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Pre-operative physiotherapy impact in early post -operative recovery in a case of bilateral avascular necrosis secondary to sickle cell disease in rural India - Case report

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ABSTRACT

An inherited disorder of red blood cell, Sickle cell disease is characterized by inadequate amount of healthy red blood cells in the blood to therefore reducing the body's oxygen carrying capacity. In sickle cell disease RBC are crescent shaped so while passing through the blood vessel they block the blood flow and reduce the oxygen supply to the tissues. Normally the average lifespan of RBC is about 120 days but in sickle cell anemia it is reduced to only 10 to 20 days. So leading to various complications, out of all of them Avascular necrosis of femoral head is the most common due to precarious blood supply to that region. Presenting a case of 45 year old elderly male, who visited our hospital with the major complaints of pain in both hips since 1 month along with difficulty in walking and after investigations was diagnosed with bilateral Stage 4 avascular necrosis of femoral head. Pre-operative Physiotherapy was started. Due to low economic status patient underwent only unilateral total hip replacement. Post-Operative physiotherapy was started on the same day of operation and rehabilitation was progressed into 3 phase that is immediate post-surgical phase, motion phase and advanced strengthening and higher level function stage. The outcomes were satisfactory and pre-operative physiotherapy was found to be highly beneficial in post-operative period in gaining early recovery. Multidisciplinary approach and patients positive attitude was the key to get the favorable outcomes.

Keywords: Sickle cell disease, avascular necrosis, physical therapy, total hip replacement arthroplasty, case report

1. INTRODUCTION

Sickle cell disease is an inherited disorder caused by mutation in hemoglobin gene which makes the red blood cell (RBC) rigid, sticky and crescent shaped

which causes reduced lifespan of RBC and insufficient supply of oxygen rich blood to various tissues (Ilesanmi, 2010). Femoral head having precarious blood supply so is prone to undergo avascular necrosis (Narayanan et al., 2017). Avascular necrosis of femur causes pain in the ball and socket hip joint, along with difficulty in bending activities. Treatment of choice depends on the stage of necrosis, bone stock, age, occupation and life expectancy of the patient. Total hip replacement arthroplasty is treatment of choice in severe stages where there is complete deformation of bone with significant reduction in the joint space (Agrawal et al., 2021). Method of fixation varies from cemented to non -cemented type, which has its advantages like non cemented type provides ease in revision surgery (Moya-Angeler et al., 2015).

Physiotherapy is found to be highly beneficial in pre as well as post- operative period to gain functional independence, reduce pain and in prevention of secondary complications. In sickle cell disease there are increased chances of deep vein thrombosis and respiratory complications due to hampered blood flow (Gabada et al., 2021; Martí-Carvajal et al., 2019). A physiotherapy rehabilitation with adequate intervals is found to be very effective in improving the endurance (Torisho et al., 2019). We present a case of patient who was managed in a multispecialty hospital with total hip replacement arthroplasty.

2. PATIENT INFORMATION

45 year old male resident of Gadchiroli district and farmer by occupation was apparently alright 3 months back until he started experiencing pain over both the hips along with restricted mobility bilaterally, right side more than the left. Pain was deep dull aching at rest in nature and sharp during activities which increased in intensity in the past 3 months and got relieved medications. On Numerical pain rating Scale patient rated 8/10 on rest. The complaints progressed over the period of last 30 days to an extent that it started affecting the activities of daily living like sitting crossed legged, walking and squatting. The patient is a known case of sickle cell disease which was diagnosed in 2011. Patient was previously admitted in government hospital at Gadchiroli where he was managed conservatively but there was no relief in pain so from there he was referred to hospital for further evaluation and management. On orthopedic consultation the patient underwent various investigations including X -ray, which revealed stage 4 Avascular Necrosis bilaterally according to Ficat and Arlet Classification. So, considering the stage of disease, age, bone quality and occupation, patient was planned for bilateral total hip replacement arthroplasty. Physiotherapy was started pre operatively on 30th December 2021. Due to financial constraints patient underwent total hip replacement arthroplasty for only right side due to more restricted mobility compared to left. Post-operative rehabilitation was started on the same day of operation that is 2nd January 2022 with a protocol made with consulting the treating orthopedic surgeon.

3. CLINICAL FINDING

Physical examination showed presence of conjunctival pallor due to anemia. There was grade 2 tenderness over joint line bilaterally with limited and painful Range of motion on right side more than left side (Table 1). Thigh muscles showed atrophy on both sides when compared to patient's general body built. There was weakness in both lower limbs right more than the left (Table 2). On examining tightness there was positive finding in hamstrings muscle on right side. Gait assessment revealed severe disturbance in gait due to painful flexion and rotation and bilateral positive Trendelenburg sign leading to waddling gait. On neurological examination there were normal reflexes and intact sensation on both the sides.

Table 1 Pre Rehabilitation Range of Motion

Joint	Movement	Right Active	Right Passive	Left Active	Left Passive
Hip					
	Flexion	0-60^0	0-80^0	0-80^0	0-110^0
	Extension	NA	0-30^0	0-10^0	0-45^0
	Abduction	0-20^0	0-40^0	0-30^0	0-50^0
	Adduction	0-20^0	0-30^0	0-35^0	0-45^0
	Internal Rotation	0-30^0	0-35^0	0-35^0	0-45^0
	External Rotation	NA	0-40^0	0-10^0	0-50^0
Knee					
	Flexion	0-110^0	0-120^0	0-120^0	0-125^0
	Extension	110-0^0	120-0^0	120-0^0	125-0^0

Ankle					
	Dorsiflexion	$0\text{-}10^0$	$0\text{-}20^0$	$0\text{-}30^0$	$0\text{-}40^0$
	Plantarflexion	$0\text{-}30^0$	$0\text{-}45^0$	$0\text{-}35^0$	$0\text{-}45^0$
	Inversion	$0\text{-}25^0$	$0\text{-}35^0$	$0\text{-}30^0$	$0\text{-}40^0$
	Eversion	$0\text{-}25^0$	$0\text{-}25^0$	$0\text{-}30^0$	$0\text{-}35^0$

Table 2 Pre Rehabilitation- Manual muscle grading

Muscles		Right	Left
Hip			
	Flexors	3/5	4/5
	Extensors	3/5	3/5
	Abductors	3/5	3/5
	Adductors	3/5	3/5
Knee			
	Flexors	4/5	5/5
	Extensors	4/5	5/5
Ankle			
	Dorsiflexors	4/5	5/5
	Plantar flexors	4/5	4/5
	Invertors	4/5	5/5
	Evertors	4/5	4/5

Timeline

In April 2011 he was diagnosed with sickle cell disease. Since October 2021 he started experiencing pain in the hip joint over right side and after few days over the left side. In November 2021 patient was admitted in Gadchiroli and was managed conservatively but due to no relief of symptoms he visited to the hospital OPD on 26th December 2021 and was admitted to Male Ortho Ward on the same day X-Ray was performed and was diagnosed with bilateral avascular necrosis of femoral head. On 27th December 2021 laboratory investigations were done along with sickling and then on 30th December 2021 again CBC was carried out and Pre-operative physiotherapy was started. Due to low hemoglobin there was a delay in the intervention. On 2nd January 2022 patient underwent total hip replacement arthroplasty for right side and on the same day Post-Operative Physiotherapy Rehabilitation was started.

Diagnostic assessment

CBC, Coagulation profile, E.S.R, sickling, CRP, peripheral smear was performed. CBC revealed low hemoglobin that is 7.2% with reduced RBC count (Table 3). There was raised E.S.R. and CRP suggesting of inflammatory changes. Early sickling was negative but late sickling was positive, patient was further adviced Hb electrophoresis for confirmation. Peripheral smear revealed RBC as microcytic hypochromic with anisopoikilocytosis with few pencil cells and occasionally fragmented RBCs platelets. KFT and LFT findings were normal. Due to economical restrains patient was unwilling to perform the MRI scan. X-ray was therefore the main diagnostic tool (Figure 1-2).

Figure 1: The X- ray pelvis Antero-posterior (AP) view showed increased density with lytic areas on right and reduced joint spaces between left head of femur and acetabulum. The imaging at this point indicates bilateral Stage 4 avascular necrosis of femoral head

Table 3 Pathological investigations post admission with clinically positive findings

Investigations	Investigation Date	Positive Findings				
CBC		Hb %	RBC count	WBC count	MCHC	MCH
	27-12-2021	7.2	4.12	4300	30.8	17.6
	31-12-2021	11.1	5.41	7300	32.1	20.6
ESR	27-12-2021	54				
Sickling	29-12-2021	Early		Late		
		Negative		Positive		
Peripheral Smear	27-12-2021	RBC -Microcytic hypochromic				
	31-12-2021	RBC-Normocytic Hypochromic				

Figure 2 Post total hip replacements X-ray showing Prosthesis placed for right hip joint

Diagnosis

The clinical and diagnostic findings were suggestive of bilateral stage 4 avascular necrosis of femoral head secondary to sickle cell disease.

Therapeutic intervention

Prior to the surgery patient was treated for sickle cell anemia by blood transfusion .Patient was then planned for bilateral total hip replacement. Pre – operative Physiotherapy was started as soon as surgery was planned as mentioned in (Table 4) but due to financial constraints unilateral right side hip replacement arthroplasty was done. The approach used was posterolateral so primary focus of physiotherapist is brought into the muscles incised that are Piriformis and short external rotators .The implant fixation method was non cemented type. The rationale behind using this method is presence of good bone stock and has an advantage of relative ease during revision THR surgery. The post-operative physiotherapy rehabilitation was planned with consulting the

orthopedic surgeon. It was divided into 3 phases that were immediate post-surgical phase, Motion phase and advanced Strengthening and higher level function stage (Table 5).

Table 4 Pre-operative physiotherapy Rehabilitation

Intervention	Duration and Dosage	Rationale
Patient Education	Educating the Importance of physiotherapy Before and after operation.	Helps to improve treatment effectiveness and gains early recovery
Deep Breathing exercises	5 Repetitions every hourly throughout the day.	Improve vital capacity of the lungs and getting rid of post anesthetic secretions
Ankle toe Movements	20 repetitions 3 times a day	Prevent accumulation of blood and improves circulation
Strong and sustained isometrics contractions to Gluteus, Hams, Quads.	20 repetitions 3 times a day with 10 second hold each	Helps to improve strength and endurance in both the limbs.
Resistive Exercises for upper limbs and foot	10 repetitions using yellow theraband progressing to green for 2 times a day	Facilitate Early ambulation with walking aids

Table 5 Post-Operative Physiotherapy Rehabilitation

Phase of Rehabilitation	Intervention	Duration and Intensity	Rationale
Phase 1 - Immediate Post Surgical Phase	Ankle Pumps	20 Reps × 2 set	Improves blood circulation
	Isometric Quads , Hams and gluteus	10 Reps × 2 set with 5 sec hold	Increases strength and endurance
	Unilateral Bridging	10 Reps × 2 set with 5 sec hold	Strengthens core abdominal muscles
	Active assisted Hip Abduction	10 Reps × 2 set	Prevents Adduction deformity
	VMO strengthening	20 Reps × 2 set	Helps in gaining end range knee extension
	Dynamic Quads	10 Reps × 2 set	Increases strength of Quads
Phase 2 - Motion Phase	Stretching of hip Abductor	3 Sets with 30 second hold	Maintain Muscle Elasticity
	Heel Slides	10 Reps × 2 set	Improve Hip Flexion
	Straight Leg Raise	10 Reps × 2 set with 10 second hold	Improve Strength of lower limbs
	Gait Training with assistive aid	2 times a day	Improve weight bearing
	Front and lateral step ups and down	Twice a day for 3-5 minutes initially.	Improves limb performance
	Sit to Stand	5 Reps × 2 set	Increase strength of hip extensor during functional task
	Resistance Exercises using Red Theraband	10 Reps × 2 set	Strengthens lower limb muscles
Phase 3-Advanced Strengthening and higher level function Phase	Ambulation Without Assistive device	Twice a day	Help to improve weight bearing and gain functional independence
	Ambulation on uneven surface	Twice a day	Helps to improve balance

Follow up and outcomes

Patient reported outcome after physiotherapy were improvement in right hip mobility after 3 weeks of post-operative physiotherapy rehabilitation and reduced pain in the hip. The other outcome measures used were NPRS, Goniometer and Harris Hip score (Table 6). Harris hip score has reliability of 0.94 that is good to excellent. Further follow up was required for Left side and sickle cell disease.

Table 6 Post Physiotherapy Rehabilitation

Sr. No	Outcome Measure		Pre Rehabilitation		Post Rehabilitation	
1	NPRS		8/10		3/ 10	
2	Joint	Movement	Right Active	Right Passive	Left Active	Left Passive
	Hip					
		Flexion	0-110⁰	0-120⁰	0-90⁰	0-110⁰
		Extension	0-30⁰	0-40⁰	0-10⁰	0-45⁰
		Abduction	0-50⁰	0-50⁰	0-30⁰	0-50⁰
		Adduction	0-40⁰	0-45⁰	0-35⁰	0-45⁰
		Internal Rotation	0-35⁰	0-40⁰	0-40⁰	0-45⁰
		External Rotation	0- 40⁰	0-40⁰	0-30⁰	0-50⁰
	Knee					
		Flexion	0-120⁰	0-130⁰	0-120⁰	0-125⁰
		Extension	120 -0⁰	130-0⁰	120-0⁰	125-0⁰
	Ankle					
		Dorsiflexion	0-30⁰	0-40⁰	0-30⁰	0-40⁰
		Plantarflexion	0-40⁰	0-45⁰	0-35⁰	0-45⁰
		Inversion	0-25⁰	0-35⁰	0-30⁰	0-40⁰
		Eversion	0-25⁰	0-35⁰	0-30⁰	0-35⁰
3	Harris Hip Score		27/100		63/100	

4. DISCUSSION

Avascular necrosis most commonly occurs in femoral head due to the precarious blood supply. According to the study on prevalence of avascular necrosis among sickle cell disease patient was 41% over unilateral hip (Adesina et al., 2017). In a on 215 sickle cell disease patients they concluded that considering the risk factors and symptoms early evaluation and management helps in improving the outcomes (Mukisi-Mukaza et al., 2000). The Challenges during managing this case were the complications occurring due to sickle cell disease. Improving the endurance was very difficult due to early fatigue. Production of a generalized protocol was not possible and can be considered as a limitation of the study. According to another study the Pre-operative physiotherapy was found to be effective as it helped to increase the patient compliance and easy understanding in post-operative (Ferrara et al., 2008). A study showed outcomes of hip replacement in ankylosing Spondylitis and AVN of femoral head and concluded that cemented THR is effective for both the conditions.

5. CONCLUSION

Through this case report we brought the light onto the importance of physiotherapy not only post operatively but pre-operative too. Multidisciplinary approach has found to be advantageous in reducing pain, gaining functional independence and improving strength in a patient with bilateral necrosis of femoral head secondary to sickle cell disease.

Author's contribution

NHS and MVJ conceptualized and took the case, MVJ and PAP implication of treatment. MVJ assisted in documenting the case. NHS wrote the manuscript. All the authors previewed and have approved the case report before submission.

Informed consent

Written & Oral informed consent was obtained from all individual participants included in the study. Additional informed consent was obtained from all individual participants for whom identifying information is included in this manuscript.

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Conflicts of interest

The authors declare that there are no conflicts of interests.

Data and materials availability

All data associated with this study are present in the paper.

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