculation during acute painful crises. Osteoporosis and extramedullary hematopoiesis are also consequences of the disease. The involvement of the spine is common, with clinical features ranging from simple changes in spinal morphology ("fish-mouth" appearance) up to vertebral bodies fractures with kyphotic deformity. In the presence of vertebral fracture, treatment options listed in the literature are conservative (rest, symptomatic therapy, orthosis), because of the high incidence of intra- and perioperative complications (acute respiratory syndrome, vaso-occlusive crisis…), in addition to the increased rate of implant failure. We report here a case of a young man affected by multiple pathological symptomatic vertebral fractures at the thoraco-lumbar junction, secondary to a well controlled but severe form of sickle cell disease. We decided to treat the patient surgically because of a worsening and potentially invalidating kyphosis deformity. We describe the surgical procedure and the management of a later complication consisting in the collapse of the osteoporotic vertebra below the instrumentation that required a surgical revision. Although a conservative approach is most frequently indicated, we believe that the surgical option should be considered when a clinical worsening occurs in a young patient with otherwise well-controlled disease.

Key Words:
Sickle cell disease, Vertebral fractures, Arthrodesis.

Introduction

Sickle cell disease or sickle cell anemia is an inherited autosomal recessive disorder, characterized by the presence of a pathological hemoglobin (HbS), which replaces the fetal hemoglobin to varying degrees from the age of 6 months. It is a structural hemoglobinopathy where a gene mutation of β-globin determines an increase in viscosity and adhesivity of the erythrocyte membrane, causing the typical "sickle" deformation of red blood cells, which tend to piling up on top of each other and are more rigid and brittle than normal[1].

The main effects are an hemolytic anemia due to premature splenic red cells destruction and the vaso-occlusive episodes in the microcirculation, favored by the reduced flexibility of the erythrocytes crossing the capillary bed. The consequent tissue ischemia is manifested by acute painful crises of varying duration, and over time leads to a progressive damage to organs. Spleen, skeleton, central nervous system, liver, kidneys and lungs are the most affected organs. The vaso-occlusive crises can occur each time there is a local tissue hypoxia, or acidosis, or dehydration; the factors precipitating the crises may be a viral infection, fever, vomiting and diarrhea, fatigue from exercise, exposure to cold, pregnancy, alcohol intake, up to emotional stress. The extreme fragility characterizing these patients is evident. The most common clinical manifestations of sickle cell disease are secondary to the osteo-articular involvement. The sites affected are different, and different pathogenetic mechanisms are involved: occlusion of the microcirculation, tissue ischemia and bone necrosis, osteoporosis due to bone marrow hyperplasia, extramedullary hematopoiesis. The involvement of the spine is common, with clinical features ranging from simple changes in spinal morphology ("fish-mouth" appearance) until to one or more vertebral bodies fractures with kyphotic deformity of the affected section[1-3].

The treatment is multidisciplinary and includes: symptomatic treatment of acute painful episodes and recurrent infections, osteoporosis medical therapy, periodic blood transfusions to reduce the amount of HbS, iron chelating agents; treatment of chronic renal failure, from hemodialysis until renal transplantation; splenecto-

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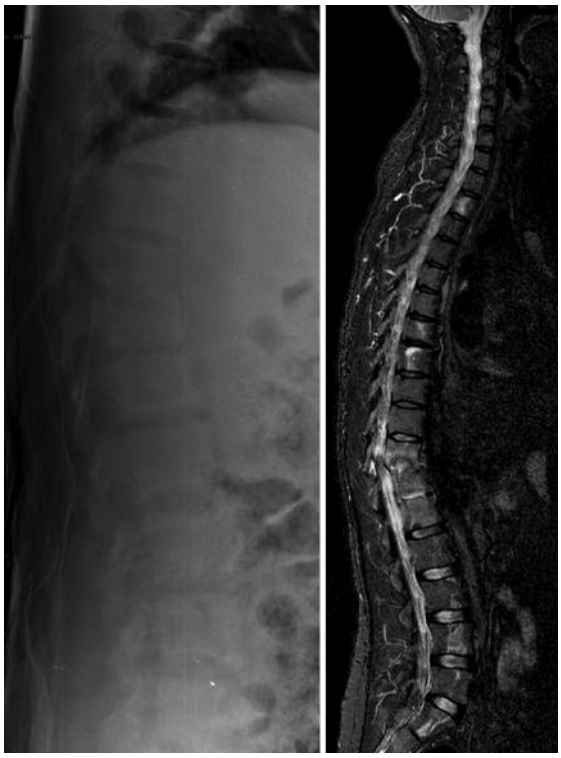

Figure 1. Twenty-one years old patient affected by drepanocytosis. Preoperative lateral X-ray and MRI imaging showing T7, T11, T12 and L1 vertebral fractures.

my, prosthetic joint replacement; prevention of complications by adequate hydration, pain control, thermal equilibrium[4].

The treatment of vertebral fractures reported in the literature is mainly conservative. This article describes the clinical case of a young patient with sickle cell disease, showing multiple vertebral collapses, treated surgically.

Case report

A man aged 21, suffering from sickle cell disease with a known diagnosis (since the age of 5) was admitted because of the onset of spontaneous worsening pain at the thoraco-lumbar spine, associated to the radiographic imaging of multiple vertebral collapses (T7, T11, T12) (Figure 1). The clinical setting was characterized by severe and diffused osteopenia, hypovitaminosis D, relapsing broncho-pneumonic foci after sickling crises, and recurrent fever. The patient, closely followed by Hematologists, had been chronically treated with Neridronic Acid and Vitamin D, and had periodically undergone red blood cell exchange transfusions in order to reduce the percentage of HbS. At the admission it was impossible for the patient to stay in standing position because of the pain, even in the absence of peripheral neurological deficits. The HbS was present to the extent of 31%. Thoraco-lumbar spine CT and MRI showed a marked osteo-morphostructural remodeling with predominantly lytic features of all the examined vertebral bodies, pathological fractures of T11 and T12, angular kyphosis centered in T12 and height reduction of T7 as "fish mouth" appearance. After a careful assessment involving internist, hematologist and anesthesiologist, the patient was considered able to undergo laminectomy, bilateral artrectomy according to Ponte[5] at T12 level and T10-L1 arthrodesis with fenestrated cemented screws (Figure 2). Great attention was paid dur-

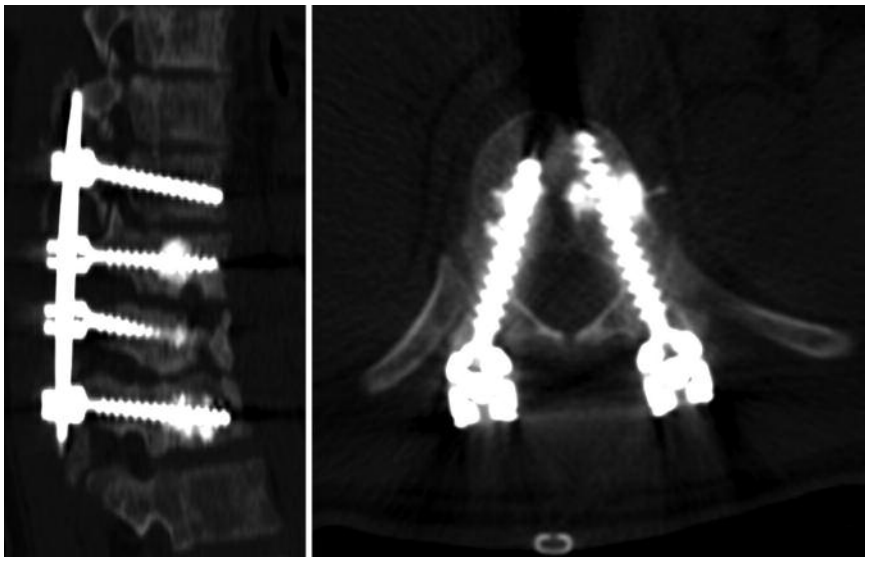

Figure 2. Post-operative sagittal and axial CT scan showing the bilateral artrectomy according to Ponte and posterior arthrodesis T10-L1 with fenestrated screws.

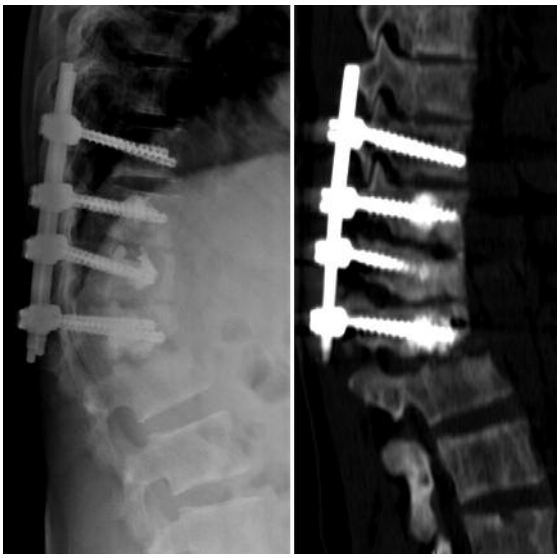

Figure 3. Lateral X-ray and sagittal CT scan performed four months after the first surgery, showing L1 fracture.

ing surgery and in the immediate postoperative period to control the body temperature, pain, post-surgical anemia; rehabilitation was carried out gradually and carefully, in order to minimize the risks for vaso-occlusive crisis. The post-operative course was uncomplicated. At the discharge the patient walked independently by wearing a three points corset. At the first examination, carried out 3 months after discharge, the patient's clinical condition was good: occasional back pain related to postural changes was observed, his walking was autonomous, the sagittal balance was correct.

Four months after the surgery an acute back pain appeared during the course of physiotherapy. A spontaneous fracture of L1 (Figure 3) was found and the patient underwent revision surgery with a new instrumentation T8-L3 and vertebroplasty of L4. Subsequently, standing was restricted for 40 days, followed by rehabilitation with corset and assisted ambulation. Four months later the patient was pain-free and the CT scan showed good evolution of arthrodesis, alignment preserved, no new fracture (Figure 4).

Discussion

The involvement of the osteo-articular system in the sickle cell disease gives rise to different clinical manifestations, in both acute and chronic presentation[2].

The microvascular occlusive episodes occur mainly in the bone marrow, causing its infarction and intense, acute pain located in one or more skeletal regions. The most common site of bone infarction is the intermediate region (meta-diaphysis) of the long bones[6]. All the long bones are involved, especially tibia and femur, and the presentation is often bilateral. A typical site, especially in childhood, are the terminal phalanges of hands and feet, with a picture of dactylitis. Another common problem is acute osteomyelitis (rarely septic arthritis), favored by hyposplenia, altered serum complement activity, and tissue ischemia with the presence of necrotic bone; the microorganism most frequently isolated in patients with sickle cell disease is Salmonella. Probably, a spontaneous vertebral body collapse can cause an acute pain.

The chronic involvement of the skeleton leads to progressive disability in patients affected by sickle cell disease. Epiphyseal ischemic necrosis occurs when repeated bone infarcts affect the subchondral region of the articular surfaces of the long bones; the most frequent site is the head of the femur, followed by proximal epiphysis of the humerus. Many studies have shown that pa-

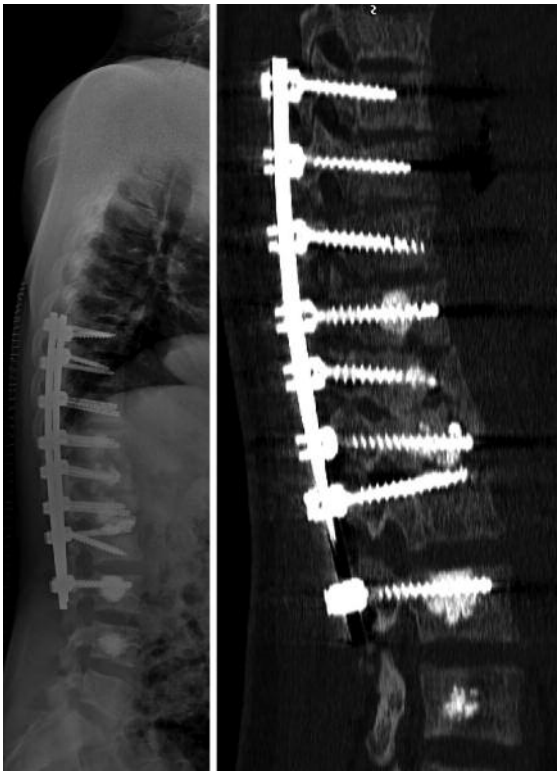

Figure 4. Lateral X-ray and sagittal CT scan performed after the second surgery, showing the revision arthrodesis T8-L3 and vertebroplasty in T4.

tients suffering from sickle cell disease have reduced bone mineral density, correlated to the medullary hyperplasia, which causes the biconcave deformity of the vertebral bodies called "fish-mouth".[1-3] As in elderly osteoporotic vertebra, spinal fracture may be nearly asymptomatic or otherwise may cause severe chronic pain, and eventually progressive deformity.

The involvement of lumbar and thoracic spine is common and widely described in literature. The pathogenesis is mixed: bone infarcts and osteopenia contribute to bone fragility leading to vertebral collapses; infectious spondylitis or spondylodiscitis are also relatively common. In the presence of fracture the treatment options listed are only conservative (rest, symptomatic therapy, orthosis)[8,9]; only Sadat-Ali cited, in a paper of 1994[3], the case of a patient presenting ischemic necrosis of two vertebral bodies which resulted in an acute kyphosis with incomplete paraplegia; treated by decompression and stabilization, he recovered the neurological deficit. On the contrary, the surgical option is often described in the column for the treatment of infections, because in the presence of extended abscesses the antibiotic therapy alone is insufficient[3,10,11]. In the literature there are also several papers regarding the surgical treatment of skeletal extravertebral manifestations, especially ischemic epiphyseal necrosis[12-14]. The high incidence of intra- and perioperative complications (transfusion reactions, acute respiratory syndrome, vaso-occlusive crisis) is always emphasized, in addition to the higher rate of implant failure than in patients treated for idiopathic avascular necrosis[15-17]. The high incidence of mechanical failure is related to the youngest average age of patients, but in particular to the altered quality of the bone in patients affected by sickle cell disease. The aim of the Anesthesiologist during surgery is to avoid acidosis secondary to hypoventilation, circulatory stasis, hypovolemia and hypothermia, in order to prevent vaso-occlusive crises triggered by any of these events[18].

In the case we treated, the choice of treatment was based on general and local clinical evaluations. The patient, a young adult man, was affected by sickle cell disease in its more severe form, but well-controlled with clinical medical treatment. The vertebral involvement resulted in very disabling symptoms and the pathological fracture of three vertebral bodies at the thoraco-lumbar spine junction caused a kyphosis deformity which was not serious at the time of observation, but it will certainly be expected to worsen, with possible consequent neurological complications. These considerations led us to choice a surgical option.

The patient was accurately prepared and specific care was taken both during and after surgery, in order to avoid any psycho-physical stress, particularly temperature changes, poorly pain control, hemodynamic changes. Regarding the pre-operative planning, we considered that the reduced bone density may cause a deterioration of mechanical properties at the bone-screw interface: the widening of the meshes of the trabecular bone, besides limiting the mechanical seal of the screws, also compromises the integration at the interface bone-prosthesis and causes a reduction of the force sufficient to mobilize the implant. As in the case of surgical treatment of the osteoporotic spine, we thought that surgery in a sickle cells patient is burdened by a high incidence of implant failure, with loosening of pedicle screws and pull-out[19]. There are several technical solutions to improve the tightness of the pedicle screws: use of larger screws (greater diameter, greater length), use of expansion screws, use of screws coated with osteoinductive material, and augmentation of pedicle screws fixation with acrylic cement (PMMA) or with resorbable cement. In the recent years our group has successfully used screws fenestrated in their distal part, through which the acrylic cement (PMMA) is introduced into the vertebral body; the injection of PMMA through cannulated and fenestrated screws provides additional stability in instrumented arthrodesis of an osteoporotic spine, may help to make shorter implants, saving the mobility of some segments, and reduces the risk of implant failure. The patient, despite the young age, presented severe secondary osteoporosis; therefore we considered this technique suitable for his case, in order to increase the fixation strength to the pedicle. Despite this, only few months after surgery we faced a complication, *i.e.* the collapse of the vertebra below the instrumentation. Once again we privileged the surgical option and this time we protected the lower junction with a vertebroplasty and promoted bone fusion by bed rest.

Cares and accurate technique were not sufficient to avoid an early mechanical complication (treated by a revision surgery), confirming reports in the literature.

Conclusions

Pathological vertebral fractures in course of sickle cell anemia are secondary to the avascular

necrosis of the vertebral bodies, which is associated with a context of severe osteoporosis. Although a conservative approach is most frequently indicated, we believe that the surgical option should be considered when a clinical worsening occurs in a young patient with otherwise well-controlled disease. The goal is to improve the quality of life; however, the risk of complications, even with the best preoperative preparation, remains high.

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Conflict of Interest

The Authors declare that no financial relationships and no conflict of interest concerning this manuscript.

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