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Client Centered Approach in the management of post-varicella purpura fulminans with multiple limb amputation in occupational therapy center –A case study

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Abstract
Background: *Purpura fulminans (PF) is a life-threatening emergency, in which there is skin necrosis and disseminated intravascular coagulation (DIC). A single case study was carried out by using client centered approach, in a 25-year-old, right hand dominant, shopkeeper by occupation, presented with chicken pox in February 2019. Later, patient complained of dark lesions appearing on left foot and spreading symmetrically and distally to all four limbs and was admitted to hospital in March 2019. The result of thrombophilia report showed heterozygous mutation of factor V Leiden. Patient was referred to occupational therapy department for rehabilitation of multiple amputation (Bilateral tarsometatarsal amputation, righthand 2nd,3rd, 4th, 5thtransmetacarpal amputation, left hand 2ndproximal phalanx mid-shaft amputation and 3rd, 4th, 5thtransmetacarpal amputation). Patient presented with complaints of pain in calf muscles while walking (claudication) along with tingling and numbness of lower limbs and difficulty in self-care activities.*
Materials and Methods*- The patient underwent acute inpatient rehabilitation in occupational therapy (OT) department from June 2019 to August 2019. It was a rare case referred to the OT department. He was assessed on Canadian Occupational Performance Measure (COPM), Six Minute Walk Test (6MWT) and Functional independence measure (FIM) scale. Occupational Therapy intervention included Buerger-Allen exercises to improve circulation and reduce tingling sensation in lower extremities. These exercises helped to improve functional mobility, whereas adaptive devices were given to improve self-care skills.*
Result*- Patient showed improvement from maximum assistance level to minimal assistance level inmost of his self-care and mobility skills with significant reduction in pain.*
Conclusion*-Client centered approach used by occupational therapist was effective in improving patient's performance as well as satisfaction level with functional independence and functional mobility.*
Keywords*: Purpura fulminans,Occupational therapy, Multiple limb amputation, Client centered approach.*

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I. Introduction

Purpura Fulminans (PF) is a rare but fatal thrombotic disorder. It is rapidly progressive syndrome characterized by intravascular thrombosis with necrosis of the skin and subcutaneous tissues along with disseminated intravascular coagulation (DIC). Purpura fulminans begins with erythema which develops into blue-black hemorrhagic necrosis in form of vesicles and bullae. It is a devastating illness, with mortality rates ranging from 50% to 90%. Symmetric involvement of all the extremities is typical of PF along with organ failureand up to 25% of patients with PF require at least partial amputation of all 4 extremities[1]. Buerger- Allen exercise is one of the interventions to stimulate the development of collateral circulation in the legs. It is a system of exercise for arterial insufficiency of lower limbs, consisting of 45-degree leg elevation, followed by 90-degree dependency of the legs, and finally 180-degree horizontal position of legs for rest. These exercises are to be administered for 7-11 minutes, two times a day for a week. One time comprises of five cycles of Buerger- Allen exercise.Thus, the use of Buerger-Allen exercise is effective in improving the foot perfusion by reducing the capillary refill time[2].Patients who have undergone lower limb amputation have usually spent a significant period of immobilization, deconditioning as well as decline in their independence. Rehabilitation following amputation must not only involve helping the patient to be physically independentbut also tobe functionallyindependent[3].To date, mobility has been the functional outcome most commonly examined in post-amputation patients.The surgical care and subsequent rehabilitation process after amputation seeks to

optimize and restore the physical and vocational outcome of the patient.Thus, in mobility-limited patient, the 6MWT would be a good measure of functional performance[4].The "client-centered approach" is an important aspect in the quality of rehabilitation processes which includes active participation of clients. It focuses on the concerns expressed by the client. It helps to identify changes in the client's personal perception of occupational performance over time[5].Since it was a rare case of an adult patient,the goal of occupational therapy was to return the patient to the highest level of independence and functional mobility.

II. Materials and Methods

This single case study was carried out atLokmanya Tilak Municipal Medical College and General Hospital, Sion, Maharashtra from June 2019 to August 2019

Study Design: Single Case Study

Study Location: This was a tertiary care hospital based study done in Department of occupational therapy, atLokmanya Tilak Municipal Medical College and General Hospital, Sion, Maharashtra.

Study Duration: June 2019 to August 2019

Methodology: A 25 year old, right handdominant, shopkeeper by occupation, presented with chicken pox in February 2019 which resolved within a week. Later, he complained of dark lesions gradually appearing on left foot which started spreading symmetrically and distally to all four limbs. Following which patient was admitted to tertiary care hospital in March 2019.Further, investigations were done and diagnosed as a case of acute infectious post varicellaPF. Multiple transfusions of blood and blood products were done for him. He underwent surgery in March 2019 with multiple amputation(Bilateral tarsometatarsal amputation,right hand 2^{nd}, 3^{rd}, 4^{th}, 5^{th} transmetacarpal amputation, left hand 2^{nd} proximal phalanx mid-shaft amputationand 3^{rd}, 4^{th}, 5^{th}transmetacarpal amputation) (Fig 1,3)followed by split thickness skin grafting (STSG) for bilateral lower limb.After acute care treatment, patient was referred to occupational therapy department for rehabilitation. On assessment,patient complained of inability to stand on his bilateral residual foot, phantom limb sensation, pain in calf muscles while walking (claudication) along with tingling and numbness of lower limb, neuroma over the stump and itching over the graft site. Patient had difficulty in self-care and ambulation. During rehabilitation phase, he was educated for residual limb care, oedema control, and desensitization techniques for lower extremities. Patient was assessed on FIM scale for functional independence, 6MWT for functional mobility and COPM. On COPM,patient's perspective was sought through the interview and occupational performance problems were defined by him. After the interview, his perceived performance on each of the identified tasks and his satisfaction with his own performance was rated by using a 0-10 scale. The patient identified total eight activities (Table no 1). Out of which five activities were identified as per the importance to him.

Table no1: Scoring of activity importance on COPM

PERFORMANCE DOMAIN	PROBLEMS IDENTIFIED	Importance
SELF CARE	Eating	8
	Bathing	7
	Dressing	5
PRODUCTIVITY	Standing at work place	10
	Walking	10
	Signing bills at work	8
Leisure	Use of phone to play games	5
	Use of remote control of TV unit	4

Table 1: Shows the problem areas identified by patient. All these areas were weighted as per their importance to the patient.

Table no 2: Therapeutic Intervention

Timeline	Clinical Findings	Therapeutic intervention
1ST WEEK –2NDWEEK	➢ **Self-confidence and self-image**- Poor. ➢ **Phantom limb sensation** present ➢ **Neuroma** present over the scar of amputed extremity. ➢ **Range of motion** -normal for Bilateral amputed extremities except left hand- Index finger metacarpophalangeal (MP)joint is 20-50 degrees ➢ **Muscle strength-** grade 3 (upper and lower limb muscles) ➢ **Hand function** Grip –poor (Difficulty in holding a pen for longer duration)Grasp-poor (Unable to open a bottle cap, holding glass of water) ➢ **Activities of daily living** - (FIM Score =83)*Eating -2,bathing -3,toileting-3,dressing-4,transfer-3,ambulation-2* ➢ **Standing Balance**- poor (Patient could achieve standing position for <5 minutes) ➢ **Walking –** Poor(On 6MWT- 10meters in 2 minutes)	➢ Counselling the relatives about patient's present condition. ➢ Strengthening exercises of upper and lower limb muscles. ➢ Sensory desensitization techniques over the scar of upper/lower limb. ➢ Skin management of recipient and donor site of grafting ➢ Gross and fine motor activities were started. ➢ Long handled scrubber and universal cuff to improve self-care skills. ➢ Standing on soft to hard surfaces initiated.
3RD WEEK – 4THWEEK	➢ **Phantom limb sensation**-Absent ➢ **Neuroma-** Absent ➢ **Range of motion**- Left hand Index finger MP joint is 20-70 degrees ➢ **Muscle strength-** grade 3+ (upper and lower limb muscles) ➢ **Hand function** Grip –Fair Grasp- Fair ➢ **Activities of daily living** - (FIM Score =99)*Eating-4,bathing -4,toileting-4,dressing-4,transfer-5,ambulation-5* ➢ **Standing Balance**- Fair (>10 minutes) ➢ **Walking –** Fair .Complains of claudication present while walking(6MWT- 50meters in 4minutes) ➢ **Climbing stair**- Poor (<5 flight of stairs - ascending and descending stairs)	➢ 1st and 2nd week intervention continued ➢ Training for donning and doffing of T-shirt and Elastic Pants were taught to the patient. ➢ Writing device ismade for dominant hand.(Fig 2) ➢ Buerger-Allen exercises were started to improve vascularity. ➢ Walking on soft to hard surfaces initiated. ➢ Modified footwear for indoor mobility.(Fig 4)
5TH WEEK – 6TH WEEK	➢ **Range of motion**- Left hand Index finger MP joint is 20-90 degrees ➢ **Muscle strength-** grade 4 (upper and lower limb muscles) ➢ **Hand function** Grip –Good ,Grasp- Good ➢ **Activities of daily living** - (FIM Score =105)*Eating-4,bathing -5,toileting-5dressing-5,transfer-5,ambulation-5* ➢ **Standing Balance**-Good(> 20minutes) ➢ **Walking –** Good(6MWT-150 meters with modified footwear) ➢ **Climbing stair**-Fair (>10 flight of stair) with modified footwear.	➢ 1st, 2nd, 3rd, 4th week intervention continued. ➢ Activities with writing device were initiated like dot game, drawing figures, writing alphabets and Names of family members, signing on the paper.
7TH WEEK – 8TH WEEK	➢ **Range of motion**- Left hand Index finger MP joint is 20-90 degrees ➢ **Activities of daily living** - (FIM Score =110)*Eating-4,bathing -5,toileting-*	➢ Continuation of above exercises. ➢ Recommendations- Filler shoes for outdoor mobility ➢ Patient referred for prosthetic hand.

	5dressing-5,transfer-6,ambulation-6 ➢ **Standing Balance**-Good(>30 minutes) ➢ **Walking –** Good (6MWT-300 meters with modified footwear) ➢ **Climbing stair**- Good (Independently ascend and descend stairs)	➢ Home modifications were advised.

Figure 1.Upper Limb AmputationFigure 2.Adaptive Device for writing

Figure 3.Lower limb amputation Figure 4.Modified Footwear

III. Result

Table no 3: Follow up and outcome measure**(**Pre and post intervention score for 6MWT, COPM and FIM)

SCALE	PRE-INTERVENTION SCORE (Baseline evaluation)	POST-INTERVENTION SCORE (After 2 months of follow-up)
6MWT	10 Meters	300 Meters
COPM		
Performance Level	2	7
Satisfaction Level	3	8

FIM SCALE	83	110

Table no 3: Shows pre and post intervention change in the score of 6MWT, COPM andFIM.

IV. Discussion

PF is life threatening disorder of acute onset characterized by cutaneous hemorrhage and necrosis.In this study, a case of acute infectious post varicella PF was referred for occupational therapy intervention. The patient was hospitalized for longer period for medical and surgical management, so he was showing most of the complications of immobilization. Due to multiple amputation and extensive skin and soft tissue lesions, patient was experiencing severe pain and was also showing psychological issues like feeling of loss, worried about severe disability and homesickness. All this was affecting patient's participation in exercise therapy during initial management of disease. Counseling helped to improve his self-confidence. Early use of desensitization techniques for hypersensitivity could have facilitated active participation of patient in rehabilitation program, thus improved self-image.Due to consistent stimulus to hypersensitive area of the stump throughout the day, showers the brain with sensory input. The brain responds to this demand by acclimating to the sensation, therebygradually decreasing the body's pain response to the particular stimuli. Thus, desensitization is beneficial as it has been shown to aid the reversal of cortical reorganization. It is also helpful in perceiving the affected limb in a more normal way[6].This also helped the patient to achieve standing posture comfortably. After the improvement in standing tolerance, patient was fitted with modified footwear with elastomeric padding material. In the present study, pre-intervention patient could walk for < 5 minutes at a time. PF impairs peripheral oxygen extraction that limits the availability of oxygen to lower extremity skeletal muscle during the 6MWT. Due to which patient could hardly walk for a shorter distance.During fast walking, the most metabolically active tissue is skeletal muscle especially within the legs[7].Whereas post-interventionhe could walk > 20 minutes at a stretch. This could have been due to Buerger-Allenexercises which improved the circulation in lower limbs with reduction of claudication. The mechanism of Buerger- Allen exercises uses gravitational changes in positions that are applied to the smooth musculature of vessels and to the vascular. Gravity helps alternately to empty and fill blood columns, which can eventually increase transportation of blood through them, thus improving the lower limb circulation.Also modified footwear could have reduced shear and friction during walking and so the difference was observed on 6MWT from Pre10meters to Post300 metersintervention.This finding is supported by a study (2015), thatreported the effect of Buerger-Allen exercise has significantly improved blood flow and walking ability[8].

Since it was a rare case it needed unique care. Therefore, COPM was administered through which patient's problem areas could be identified and occupational therapy program could be planned accordingly.In this study, we observed difference in performance levels from score 2 to score 7 and satisfaction levels from score of 3 to score of 8respectively. For self-care, patient was given various adaptive devices which had facilitated the use of residual limb of upper extremity. He was able to write by using writing device. Thus, regular training along with motivating thepatient consistently for the use of prescribed devices in his day to day activities has showed improvement in his performance and satisfaction level.One of the case series (2009) reported that the use of customized universal cuff fitted to forearm of the residual limb, improved independence in feeding and grooming.Also, two patients of this case series with lower limb amputation could ambulate more than 150 feet and required minimal assistance with stair climbing[1].As there was overall improvement observed in performance as well as satisfaction level for identified activities, which ultimately laid to change in functional independence from 83 to 110 intervention. This patient gained independence in his ADL activities and functional mobility within short period of time. It could have been due to his younger age and early rehabilitation program post amputation. This goes in accordance with the study by U.R. Madsen et al (2018), that short-term functional outcome (independence in ADL) was positively associated with lower age and rehabilitation initiated after discharge[9].Unfortunately, there is a paucity of literature that addresses quality of life for patients with PF, and the limited evidence of functional independence in these cases. So the treatment to improve peripheral circulation and appropriate adaptation remains the only source for planning rehabilitation program.

V. Conclusion

PF is an uncommon but severe and life-threatening thrombotic disorder that needs extensive rehabilitation. Client centered approach was found effective, as it weighted the important problem areas faced

by the client and rehabilitation program was planned accordingly. Buerger- Allen exercises improved peripheral circulation which helped to improve functional mobility. Thus, in this case the use of adaptive devices appeared to have impact on increased functional independence with better performance and satisfactionlevel.

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