Achievement of Functional Independence in a Patient with Sickle Cell Disease with Autoimmune Hepatitis, Osteomyelitis, Wilson's Disease, and Pathological Fracture Following Physiotherapy

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INTRODUCTION

Sickle Cell Anaemia (SCA) is an autosomal recessive condition caused by malfunction in the synthesis of haemoglobin (Hb) where a single amino acid glutamic acid in the β globin gene is exchanged with valine.[1] The red blood cells assume an abnormal biconcave disk into elongated, rigid, crescent-shaped sickle cells, causing haemolysis and occlusion of microcirculation, which then in turn will lead to hypoxia and sickling of the tissue,[2] reducing their capacity to microcirculation, with frequent clotting and thrombosis, leading to obstruction, ischemia and infarction, damaging many organs.[1]

Auto-Immune-Hepatitis (AIH) has high risk of steroid-related harmful reactions or other immunosuppressive therapies.[3] Blood transfusions is useful in prevention and treatment of acute and chronic conditions.[4] The subsequent elevated excretion of bilirubin predisposes to cholelithiasis, which might be attributed to several other hepatological complications such as haemosiderosis related to transfusion, persistent viral hepatitis, and autoimmune liver disease (AILD).[5]

Tissue hypoxia causes a supplementary inflammatory response resulting in higher intramedullary pressure and bone pain leading to osteopenia, fractures of stress, vertebral collapse, and growth anomalies. The combination of ischemic and compromised immunity will trigger osteonecrosis and haematogenous osteomyelitis.[2] Immunodeficiency leading to splenic dysfunction, tissue infarction and excessive iron content contributes to elevated incidence of vaso-occlusive-crisis (VOC) and haematogenous-osteomyelitis. Medullary bone infarction and necrosis generate an appropriate environment for development and spread of bacteria. Osteomyelitis is commonly described as infection that involves tibia, femur and humeral diaphysis, along with vertebral infection. *Salmonella* preceded by *Staphylococcus aureus* are organism causing sickle cell-related haematogenous osteomyelitis.[1]

Sickle Cell Anaemia (SCA) is an autosomal recessive genetic disease where single amino acid; glutamic acid is exchanged by valine in β globin gene. Red blood cells assume an abnormal, rigid, and sickle structure, damaging many organs. The combination of ischemic and compromised immunity will trigger osteonecrosis and osteomyelitis. Immunodeficiency leading to splenic dysfunction, tissue infarction and excessive iron content contributes to elevated incidence of VOC and osteomyelitis.

In this case report we present a 25 years old student who was referred to physiotherapy department for further management of a pathological fracture. She has a history of SCA, Wilson's disease, autoimmune hepatitis and osteomyelitis with frequent requirement for blood transfusion.

In this study a patient having sickle cell anaemia, Wilson's disease, autoimmune hepatitis, osteomyelitis and external fixator fracture for displaced mid-shaft implant was able to resume ADLs (Activities of Daily Living) and independent ambulation.

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PRESENTATION OF CASE

In this case report, we present a 25 years old student who was referred to physiotherapy department for further management of a pathological fracture. While doing bed mobility which was assisted by her mother, a loud cracking sound was heard. She was taken to Acharya Vinobha Bhave Rural Hospital (AVBRH), DMIMS (DU), Wardha, Maharashtra, India, where she was diagnosed with pathological fracture (Figure 1, 2). One-month prior, a sequestrectomy was done and around 1.5 litres of pus was exerted, which lead to breaking of bone in the area leading to a compound midshaft femur fracture on the same site from where the pus was extracted. An external fixator was applied (Figure 3, 4, 5, 6) to stabilize the fracture. She has a history of SCA, Wilson's disease, and autoimmune hepatitis and haematogenous osteomyelitis with frequent requirement for blood transfusion. In the past year she was hospitalized several times for concerns such as fever, abdominal pain, vomiting, swelling over the face, chill fever, dry cough, pain in both lower limbs, pain and swelling over the left thigh, swelling of the lower limbs, breathlessness, etc. for which she was undergoing treatment in same hospital.

Clinical Findings

She was evaluated in supine lying position with both shoulders at the same level with external fixator in left thigh. On inspection, left leg was abducted and slightly externally rotated, knee extended with pillow support below, leg and ankle where ankle was slightly plantar flexed. Swelling was present on left mid-thigh, knee and operative site. Local temperature was increased on palpation and non-pitting oedema (Figure 7) (Table 1, 2).

Muscles	Right	Left
	Hip	
Flexors	Poor	Poor
Extensors	Poor	Poor
Abductors	Poor	Poor
Abductors	Poor	Poor
	Knee	
Flexors	Poor	Poor
Extensors	Poor	Poor
	Ankle	
Plantar Flexors	Poor	Poor
Doris Flexors	Poor	Poor
Invertors	Poor	Poor
Evertors	Poor	Poor

Table 1. Isometric Strength

Joint	Active	Passive	Limitation
		Help	
Flexion	NA	NA	Unable to perform due to pain
Extension	NA	NA	Unable to perform due to pain
Abduction	NA	NA	Unable to perform due to pain
Abduction	NA	NA	Unable to perform due to pain
		Knee	
Flexion	NA	NA	Unable to perform due to pain
Extension	NA	NA	Unable to perform due to pain
		Ankle	
Plantar Flexion	0 - 20^0	0 - 25^0	NA
Doris Flexion	0 - 10^0	0 - 15^0	NA
Inversion	0 - 15^0	0 - 15^0	NA
Eversion	0 - 10^0	0 - 10^0	NA

Table 2. Range of Motion

Figure 1, 2. Pre-Op X-Ray A-P and Lateral View

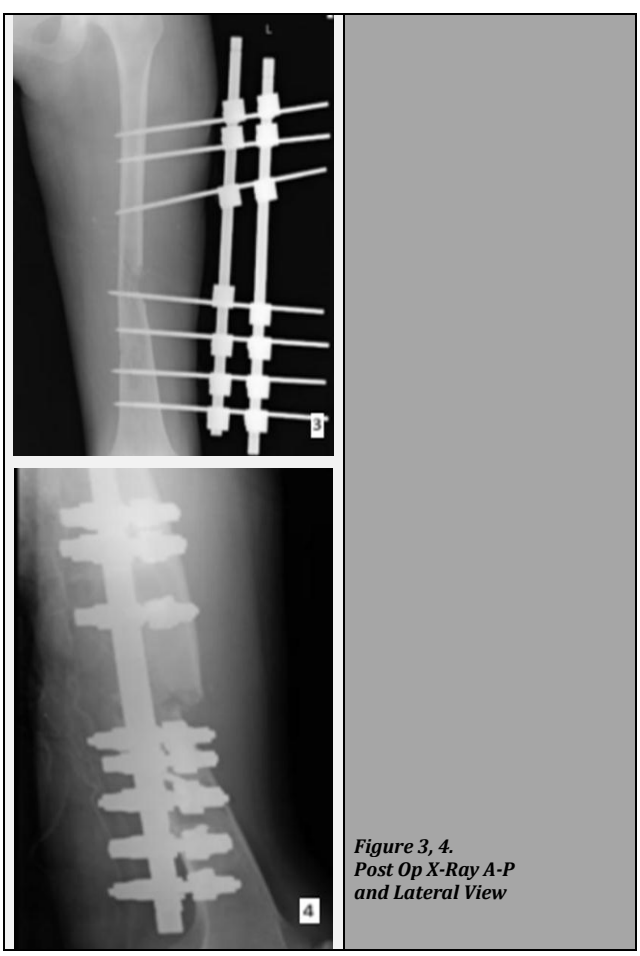

Figure 3, 4. Post Op X-Ray A-P and Lateral View

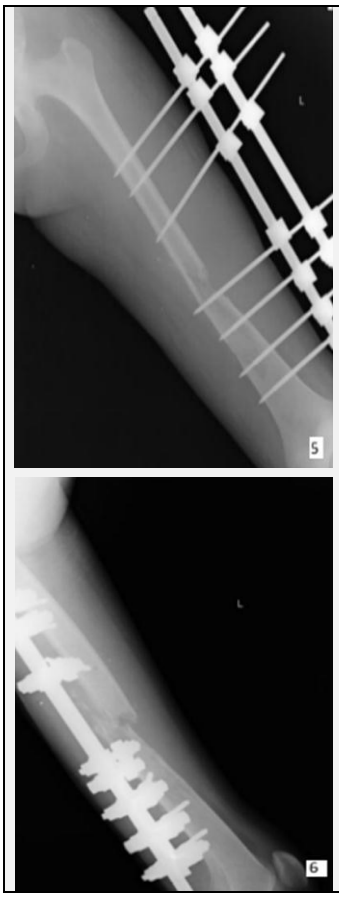

Figure 5, 6. Post Op A-P and Lateral View

Events	Date
Diagnosed with Sickle Cell Anaemia, Wilson's Disease and Autoimmune Hepatitis	12 / 07 / 2019
Visited AVBRH with complains of pain swelling over the left thigh	09 / 04 / 2019
Diagnosed with Osteomyelitis.	09 / 03 / 2020
Sequestrectomy and Saucerization of left tibia and femur.	11 / 03 / 2020
Revisited AVBRH as tired to lift up pelvis when a cracking sound was heard along with complains of sudden pain inability to move leg which didn't reduce.	01 / 04 / 2020
Diagnosed with compound midshaft femur fracture	01 / 04 / 2020
External Fixator was applied for compound midshaft femur fracture	02 / 04 / 2020
Referred to Physiotherapy for further management	05 / 04 / 2020

Figure 7. Timeline

DISCUSSION OF MANAGEMENT

Medical Management

She is a known patient of sickle cell anaemia, Wilson's disease, autoimmune hepatitis, and haematogenous osteomyelitis with frequent requirement for blood transfusion (haemoglobin: - 4 gm %), was managed with saline washed PRC transfusions and injectable antibiotics and tablet hydroxyurea once daily, Tablet Folic Acid 5 mg once daily and Tablet Sodamint thrice daily. One-month later patient developed tense red hot swelling around the knee and midshaft femur, for which an emergency incision and drainage of the swellings and 1.5 litres of pus was drained out from the midshaft femur and proximal tibia. Sequestrectomy was done for it. The reports of culture showed growth of *staphylococcus aureus* with sensitivity to amikacin. The patient was started on injection amikacin for two weeks with regular KFT monitoring and regular blood transfusions.

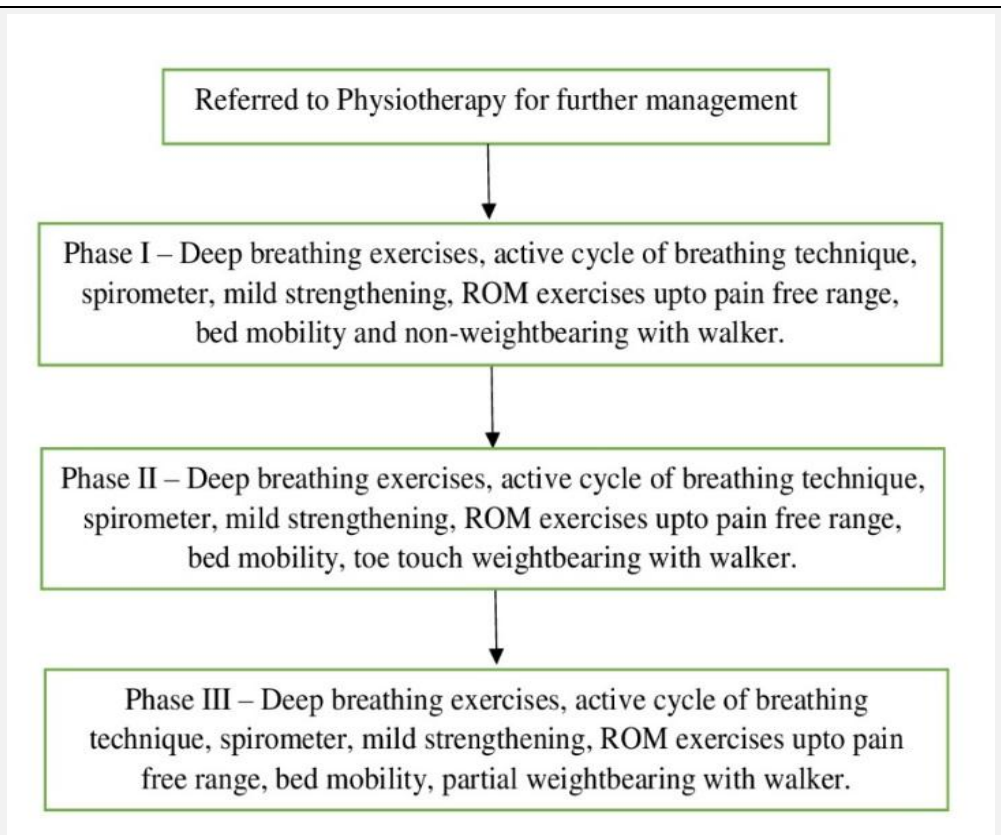

Figure 9. Rehabilitation

The patient was then discharged on oral ciprofloxacin. After one-month patient again presented with pathological fracture midshaft femur left side. She was managed with realignment of the fracture and external fixator application with three proximal and three distal pins and connected with two connecting rods and injectable antibiotics.

Post-Operative Goals

The short-term goals were to avoid respiratory problems, minimize pain and oedema, preserve and improve the joint range of motion and strength, facilitate early mobility, prevent pressure sores, facilitate walking (non-weight bearing) and independent ADLs. Long-term goals involved improve joint range of motion and strength, encouraging walking, balance, independent ADL and ergonomics.[6–12] (Figure 7).

Post-Operative Management

Phase I (Inpatient. Postoperative: 0 - 4 Weeks)

Phase I activities concentrate on breathing exercises, joint mobility in the hip and knee, non-weight-bearing strengthening, statics and active movements.

Relaxation techniques, active cycle of breathing technique, deep breathing and spirometry avoid problems in the respiratory system. Movement of the ankle toe to prevent oedema. Static quads, static hams and static glutei which sustain the quadriceps, hamstring, and glutes strength. Half bridging to sustain core strength (using leg and elbows that are not affected). Active exercises for unaffected limbs were continued along with proper positioning and bed mobility to reduce the occurrence of pressure sores, active assisted SLR (Straight Leg Raise) to retain quadriceps and hamstring strength, to decrease joint stiffness and increase range, active assisted hip abduction, adduction to preserve hip abductor and adductor flexibility, to decrease joint stiffness and increase range. Heel slides to retain quadriceps and hamstring strength, decrease joint stiffness and increase range, active quadriceps assisted dynamic. Long sitting and bed side were promoted sitting with or without a bed support. Bed side standing with walker and walking with walker (non-weight bearing) to enhance balance, reduce fear of falling, obtain confidence and strength.

Phase II (Outpatient. Postoperative: 4 - 8 Weeks)

Most phase I aspects persisted through phase II, as needed. Weight bearing with walker (toe touch). Phase II workouts featured a transformation of phase I exercises with further weight-bearing activities, additional gait retraining, and strength conditioning incorporation.

Resisted exercises were performed with increased motion range, such as SLR, prone SLR, side SLR, heel slides, and dynamic quadriceps. It was encouraged to stand and walk with a walker (partial weight bearing).

Phase III (Outpatient. Postoperative: 8 - 12 Weeks)

The goal during Phase III is to promote balance, mobility and endurance training. The exercises focused on increasing the strength of the lower extremity as tolerated lower extremity walker activities.

Resisted exercises were done with moderated resistance with increased range of motion such as SLR, prone SLR, side SLR, heel slides, dynamic quadriceps. Standing and walking with walker was facilitated. Gait training exercises such as marching on the spot, stepping, ascending stairs, stride walking retro walking, tandem walking, high step walking to enhance coordination, promoting strength.

Limitations

As she was confined to bed with less to no bed mobility, the cardiovascular endurance and strength was less. As a result, the patient would get fatigue soon resulting in less output than required which delayed the rehabilitation process of making patient self-dependent.

DISCUSSION

In this case report, we found, a patient with history of osteomyelitis, for which sequestration was done for left leg with 1.5 litres of pus extracted leading to pathological fracture which was managed with an external fixation. Goals for rehabilitation were set, starting from mild exercises to strengthening and assisted weight bearing ambulation. All the exercises were performed with each one three times a day 10 sets. There are literatures that show the effects of internal fixation and Ilizarov fixation its effect on rehabilitation, but research on external fixation and routine rehabilitation are missing.

Early mobilization and total weight bearing[8] in patients with femur fractures was commonly correlated with a quick and stress-free recovery with very little effect on pain, hip mobility and walking capacity. We couldn't mobilize her early as due to weakness due to her prolonged stay in hospital.

As program comprising hip range of motion exercises, isometric exercises and eccentric strengthening as suggested by recent researches,[9,10] enhances strength of hip flexors, abductors, walking rhythm and cadence. We incorporated isometrics, eccentrics and range of motion exercises.

Functional limitations mostly influence the result as study done by; of patient who underwent femoral fractures included hip abductor weakness, quadriceps femoris muscle weakness and anterior knee pain might relate to an altered gait pattern individually or collectively.[11] Thus, we enforced more on strengthening them

A flexible protocol was made by our team considering her weakness and to enhance her strength and endurance,[12] sufficient considerations should be offered to exceptional cases with reference to injuries and the rehabilitation protocol, flexibility with certain circumstances might be required; furthermore, all attempt must be made to proceed with a goal-based development of rehabilitation protocol, specific with progress to weight-bearing activities as quickly as possible, since this guides rehabilitation advancement.

CONCLUSIONS

Rehabilitation program successfully minimizes post-operative pain, oedema, increased strength and range of motion. In this

study, a patient having sickle cell anaemia, Wilson's disease, autoimmune hepatitis, osteomyelitis, and external fixator fracture, for displaced mid-shaft implant was able to resume ADLs and independent ambulation.

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Disclosure forms provided by the authors are available with the full text of this article at jemds.com.

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