Rehabilitation of the patient after total resection of the femoral bone as part of the Ewing's sarcoma treatment

(Usprawnianie pacjenta po zabiegu totalnej resekcji kości udowej w wyniku przebytego leczenia mięsaka Ewinga)

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Abstract – Introduction. Ewing's sarcoma is a small-round-cell tumour, the second tumour (after osteosarcoma) as to the prevalence and the most malignant primary tumour of bone in children and adolescents. As much as 90% of the incidence occurs in people between 5 and 25 years of age, and in the population below 20 years of age the frequency is below 3 persons / 1,000,000.

The aim of the study. The objective of the thesis is to present the course and results of rehabilitation of a 45-year-old patient after total resection of the femoral bone as part of the Ewing's sarcoma treatment.

Case report. In 1996, a patient with diagnosed Ewing's sarcoma underwent a resection arthroplasty of hip joint with allograft implantation. In 2016, due to pathological fracture of the femoral bone and loosening of the femoral part of the prosthesis in the presented patient, total femoral bone resection with alloplasty of the knee joint was performed. The rehabilitation process after the surgery lasted for two years and included intensive physiotherapy, which allowed to achieve the goals assumed in the individual treatment plan. Thanks to surgery and rehabilitation, pain relief and improvement of the right lower extremity function were achieved, which resulted in increased gait efficiency. Due to the persistent weakness of the right lower extremity, continuation of rehabilitation in subsequent years is recommended.

Conclusions. The presented case shows that physiotherapy after total resection of the femoral bone allows you to regain lost functions.

Key words - Ewing's sarcoma, resection arthroplasty, rehabilitation.

Streszczenie – Wstęp. Mięsak Ewinga (ME) jest guzem drobnookrągłokomórkowym, drugim nowotworem (po mięsaku kościopochodnym) co do częstości występowania oraz najbardziej złośliwym guzem pierwotnym kości u dzieci i młodzieży. Aż 90% zachorowań występuje u osób między 5. a 25. rokiem życia, w populacji poniżej 20. roku życia występuje z częstotliwością poniżej 3 osoby /1 000 000.

Cel pracy. Celem pracy jest przedstawienie przebiegu i wyników usprawniania 45. letniego pacjenta po zabiegu totalnej resekcji kości udowej w wyniku przebytego leczenia mięsaka Ewinga.

Opis przypadku. W 1996 roku u pacjenta z rozpoznanym mięsakiem Ewinga wykonano zabieg resekcyjnej endoprotezoplastyki stawu biodrowego z implantacją allograftu. W 2016 z powodu złamania patologicznego kości udowej i obluzowania elementu udowego protezy u przedstawionego pacjenta przeprowadzono totalną resekcję kości udowej z alloplastyką stawu kolanowego. Proces usprawniania po zabiegu trwał dwa lata i obejmował intensywną fizjoterapię, która pozwoliła na osiągnięcie celów zakładanych w indywidualnym planie terapii. Dzięki operacji i rehabilitacji uzyskano zmniejszenie dolegliwości bólowych i poprawę funkcji kończyny dolnej prawej, co przełożyło się na zwiększenie wydolności chodu. Ze względu na utrzymujące się osłabienie kończyny dolnej prawej wskazana jest kontynuacja rehabilitacji w kolejnych latach.

Wnioski. Przedstawiony przypadek pokazuje, że fizjoterapia po zabiegu totalnej resekcji kości udowej pozwala na odzyskanie utraconych funkcji.

Slowa kluczowe – mięsak Ewinga, resekcyjna endoprotezoplastyka, rehabilitacja.

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I. INTRODUCTION

wing's sarcoma is a small-round-cell tumour, the second tumour (after osteosarcoma) as to the prevalence and the most malignant primary tumour of bone in children and adolescents. As much as 90% of the incidence occurs in people between 5 and 25 years of age, and in the population below 20 years of age the frequency is below 3 persons/ 1,000,000. [1]. The peak of morbidity is closely related to the period of the highest bone structure growth and in boys it falls between the ages of 10 and 14, whereas in girls between 5 and 9 years of age [2]. Almost only representatives of the white race fall ill (Ewing's sarcoma very rarely occurs in the African or Asian population), more often men than women (the incidence ratio is 1.5: 1).

In Poland, the incidence is estimated at about 40 cases per year [2,3,4]. Ewing's sarcoma develops in the body of the bones, most often in long bones (60%), of which 22% are femoral bones [1,2,4].

In 25-30% of patients, at the time of diagnosis, metastases are found, mainly to the lungs, bones, lymph nodes and bone marrow. [1,3–5].

The most common symptom of cancer is pain, which intensifies during the night hours and often limits the range of movement. In 40% of patients, there are also symptoms of local and generalised inflammation [1,3,4].

Ewing's sarcoma treatment is combined and includes chemotherapy (CHT), surgery and radiotherapy (RT). The basic form of treatment is multi-drug chemotherapy. It is used before surgery (so-called induction, pre-treatment, neoadjuvant or pre-operative) and after surgery (so-called adjuvant or postoperative). The combination with radical irradiation of the primary lesion gives a chance of 5 years of living without symptoms of malignant disease in 50-55% of sick children and in about 40% of adult patients [5,6,7]. Ewing's sarcoma belongs to radiosensitive cancers, i.e. the ones susceptible to treatment with ionising radiation. [1,3,5,6].

The partial or radical removal of the malignant bone is part of the surgical treatment. However, this treatment should be combined with chemotherapy and often with complementary radiotherapy [4]. Limb sparing treatments are currently performed in over 85% of patients with a good response to initial treatment. They consist in removing the entire tumour along with the surrounding tissues and bone. The loss of bone is replaced by a bone graft, in some cases it is so large that it becomes necessary to replace the entire joint with the endoprosthesis. In some patients, amputation of the entire limb with a tumour is necessary [4,9].

The possibilities of reconstruction after removing the femoral tumour assume the filling of the bone defect with the use of autograft, allograft, rotational plastic and an individually made modular prosthesis [10,11,12]. High frequency of fractures, lack of adhesions and infections make reconstruction using allografts not beneficial for the patients. [11]

The most radical form of arthroplasty is total resection of the femoral bone with concurrent alloplasty of the hip and/or knee joint. This operation may be an alternative to amputation with enucleation in the hip joint due to a large bone loss as a result of the excision of a large femoral tumour, in the case of aseptic loosening of a previously implanted knee or hip joint prosthesis, infection or the periprosthetic fracture [13].

Factors that worsen the prognosis in the course of Ewing's sarcoma include: the presence of remote metastases, the size of the primary tumour above 5 cm, the placement of Ewing's sarcoma in the pelvic bones and chest bones, infiltration of soft tissues. [1,4,6,17–19].

The objective of the thesis is to present the course and results of rehabilitation of a patient after total resection of the femoral bone as part of the Ewing's sarcoma treatment.

II. CASE REPORT

A 45-year-old patient, who in 1995 was diagnosed with an Ewing's tumor located in the right thigh. The tumour covered ½ proximal femoral bone including the neck. The tumour infiltrated adjacent soft tissues. After the induction chemotherapy, the patient underwent resection hip replacement with allograft implantation. Surgical treatment was supplemented with postoperative chemotherapy.

In the following years, the patient without the recurrence of malignant disease.

After performing the hip arthroplasty, the length of the lower extremities in the patient was equal, but as a result of progressing changes in the femoral bone, the length of the right extremity decreased in relation to the left one. The progressive shortening, which in 2016 amounted to 5 cm, impaired the gait of the patient and contributed to the appearance of pain in the thoracic and lumbar spine. (Figure 1).



Figure 1. Difference in the length of lower extremities (2014) (own material)

Since 2009, the patient complained about pain in the area of the right knee joint, which appeared after prolonged walking. He also had the feeling of instability of this joint. These symptoms intensified in 2015. Based on the X-ray picture, a pathological fracture of the femoral bone and loosening of the femoral part of the prosthesis were found, which qualified the patient for reoperation (Figure 2, Figure 3).



Figure 2. Loosening of the femoral part of the prosthesis (own material)



Figure 3. Loosening of the femoral part of the prosthesis, image during knee bending (own material)

In September 2016, the patient underwent total resection of the femoral bone and implantation of the prosthesis together with the knee replacement (Figure 4, Figure 5, Figure 6). After two weeks, the patient was discharged from hospital with recommendations for walking with the assistance of 2 elbow crutches, with partial loading of the operated lower right extremity, with contact of the foot with the ground and recommendations for performing exercises strengthening the gluteus muscles, quadriceps and exercises improving the range of motion in the hip joint. The patient was banned from taking a full bending position, performing the retraction movements and internal rotation of the operated limb due to the risk of prosthetic dislocation.



Figure 4. Hip joint endoprosthesis (own material)



igure 5. Knee joint endoprosthesis with femoral implant (own material)



Figure 6. Knee joint endoprosthesis with femoral component (own material)

Description of the rehabilitation process

One and a half months after total resection of the femoral bone with the knee joint alloplasty, the patient started a 6-week physiotherapy in the day rehabilitation ward. Before the rehabilitation, the patient moved with the assistance of two elbow crutches. After establishing therapeutic goals, physiotherapy was initiated, which included the following procedures: exercises to strengthen the muscles of the right lower extremity, proprioception and increasing the range of mobility in the right and left knee joints, mobilisation of postoperative scar and learning the locomotor functions. After the completion of rehabilitation, the range of movements and strength of the right lower limb muscles was improved, which allowed walking with the assistance of only 1 crutch. In the following months, the patient participated in physiotherapy as part of the stationary rehabilitation department (for 4 weeks) and then in the day rehabilitation ward (for a period of 6 weeks). The frequency and intensity of physiotherapy during the first year after surgery was high, which positively affected the patient's efficiency. Physiotherapy cycles in the first year after total resection of the femoral bone with alloplasty of the knee joint are presented in Table 1.

Table 1. Physiotherapy of a patient after total resection of the femoral bone with hip and knee alloplasty in the first year after surgery

Forms of rehabi- litation	The cycle duration	The goals of phy- siotherapy	Physiotherapeutic procedures
Daily rehabilita- tion ward	6 weeks	 improvement of hip and knee ROM improvement of proprioception strengthening the muscles of the lower extremities enhancing the elastic- ity of tissues in the area of postoperative scar reduction of inflam- mation and pain 	 active and passive exercises assisted exercises isometric exercises individual exer- cises according to the PNF method classic massage mobilisation of postoperative scar whirlpool bath cryotherapy
Stationary reha- bilitation ward for the locomo- tor system	4 weeks	 strengthening the muscles of the lower extremities and the trunk improvement of the efficiency of the glute- us medius muscle gait re-education, improvement of the load phase of the right lower extremity enhancing the elastic- ity of tissues in the area of postoperative scar reduction of inflam- mation and pain 	 individual exer- cises according to the PNF method assisted exercises active exercises classic massage laser therapy magnetotherapy
Daily rehabilita- tion ward	6 weeks	 strengthening the muscles of the lower extremities and the trunk improvement of the efficiency of the glute- us medius muscle gait re-education, improvement of the load phase of the right lower extremity reduction of pain 	- isometric exercis- es - slow active exer- cises - individual exer- cises according to the PNF method - acquiring locomo- tion abilities - cryotherapy

Due to the alignment of the extremities, the efficiency and aesthetics of walking improved. During about 6 months after the surgery, the patient moved alone without crutches. The patient's gait remained slightly limping, which is why in the second year after the surgery, the main goal was to work on particular stages of gait and pelvic stabilisation. The rehabilitation process in the second year is presented in Table 2. Table 2. The process of rehabilitating the patient after totalresection of the femoral bone with knee alloplasty in thesecond year after surgery

Forms of rehabili- tation	The cycle duration	The goals of phy- siotherapy	Physiotherapeutic procedures
Daily rehabilita- tion ward	6 weeks	 learning how to properly load the lower extremities during individual stages of gait improving the stability of trunk and pelvis during gait maintaining the correct range of hip and knee ROM reduction of pain in the lumbar and thoracic spine 	 acquiring locomo- tion abilities individual exercises according to the PNF method assisted exercises manual therapy local cryotherapy
Physiotherapy as part of a project co-financed from the European Union budget	6 weeks	 improvement of the gait dynamics balance training prophylaxis of pain in the lumbar and thoracic spine 	 acquiring locomo- tion abilities individual exercises according to the PNF method equivalent exercises manual therapy local cryotheapy
Outpatient phy- siotherapy	2 weeks	 improvement of the gait dynamics balance training prophylaxis of pain in the lumbar and thoracic spine 	 acquiring locomo- tion abilities equivalent exercises manual therapy

The shortened Harris scale, which examines the hip function, was used to assess the progress of the rehabilitation process. Harris Hip Score contains questions about pain, weakness of the extremity, gait, orthopaedic supplies, as well as the activities of putting on shoes and the function of sitting. The maximum number of points that a patient can get in all ranges is 100. The examination was performed before the surgery of total resection of femoral bone and after two years of rehabilitation. In the final examination, after the two-year improvement period, the presented patient received 74 points, thus obtaining a moderate result. The evaluation of progress in the improvement process is presented in Table 3. Table 3. Results of rehabilitating the patient after total resection of femoral bone based on the Harris scale

Range tested	Before the surgery of total resection of femo- ral bone	After two years of post- surgery rehabilitation
Pain	sharp, seriously limits daily activities	none or almost unnoti- ceable
Extremity weakness	significant	large
Orthopedic supplies	a walking cane used most of the time (orthopedic shoe)	a cane used only for long walks
Walking distance	600 metres	unlimited
Climbing the stairs	usually with a handrail	usually without a handra- il
Putting on shoes and socks	with difficulty	with difficulty
Sitting	comfortably in an ordi- nary chair for 1 hour	comfortably in an ordi- nary chair for 1 hour
Public transport	No	No
Flexion contracture	No	No
The difference in the length of the extremi- ties	5 cm	0,5 cm
Test result	32 p.	74 p.

III. DISCUSSION

Currently, the combined Ewing's sarcoma treatment in children and adolescents is based on initial chemotherapy followed by surgery and post-operative chemotherapy and/or radiotherapy. The intention of surgical treatment is to perform a radical operation. [14] Amputations have been replaced by limb-saving operations. [13]

After total resection of the femoral bone, the general principles of rehabilitation used after hip or knee replacement apply. However, these patients require a special approach, because the surgery of total resection of the femoral bone involves not only replacement of both hip and knee joints, but also the excision of some muscles and resuturing of other muscles. Patients with extensive implants and muscle transfers require careful implementation of limb loading and a modified muscle enhancement programme. [15, 16, 17, 18]

The course of rehabilitation depends, among other things, on the type of endoprosthesis used, the extent of surgery in the area of removed muscles, e.g. quadriceps, ischiadic and shin muscles or gluteal muscle, as well as the method of muscle suturing. The patient is instructed as to the loading the operated limb and using the assistance (balcony, elbow crutches). Initially, isometric exercises or passive or active assisted exercises are implemented in the indicated range of movement. To minimise the risk of dislocations and to secure muscle attachments, active flexion above 90 °, adherence and internal rotation of the hip are not allowed for the first 10-12 weeks [17].

At the Royal National Orthopedic Hospital NHS Trust (RNOH) in London, guidelines for rehabilitation after total resection of the femoral bone were developed. The period of rehabilitation is divided into three stages: from 0 to 12 weeks, from 12 weeks to 6 months, from 6 months to 1 year. Expected results of therapy: range of motion in the knee joint 0 - 120, muscular strength of the quadriceps muscle of the thigh- 5 on the Lovett scale, elbow crutch gait, muscle strength of the hip joint abductor muscles- 4 on the Lovett scale, independence in the field of self-care and housework. At each stage, the authors set goals and indicated ways to achieve them by specifying precautions, the type of exercises (rehabilitation and occupational therapy), required orthopaedic supply and patient education [19].

With proper treatment and physiotherapy programme, patients after total resection of the femoral bone are able to achieve a satisfactory range of motion in the hip and knee joints as well as a high level of independence and function. General guidelines should help in the improvement process, but the treatment plan should be individually adapted to the patient's condition and capabilities.

IV. CONCLUSIONS

- The described case confirms the occurrence of complications after implantation of the long-stem hip joint endoprosthesis as a result of Ewing's sarcoma treatment.
- The appropriate physiotherapy programme implemented after total resection of the femoral bone allows you to regain lost functions.

VI. REFERENCES

- [1]Esiashvili N, Goodman M, Marcus RB Jr. Changes in incidence and survival of Ewing sarcoma patients over the past 3 decades: Surveillance epidemiology and end results data. J Pediat Hematol Onc 2008;30(6):425-30.
- [2] Ruka W. Mięsaki kości. [W:] Onkologia kliniczna. Krzakowski M (red.). Warszawa; Borgis – Wydawnictwo Medyczne, 2001:405-36.
- [3] Skowrońska-Gardas A.: Radioterapia w nowotworach wieku dziecięcego. Warszawa; Wydawnictwo Centrum Onkologii – Instytut im. M. Skłodowskiej-Curie, 2003;25-7.
- [4] Ruka W, Falkowski S, Nowacki Z, Rutkowski P. Mięsaki kości u dorosłych. [W:] Zalecenia postępowania diagnostycznoterapeutycznego w nowotworach złośliwych u dorosłych. Krzakowski M (red.). Warszawa; Wydawnictwo Polska Unia Onkologii, 2003:358-69.
- [5] Thomas P. Ewing's Sarcoma. [In]: Principles and practice of radiation. Oncology. Perez CA, Brady LW (ed.). Philadelphia; Lippincott Williams and Wilkins, 1998; 2038-45.
- [6] Dunst J, Jurgens H, Sauer R, Pape H, Paulussen M, Winkelmann W, Rube C. Radiation therapy in Ewing's Sarcoma: an update of the CESS 86 trial. Int J Radiat Oncol 1995;32:919-30.
- [7] Donaldson SS, Torrey M, Link MP, et al. A multidisciplinary study investigating radiotherapy in Ewing sarcoma: end results of POG8346. Int J Radiat Oncol 1998;42:125-35.
- [8] Paulussen M, Ahrens S, Dunst J, et al. Localized Ewing tumor of bone: final results of the Cooperative Ewing s Sarcoma Study CESS 86. J Clin Oncol 2001;19:1818-29.
- [9]Stowarzyszenie Pomocy Chorym na Mięsaki SARCOMA [online] [cited 2019 Feb 18] Available from: URL: http://www.sarcoma.pl/dla-chorych-iopiekunow/publikacje/miesak-ewinga-czyli-co-powinieneswiedziec-o-pierwotnych-nowotworach-kosci/
- [10] Chen WM, Chen TH, Huang CK, Chiang CC, Lo WH. Treatment of malignant bone tumours by extracorporeally irradiated autograft-prosthetic composite arthroplasty. J Bone Joint Surg 2002;84-B:1156-61.
- [11] Fox EJ, Hau MA, Gebhardt MC, et al. Long-term followup of proximal femoral allografts. Clinical Orthopaedic and Related Research 2002;397:106-13.
- [12] Randall L., Calvert G., Spraker H., Lessnick S.: Ewing's Sarcoma Family of Tumors (ESFT) [online] [cited 2019 Feb 18] Available from: URL:http://sarcomahelp.org/ewingssarcoma.html
- [13] Ramanathan D, Siqueira M, Klika A, Higuera C, Barsoum W, Joyce M. Current concepts in total femoral replacement. World J Orthop 2015;18, 6(11):919-926.
- [14] Hendershot E. Treatment approaches for metastatic Ewing's Sarcoma: a review of literature. JOPON 2005;22(6):339-52.
- [15] Shehadeh A, Dahleh M, Salem A, Sarhan Y, et. al. Standardization of rehabilitation after limb salvage surgery for sarcomas improves patients' outcome. Hematol Oncol Stem Cell Ther 2013;6(3-4):105–111.
- [16] Paramanandam S, Daptardar A, Gulia A. Rehabilitation following limb-salvage surgery in sarcoma. J Bonae Soft Tissues Tumors 2016;2(2):19-21.
- [17] Katarak P, O'Connor B, Woodgate I. Rehabilitation after total femur replacement: a report of 2 cases. Arch Phys Med Rehab 2003;84:1080-4.
- [18] Sewell M, Spiebelberg B, Hanna S, Aston W, et al. Total femoral endoprosthetic replacement following excision of bone tumours. J Bone Joint Surg [Br] 2009;91-B:1513-20.
- [19] Royal National Orthopaedic Hospital. Rehabilitation Guidelines. [online] [cited 2019 Feb 18] Available from: URL: https://www.rnoh.nhs.uk/our-services/rehabilitation-guidelines