

Physical therapy of far-advanced primary lower limb lymphedema: a case report and literature review

Fizjoterapia w dalece zaawansowanym pierwotnym obrzęku chłonnyim kończyny dolnej: opis przypadku i przegląd literatury

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Abstract

Physical therapy with good compliance remains the key of success in lymphedema management. Tissue excisions should be reserved for physiotherapy-resistant cases and always followed by extended maintenance compression therapy. We present the case of the successful physical therapy of the young female with a neglected, massive, primary lymphedema of the lower extremity present since birth, with prior extensive tissue resection. Complex decongestive physical therapy has been performed for one year, combined with ten months' of negative-pressure wound therapy at a hospice facility due to pressure ulcers complication. Obtained reduction of the affected extremity/edema volumes (by 69.4 and 88.2% respectively) enhanced patient's mobility, helping her become more independent. Ongoing therapeutical program maintained these results for three years. Physical therapy even in far-advanced and neglected cases may be effective. Lymphedema services should be offered widely through a network of fully reimbursed health care system, delivering high-quality evidence-based management tailored to individual patients.

Key words: primary lymphedema, palliative care, physical therapy.

Streszczenie

Kluczem do sukcesu w leczeniu obrzęku limfatycznego pozostaje fizjoterapia i stosowanie się do zaleceń terapeutów. Leczenie operacyjne powinno być zarezerwowane dla chorych niepoddających się fizjoterapii i stosowane zawsze w połączeniu z kompresjoterapią podtrzymującą. Prezentujemy przypadek skutecznego leczenia fizjoterapeutycznego u młodej kobiety z zaniedbanym, masywnym, pierwotnym obrzękiem limfatycznym kończyny dolnej, uprzednio poddanej rozległej resekcji tkanek. Trwająca rok kompleksowa fizjoterapia powikłana odleżynami wymagała przyjęcia do hospicjum i stosowania przez 10 miesięcy terapii podciśnieniowej. Uzyskana redukcja objętości zajętej kończyny i obrzęku odpowiednio o 69,4 i 88,2% zwiększyła możliwości ruchowe chorej, poprawiając jej zdolność do samoobsługi. Wdrożony program terapeutyczny pozwolił na utrzymanie efektów terapii przez kolejne 3 lata. Nawet w dalece zaawansowanych i zaniedbanych obrzękach kompleksowa fizjoterapia może okazać się skuteczna. Opieka nad chorymi z obrzękiem wymaga istnienia sieci ośrodków oferujących wysokiej jakości terapie o udowodnionej naukowo skuteczności, w pełni refundowane i dostosowane do indywidualnych potrzeb.

Słowa kluczowe: pierwotny obrzęk limfatyczny, opieka paliatywna, fizjoterapia.

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INTRODUCTION

The frequency of usually asymmetrical, primary lymphedema at birth is estimated to be about 1 per 10000, three times more often in females [1, 2]. De-

creasing quality of life it also leads to significant disability [3, 4]. This condition can be managed safely by a combination of Complex Decongestive Therapy (CDT) elements [5, 6]. The treatment is more effective when started early; as the condition advances, its

efficacy narrows down to the maintenance of the actual state of lymphedema and the prevention of progression [7]. Drug therapy has not proved itself to be particularly beneficial, and surgical interventions are reserved for extreme cases [8]. Patients must always take an active role in controlling their disease by means of secondary prophylaxis and compression. The case of the prolonged, while successful conservative management of edema in a 17-year-old female with a massive, neglected by the health system for years, primary lymphedema of the lower extremity presented at birth, without prior conservative treatment is presented.

CASE PRESENTATION

A 17-year-old woman with moderate mental retardation, encephalopathy and epilepsy was referred to the lymphedema clinic with massive (3rd stage by International Lymphology Society) primary leg lymphedema, complicated by skin ulceration that recurred several months after radical tissue Sistrunk's resection (performed at the age of 14). Her edema presented at birth (suspected of Klippel-Trénaunay Syndrome since the age of 9) had never been treated conservatively before, except for a year of compression stocking following the surgery. She was hospitalized three times due to foot and calf inflammatory skin episodes with empirical antibiotic therapy introduced each time without additional physiotherapy in spite of progressive limb swelling. Patient agreed for the proposed conservative physical therapy and consent for the publication was signed by her father.

On admission the patient was in poor condition with moderate left leg pressing pain. Her left lower extremity had positive Stemmer's sign, the calf had a hard consistency with *peau d'orange* dermatolymphangioadenitis, while the skin on the thigh area was intact and pitting. Large skin folds in the ankle region which rubbed the ground and limb heaviness impairing her mobility, especially on walking, problems with finding appropriate clothes and footwear were particularly problematic. The affected and

healthy extremities volumes assessed on the basis of circumferences at 4 cm intervals, using simplified frustum formula [9], were 27.1 L, and 5.4 L respectively (403.5% of excess limb volume).

The treatment plan was consistent of selected elements of CDT – multilayer short-stretch bandaging (SSB) with dedicated aerobic physical exercises was chosen as the principal method of treatment. She was not qualified for manual lymph drainage due to poor compliance, and the limb size made intermittent pneumatic compression impossible in this case. Over a period of four weeks, the SSB was applied ambulatory for 23 hours a day five times a week (Saturday and Sunday – 48 hours in bandages). Within two weeks the additional foam pads were applied underneath the bandage on calf to enhance skin relaxation. This SSB has been continued at home for the next four weeks by the educated family, showing edema increase and extensive infected skin erosions in the heel and popliteal region. For this reason she was admitted to the hospice for 44 weeks and re-banded twice a day (over 22 weeks) combined with the intensive aerobic training (two 30-minute sessions). Two small skin erosions healed in 5 weeks with octenidine, standardized dialysate from young calf's blood topical cream, ionic silver and hydrocolloid dressings. The third, most profound ulcer in the pretibial region (3,4 × 6,0 × 1,0 cm in size), MRSA infected, required the combination of SSB and continuous negative pressure wound therapy (NPWT) with nanocrystalline silver dressings for an additional 22 weeks. Finally after achieving a steady improvement in limb volume reduction she was discharged and flat knitted hosiery (compression class 3, 34-46 mm Hg, German RAL standard) was applied with a recommendation for use during the day (2-3 times a week) interchangeably with SSB. The results maintained through the next 28 weeks (Table 1). The patient's body weight decreased by 9 kg as a result of compression therapy combined with dietary and mobility-enhancing exercises; the patient's ability to ambulate and transfer improved, helping her become more independent in the performance of daily routines (Fig. 1).

Table 1. The results obtained during the successive phases of treatment, including body weight

Treatment phase	Management duration (weeks)	ALV (L)	ULV (L)	Weight (kg)	EV (L)	EV (%)
before admission		27.1	5.4	71.5	21.8	403.5
ambulatory intensive	4	13.1	5.4	68.0	7.7	142.0
I maintenance at home	4	15.9	5.2	64.7	10.7	206.3
hospice intensive	44	9.0	5.4	64.0	3.5	64.5
II maintenance at home	128	8.8	5.5	76.2	3.3	58.4

ALV – affected limb volume; ULV – unaffected limb volume; EV – edema volume



Fig. 1. Patients' legs on admission to the lymphedema clinic (A) and after three years of conservative therapy (B). The linear scar and padding shape of the left foot distally to the previous tissue resection is still visible (B)

DISCUSSION

The management of primary lymphedema has remained a challenge for both patients and medical professionals. When left unattended, it results in lymph stasis, the steady progression of the disease and due to recurrent inflammatory episodes, in far-advanced cases, it can lead to life-threatening complications like sepsis [10, 11]. In our patient no conservative management was introduced over the years, as the girth of left lower limb increased and she made numerous medical appointments. Compression treatment was not recommended even in the aftermath of the recurrent episodes of dermatolymphangioadenitis, which required antibiotic therapy. A traumatic surgical procedure in a far-advanced state was performed instead, still without subsequent compression, complicated by skin infection and necrotic ulceration that demanded reoperation. The various types of staged subcutaneous excisions have been performed since 1911 (by O. Lanz, followed by E. Kondoleon, W. Sistrunk and J. Homans) with satisfactory clinical results and improvement of lymphoscintigraphy clearance at 1 year afterwards [12]. All invasive techniques, including less traumatic liposuction nonetheless should be limited to end stage disease irresponsive to CDT or steady progression despite the maximum involvement of therapy over two years [13, 14]. It may play an auxiliary role in supplementing unsuccessful physiotherapeutic management, although it is a long-term postoperative commitment to compression that offers the key to success [15].

The management of choice in this incurable disease is CDT, based on limb compression, manual lymphatic drainage, exercises with the affected limb in compression, and proper skin care. Approximately 95% of patients in multidisciplinary lymphedema clinics are able to be managed conservatively without operative intervention, significantly reducing infections recurrence [16, 17]. Continuous compression combined with physical exercises is considered to be the most efficient component of CDT [18]. The additional effect of manual lymphatic drainage still needs to be confirmed by randomized data [19]. In more advanced pitting edema, compression bandaging should be applied first to prepare the limb for compression garments [20]. In most cases of lymphedema, the greatest reduction in volume occurs during the first 2 weeks of therapy, but in more advanced fibrotic cases the improvement can appear later [21]. In the presented case the spectacular initial effect obtained at the expense of decubitus ulcers within the area of former skin incision required protracted in-patient care. When treating wounds of mixed etiology, the aim is to achieve a balance between safety and efficacy [22]. Skin damage due to compression or shearing stress is an uncommon complication, observed more frequently on the sensitive skin of children, but intensive compression bandaging always needs individualized risk assessment. Chronic wounds appearing in the region of lymphatic insufficiency and vascular iatrogenic malformations require a combination of topical wound care therapies, including limb compression and general pharmacotherapy – often including antibiotics. This therapy

usually demands bed rest and strict inpatient supervision. Prolonged hospitalization generates high costs of attending to such patients. In this case the critical colonization and infection of the wounds in the region of poor perfusion and compromised immune system activity delayed healing, bringing about increased pain and discomfort. Observations suggest that cryptic bacteria present in lower limb tissues in lymphedema or vascular insufficiency may lead to empiric antibiotic therapy failure [23]. For this reason, silver-containing antimicrobial dressings with proven broad spectrum antimicrobial activity including against antibiotic-resistant bacteria were used with only partial effect of wound stabilization [24]. Complete healing was observed after 5 months of NPWT combined with compression bandaging of the limb [25]. A parallel limb volume improvement was observed, concurrent with the patient's enhanced ability to perform her daily routine. Patient's weight also decreased, which possibly enhanced lymphatic flow, reversing some lymph insufficiency [26]. Within the last maintenance phase at home, the patient achieved further improvement in ability to perform all work-related activities, returning to the community. This required ongoing management, highlighting the importance of improving treatment adherence [27]. An excellent compliance with management was observed through all phases, also due to substantial engagement of her family. Non-adherence to treatment is considered to be the most important modifiable factor compromising treatment effectiveness of chronic conditions.

The costs of compression products, lack of sufficient reimbursement and also discomfort associated with SSB, are considered unacceptable to a significant number of patients thus leading to experimenting with a wide range of sometimes risky complementary therapies. Physical therapy of lymphedema still seeks the proper place in Polish health system [28]. Traditionally lymphedema clinics came in early ninety's of the last century from the grassroots of hospice movement, reducing cancer patients' unmet needs. At present, besides scarce lymphedema clinics following-up patients, ambulatory rehabilitation consists of a few days sessions without intention to further monitoring. Hospital conservative management receives insufficient reimbursement which usually covers only up to one week. Effective lymphedema treatment may reduce costs of acute hospitalizations due to its consequences, including cellulitis [29]. Treatment results are optimal when lymphoedema is diagnosed and treated early, but effective management in advanced stages also can improve the outcomes, improving quality of life, reducing patients' disability. However patients must have better access to both out and in-patient care

which are sufficiently refunded by the health care system.

CONCLUSIONS

Life-long program of compression therapy with good compliance remains the key of success in primary lymphedema even in far advanced and neglected cases.

Surgical excisions should be reserved for physiotherapy-resistant cases and always followed by extended maintenance physiotherapy.

Lymphedema services should be offered widely through a network of fully reimbursed health care system, delivering high-quality standardized management tailored to individual patients

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