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Intensive Treatment of Elephantiasis in A Child: A Case Report

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Abstract

Introduction: There are few publications on the treatment of elephantiasis in children. Objective: The aim of this study is to report on the intensive treatment of lower limb elephantiasis of a child in an outpatient setting.

Case report: We report the case of an eight-year-old boy who was born with scrotum and lower limb lymphedema. The initial diagnostic difficulty and therapeutic conduct over the years did not produce results and there were complications such as erysipelas. The lymphedema progressed to the most advanced stage, elephantiasis (stage III), with the mobility of the child limited affecting the quality of his life, including academically. A five-day intensive outpatient treatment program was proposed including Cervical Stimulation Therapy, Manual Lymphatic Therapy, Mechanical Lymphatic Therapy and compression using a custom-made stocking made of a grosgrain fabric.

Results: There was a reduction of 45% of the limb volume in five days.

Conclusion: Intensive treatment of elephantiasis in children is a new therapeutic option that allows significant reductions in volume in a short time.

Keywords: Elephantiasis; Lymphedema; Intensive treatment; Diagnosis; Lymph drainage

Introduction

Lymphedema is defined as a disorder of the lymphatic system characterized by an imbalance between the formation and absorption of lymph with an abnormal accumulation in the tissue of fluid rich in proteins and macromolecules [1]. Because of this lymphatic dysfunction, the formation of edema is progressive and can evolve with fibrosis and major deformities. Lymphedema is classified as primary or secondary, with primary being basically idiopathic or due to congenital causes, and secondary being acquired with the dysfunction of the lymphatic system owing to infections, cancer or medical interventions used to treat cancer such as lymphadenectomy and radiotherapy [1-5].

It is estimated that the worldwide prevalence of lymphedema is between 0.13 and 2%, with the highest prevalence related to secondary lymphedema in developing countries and in areas endemic for *Wuchereria bancrofti*, the major cause of filariasis [2]. The prevalence of lymphedema in children is low, approximately 1.15:100,000 in under 20-year olds [3]. There are no specific guidelines for the treatment of children, even though some characteristics are different to the disease in adults such as the higher prevalence of primary lymphedema and the possibility of the involvement of more than one limb [4].

During the clinical course of lymphedema an accumulation of proteins in the interstitial space and inflammatory cells as well as adipose hypertrophy and the presence of fibrosis can be observed; the latter condition results in less pitting under finger pressure [3]. Clinical staging of lymphedema assesses the consistency of the skin (degree of subcutaneous fibrosis) and the amount of reduction of lymphedema with the affected limb raised for 24 hours. Stage I is defined by a smooth, soft swelling without fibrosis which subsides after elevating the limb for 24 hours and evident indentation (pitting) on applying

finger pressure. Stage II lymphedema is characterized by a certain amount of fibrosis however finger pressure still causes an indentation in the skin, but the edema does not disappear completely by elevating the limb for 24 hours. Finally, stage III, or elephantiasis, has a high degree of subcutaneous fibrosis, the skin is hard, no pitting is seen under finger pressure and ulcerations, fat deposit, acanthosis and even warty skin lesions are observed [2].

There is currently no specific treatment for children. Therapeutic options consist of skin care, manual lymph drainage, control of infections, and myolymphokinetic exercises and activities associated with containment mechanisms such as compression stockings and elastic or non-elastic bandages. In addition to physical therapy, proper guidance on the treatment and parent conduct is necessary, including stimulation to participate in normal activities and psychological support to improve treatment adherence and outcome [4]. The aim of this study is to report the intensive treatment in an outpatient setting of a child with lower limb elephantiasis.

Case Presentation

We report the case of an eight-year-old male child, whose mother described the presence of scrotal edema from birth at which time she sought medical assistance. Pediatricians told her to watch and wait as the condition might resolve spontaneously. After five days, as the infant's left leg had swelled from the foot to the knee, the mother and child returned to the health center where the physicians hypothesized a possible neurological disorder. The mother continued her quest with evaluations by an orthopedist and angiologist, who diagnosed a chronic disorder, and referred the child to a specialist in edema. The mother did not consult the specialist, and one year later the infant was prescribed an elastic stocking during a pediatric consultation; the patient used an elastic stocking for two years without any problems. After that and coinciding with the first outbreak of erysipelas due to an injury to the upper foot, the child was sent for manual lymph drainage. Repetitive episodes of infection followed, and the child needed hospitalization for treatment. With the progression of the disease the patient refused to use elastic stockings and wart-like lesions developed on the foot; the edema spread to the entire left leg, scrotum and penis. Despite treatment with benzetacil every 21 days he suffered further episodes of erysipelas.

The mother sought the Clínica Godoy, where stage III (elephantiasis) congenital lymphedema was diagnosed. The patient had severe lymphedema of the left leg, foreskin and scrotum with painful and limiting elephantiasis verrucosa in the plantar region.

The leg was evaluated by volumetry and bioimpedance; the left leg measured 7804 mL below the knee and the contralateral limb measured 2441 mL giving a difference of 5363 mL. By bioimpedance (Inbody S10[®]) the left leg had 8220 mL of liquid and the right leg 2920 mL, a difference of 5300 mL. Intensive treatment was performed for five days in an

outpatient setting using a grosgrain stocking, Mechanical Lymphatic Therapy (RAGodoy[®]) and Cervical Lymphatic Stimulation (Godoy and Godoy technique), together with scrotal and penile compression bandages made of grosgrain. After intensive treatment, the left leg measured 5866 mL by volumetry, a reduction of 2440 mL (45.5%) and a reduction of 46.3% was measured by bioimpedance. Moreover, there was a significant reduction in the edema of the foreskin and scrotum. The child returned to school but must return to the clinic during his vacations for the next phases of treatment until the edema is reduced totally. The study was approved Ethical Committee in Medicine School of São Jose do Rio Preto-Brazil number# CAAE: 54631416.4.0000.5415/2016 and was child familiar responsible signed consent form.

Discussion

This study reports the case of the most advanced stage of lymphedema (stage III - elephantiasis) in an eight-year-old boy. This case report proposes a new form of treatment for children with lymphedema with eight hours per day of intensive treatment in an outpatient setting. This approach in adults can reduce the lymphedema by an average of 50% within five days; in the current case, the reduction was 45% in five days [6]. There are no reports in the literature describing this type of approach in children.

The difficulty in diagnosis and therapy of this child shows that even after diagnosis the disease is able to evolve to the most advanced stage of lymphedema. These difficulties are commonly observed in the clinical practice even in adults and relatives do not always receive adequate guidance. It is important to stress that treatment should be started as early as possible in order to prevent progression to a more advanced stage of the disease.

The therapeutic strategy of this child significantly reduced the edema within one week, and in this time the mother received counselling on how to maintain this reduction by treating at home and about adapting treatment to the child's academic life. Treatment can be carried out without affecting the child's education as intensive treatment can be performed during school vacations. In this case treatment had a positive effect on the psychosocial life of the child, reducing the physical limitations, raising his self-esteem and reinserting him into the school environment.

Intensive treatment includes about 8 hours per day of Mechanical Lymphatic Therapy. This uses an electromechanical apparatus, the RAGodoy[®] device, which passively performs ankle flexion and extension movements [6]. The most important compression mechanism used is a custom-made grosgrain stocking, which has inelastic features [7]. Another type of therapy used is Cervical Lymphatic Stimulation, a new concept of lymph drainage [3]. The manual lymph drainage technique used was created by Godoy and Godoy and consists of compression exerted by the hand using linear movements along the path of the lymphatic vessels [8].

The literature does not describe a specific form of treatment for children and so adult treatments are often adapted.

However, children have a different pace of life and do not always adhere to the conditions of therapy. In this case, the limitation of this child stimulated the care team to find the best approach; an adaptation of therapies is required for each case.

The proposal of the therapy is to normalize the edema, as generally occurs in minor clinical stages of lymphedema. The rapid reduction in this case allowed the child to return to his academic and social life. Psychological and family support is essential as it helps in the treatment.

Conclusion

Intensive treatment of lymphedema is a new therapeutic option for children with elephantiasis as it allows significant reductions in edema in a short time.

Competing Interests

The authors declare that they have no competing interests (political, personal, religious, ideological, academic, intellectual, commercial or any other) in relation to this manuscript.

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