Case Report

Complex Primary Total Hip Replacement in a Patient with Sickle Cell Disease and Contralateral Poliomyelitis: A Case Report and Review of Literature

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Abstract

Sickle cell haemoglobinopathy (SCH) and neurological sequelae of childhood poliomyelitis are still relatively common in this environment. The non-paralytic limb in polio patients is subjected to abnormal stresses due to excessive weight-bearing load, leg length discrepancy, pelvic obliquity and abnormal gait mechanics. We present this case to highlight the challenges of managing such a case and present our experience. A 47-year-old female with SCH presented with left-sided avascular necrosis (AVN) of the head of the femur and right-sided post-polio paralysis. Limb length discrepancy was 1.5 cm with a longer left lower limb. Oxford hip score (OHS) = 25 and SF12 = 16. Packed cell volume (PCV) was 20%. Radiographs revealed a destroyed left hip with secondary osteoarthritis, partial collapse, lateral subluxation and metaphyseal sclerosis. She had non-cemented left total hip arthroplasty via Hardinge approach. One-month post-operative scores were OHS = 38, SF12 = 24. Three-month follow-up scores were OHS = 46 and SF12 = 30. AVN poses a major burden. Combination with post-polio paralysis and the risk of infection with encapsulated organisms create a complex interplay of challenges. Peri-operative management requires meticulous monitoring, care and prevention of sickling crisis. Uncemented implants gave better results with fewer complications. Limb shortening at arthroplasty increases dislocation risk; so, length should be maintained. Coexisting post-polio paralysis and SCH is rare but challenging. The non-paralytic limb is subjected to excessive abnormal forces. Excellent results and low complication rates are achievable if meticulous peri-operative management, appropriate choice of cementless implants and maintenance of length are done. Ensuring adequate fixation of implants at surgery reduces risk of loosening. Rehabilitation must take the risk of falls into account. Pre-operative planning and optimisation, meticulous surgical technique and cementless implants are keys to success.

Keywords: Avascular necrosis, complex primary hip arthroplasty, hip arthroplasty, poliomyelitis, sickle cell haemoglobinopathy

Introduction

Sickle cell haemoglobinopathy (SCH) is a common haematological condition in Nigeria. It is widespread in Africa and parts of the Middle East and Mediterranean regions. The orthopaedic manifestations include bone infarcts, acute and chronic osteomyelitis (with a higher incidence of infection with encapsulated organisms than the general population) and avascular necrosis (AVN) affecting different regions of the body, most commonly the femoral head. By age 30 years, 30%–50% of patients will have developed AVN in the femoral head often involving both hips.[1] Recent advances in medical management of this condition have resulted in more people with haemoglobinopathy living into their 7th decade.

Femoral head AVN secondary to haemoglobinopathy presents with hip and groin pain, an antalgic gait and joint stiffness. Surgical treatment of AVN in the early stages involves options like core decompression with or without vascularised pedicle graft to improve the vascular supply to

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the head of the femur. Optimal management of late stages of haemoglobinopathy-induced femoral head AVN and subsequent end-stage arthritis is total hip replacement (THR). The occurrence of bone sclerosis with narrowing and sometimes obliteration of the proximal femoral medullary canal makes THR in these patients truly challenging. This is further complicated by increased medullary haemorrhage from the intense haematopoiesis, driven by the chronic haemolysis and anaemia. The osteoporosis that accompanies this haemopoietic response increases the risk of fracture and perforation during femoral canal preparation. Medical complications following this operation are not infrequent and result from the various medical co-morbidities associated with this condition and can be life-threatening.[2,3]

The hip joint of the normal non-paralytic lower limb of patients affected by lower limb post-polio paralysis is subjected to abnormal mechanical stresses due to excessive weight-bearing load, leg length discrepancy, pelvic obliquity as well as abnormal gait mechanics.[4] This limb is often longer than the paralytic limb with resulting point loading of the hip joint. The leg length discrepancy combined with the weakness in the paralytic limb as well as the joint deformities resulting from muscle imbalance, such as equinus deformity of the ankle can affect balance. A tendency to recurrent falls has been noted in patients with post-polio paralysis due to the aforementioned reasons.[5,6] There is also increased risk of dislocation of the paralytic but not the normal contralateral limb following THR.[4] The abnormal gait mechanics experienced by the normal non-paralytic contralateral hip imposes significant stresses on the hip joint with the potential risk of premature arthritis of the native hip or early loosening of a prosthetic hip joint.

The combination of haemoglobinopathy with its medical and musculoskeletal challenges combined with the challenges of THR in the normal non-paralysed contralateral limb in patients with post-polio paralysis makes hip arthroplasty a formidable proposition when both conditions are present in the same patient.

We present a case of THR for osteoarthritis secondary to AVN in the non-paralysed lower limb of a patient with contralateral post-polio paralysis who also suffers from SCH and review the literature on this rare combination.

Our objectives are to highlight the challenges of managing such a presentation and to present our experience. The challenges included the need for achieving stability intraoperatively because this patient required to be able to do immediate full weight bearing as the contralateral limb was paralytic from polio. The bone changes in sickle cell disease also make the bone sclerotic and prone to fracture with a narrow medullary canal. The risk of intraoperative crisis and post-operative surgical site infection were also significant.

Case Report

A 47-year-old female patient with SCH presented with pain and stiffness in the left hip which has progressively worsened over the years. There was no history of trauma to the hip. She had a history of poliomyelitis in childhood which resulted in paralysis of her right lower limb. The left lower limb is therefore her main weight-bearing limb and it is this hip that is arthritic. The pain was severe enough to cause limitation of daily activities and function and limit her productivity at work. She was rapidly becoming immobile and disabled.

Examination revealed a young woman walking with antalgic gait. There was marked wasting of the right lower limb with shortening and compensatory equinus at the ankle joint, the result of childhood polio. There was leg length discrepancy of about 1.5 cm with the arthritic left leg from AVN being the longer leg. This was explained by the long-standing atrophy from the poliomyelitis in the right lower limb causing shortening which was more than that produced by the AVN in left lower limb. The left lower limb revealed pain on rotation of the hip joint. There was marked painful limitation of both passive and active abduction to <10°. Her Oxford hip score (OHS) was 25, Harris hip score was 20 and her SF12 score was 16. Her steady state PCV was 20%, white blood cell count was 8600/mm^3 and electrolytes, urea and creatinine were all within normal limits. Her radiographs [Figure 1] revealed a destroyed left hip with secondary osteoarthritis of the hip, partial collapse and lateral subluxation of the femoral head with areas of luscency and sclerosis within the head. It also revealed extensive femoral metaphyseal sclerosis and obliteration of the medullary canal due to the underlying SCH. There was also radiological evidence of atrophy of the right femur as the diaphysis of the bone was much narrower than the left side.

Pre-operative templating was carried out to plan the surgery and ensure optimal restoration of anatomic biomechanics in order to achieve lasting reconstruction.

She had left THR which was done via the Hardinge approach which is the surgeon's preferred approach with the additional benefit of reduced dislocation risk. The femoral head was dislocated with some difficulty and the joint was found to be severely arthritic with florid periarticular osteophytes. The bone of the acetabulum was soft and required gentle concentric reaming. The acetabulum was reamed to size 53 mm and a size 54 mm cluster acetabular shell was impacted with primary stability augmented with 3 screws.

The femoral canal was found to be obliterated in the metaphyseal region. Rather than risk a fracture, the canal was accessed with a 4.0 mm drill bit and a guide wire inserted into the canal under image intensifier guidance. Flexible reamers were then used to widen the canal over the guide wire. The canal was reamed to size 11 mm down to the junction of the proximal and middle thirds of the shaft while the metaphysis was reamed to size 12 to accommodate the metaphyseal flare of a tapered stem. The canal was next broached to size 6 and trialled with the standard offset neck and a 32 mm by 45 mm neutral liner and 32 mm +1 trial head. The joint was

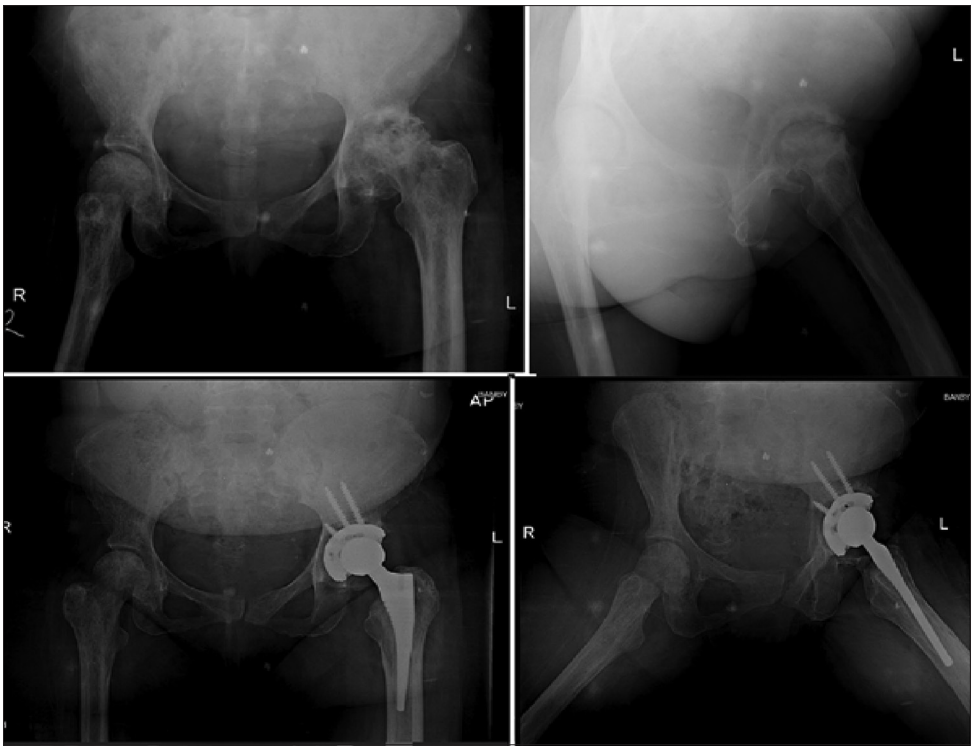

Figure 1: Pre-operative and 3-month post-operative radiographs in anteroposterior and lateral views

carefully tested for stability by putting the hip through the range of motion as well as the shuck test. Component position was confirmed with the Ranawat Coplanar test for assessing combined anteversion. These were all found to be satisfactory. Careful assessment also showed that the leg length had not been significantly altered and adequate soft tissue tension had been achieved. Definitive neutral highly cross-linked polyethylene liner was inserted. A size 6 standard offset Corail stem was impacted with a 32 mm +1 ceramic head and the joint reduced and stability reconfirmed. The cavity was copiously irrigated by jet lavage. The muscles were repaired with trans-osseous sutures and the wound closed meticulously in layers after securing haemostasis. Her post-operative radiographs were satisfactory [Figure 1]. The patient was commenced on immediate supervised full weight-bearing mobilisation with walking aids with emphasis on both dislocation and falls precautions.

At 1-month follow-up, the wound had healed completely with no wound complications. She had regained independent mobility and her pain had significantly subsided. Her scores were universally improved with Harris hip score of 64, OHS of 38 and SF 12 score of 24. At 3-month follow-up, her scores were further improved to OHS of 46 and SF 12 score of 30. She had a limb length discrepancy of 5 cm (the hypoplastic right lower limb affected by post-polio paralysis was now shortened compared to the restored left lower limb) which was compensated for with a shoe-raise. With the use of the shoe raise, she no longer walked with the right ankle joint in equinus and her mobility was much improved.

Discussion

In an environment where childhood polio infection was previously common but is now virtually eradicated, it is not unusual to see the residual effects of post-polio paralysis in middle-aged individuals. SCH is still very common, but the combination of sickle cell disease and post-polio paralysis in a patient undergoing THR is rare.

The orthopaedic manifestations of sickle cell disease often lead to lifelong deformity and suffering for patients who experience such. The predisposition to and frequent occurrence of AVN of the head of the femur places a lot of burden on the health care system and causes a lot of pain and limitation of activity for the patients. The optimum management of late stages of haemoglobinopathy-induced AVN and the end-stage arthritis that results, is THR.

The incidence of infection with encapsulated organisms due to decreased opsonisation from autosplenectomy in sickle cell disease patients is a major challenge. This makes the perioperative management and infection control even more pertinent in them than in the otherwise healthy controls. The challenges of precipitating a sickling crisis in the event of hypothermia, dehydration, severe pain, hypoxic stress and other stressors that may occur peri- or intra-operatively makes their perioperative management more challenging. There is increased medullary haemorrhage from the intense haematopoiesis, driven by the chronic haemolysis and anaemia. In the presence of an already low PCV, such blood loss is poorly tolerated; hence, the haematological management of sickle cell patients is a critical factor in performing safe surgery. In

the index case, pre-operative haemoglobin level was low, and transfusion was started early intraoperatively.

Early literature on outcome of cemented THR for AVN secondary to haemoglobinopathy was abysmal with very high medical and surgical complication rates and poor survival of the implants.[1,7-9] Thankfully more recent studies[2,10] have demonstrated significant improvement in both medical and surgical outcomes with mid-term to long-term survival approaching the results of THR for primary osteoarthritis. This has been due to overall improvement in the medical management of patients with haemoglobinopathy, improvement in surgical technique and use of uncemented femoral and acetabular implants. A 10-year survivorship of 98% has been demonstrated[10] equalling the best result of any international registry data and a 15-year survival of 94.1% for all cause failure with the use of cementless components. Equally impressive is the 95% survival free of aseptic loosening at 7 years[2] using cementless components which was equivalent to the survival of non-sickle cell AVN patients. These were accompanied by significant reduction in complications and excellent improvement in functional outcome. The complications that came to the fore relate to increased wear rates including balloon pelvic osteolysis[10] and loosening[2] and reflect the activity demands of these young patients whose average age is often around 25 years.[10,11]

We were guided by the encouraging results of these recent reports[10,2] in carrying out this THR. This we did by optimising the medical management of this patient, in our choice of cementless acetabular and femoral implants and in the careful and controlled negotiation of the sclerotic/blocked proximal femoral metaphysis with the use of image intensifier and the choice of X-linked polyethylene liner and ceramic head as a hard wearing articular couple in this relatively young patient.

With regard to the associated polio paralysis, an area of concern is ensuring that we did not reduce the length of the non-paralytic longer operative leg by resisting the tendency to equalise the leg length discrepancy as attempts to do this will result in increased risk of dislocation. Also, any further lengthening of this longer limb will affect this patient's balance and may increase the risk of falls. We ensured that by a combination of pre-operative templating, intraoperative measurements and careful reconstruction, leg length was not significantly altered. We undertook gluteal tendon repair as a trans-osseous repair using non-absorbable suture to reduce the risk of dehiscence and limp. The need for a hard-wearing articulation in this main weight-bearing lower limb was addressed by the choice of X–linked polyethylene–ceramic articular couple as stated earlier.

A review of the literature on outcome of THR of the paralytic as well as the non-paralytic lower limbs of patients with residual post-polio paralysis provided invaluable information on optimum perioperative/surgical management of this cohort of patients.

A review of the result of 10 patients with polio paralysis undergoing THR with 4 in the paralytic limb and 6 in the contralateral normal limb at mid-term follow-up demonstrated excellent pain relief in the patients with hip replacement and bone ingrowth with no evidence of loosening in all cases.[12] There was one case of dislocation which remained stable following closed reduction.

Despite the theoretical concerns, prosthetic loosening following THR in patients with post-polio paralysis does not seem to be a concern in the literature. A study found no loosening at 8 years follow-up.[12] This was also the finding in a study that showed a 10-year survival free of loosening or other complication of 91%.[13] Finally, another study found no loosening or other complication at 2–8-year follow-up.[4]

This is the first report as far as we are aware, of THR in the normal non-paralytic lower limb in a patient with both polio paralysis and haemoglobinopathy.

The challenge of replacing the hip of the non-paralytic main weight-bearing leg in this patient with post-polio paralysis as well as haemoglobinopathy, is to optimise the joint biomechanics and accommodate the demands of high weight-bearing loads. It was also necessary to ensure accurate implant positioning to reduce the risk of dislocation while ensuring long-term durable reconstruction. This was achieved by also taking cognisance of and tackling the peculiar medical and skeletal challenges posed by the patients' SCH.

Conclusions and Take-Home Message

1. Residuals of post-polio lower limb paralysis is common in our environment as is SCH, but the finding of both pathologies in the same patient is rare
2. Hip reconstruction for each pathologic condition represents a complex hip replacement as there are exceptional challenges from each pathology. The combination of pathologies in the same patient can make hip reconstruction doubly difficult
3. There is encouraging recent data showing excellent results for THR in patients with haemoglobinopathy with associated reduction in complication rates
4. The non-paralytic lower limb of a patient with post-polio palsy is subjected to excessive loads and abnormal gait mechanics with potential risk of premature wear and loosing
5. There is an increased risk of recurrent falls in post-polio patients. Surgery may increase this risk, and post-operative rehabilitation must give it due consideration
6. Optimal pre-operative planning, multidisciplinary approach to pre-operative optimisation, improvement in surgical technique and the use of cementless implants and durable articular couple are the keys to success.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have

given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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