ORIGINAL ARTICLE

# GIANT PENOSCROTAL LYMPHEDEMA: PLANNING AND SURGICAL TECHNIQUE TO TREAT A RARE DEBILITATING DISEASE

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**Background:** Giant penoscrotal lymphedema is a rare condition and is treated by surgical debulking and reconstruction with remaining skin and skin grafts. The described techniques may result in a staged surgery, multiple blood transfusions, orchidectomy and early debulking of the scrotal skin. We present a case series describing our technique to address all the concerns, discuss management to decrease progression and transmission in secondary cases and present a novel questionnaire to assess of quality of life of these patients. **Methods:** This descriptive case series was done from July 2016 to October 2019. Patients with Campisi grade 5 disease were included. Clinical assessment and relevant investigations were done to identify the cause and confirm the extent of the disease. Procedural detail, post-op haemoglobin levels (Hb), need for transfusion and weight of excised specimen were recorded. Wound healing, recurrence and body mass index were noted on follow up. A scrotal lymphedema quality questionnaire was developed and was filled on follow-up visit. **Results:** Twelve patients were operated on. The mean history was 3.0±0.5 years. 4 tested positive for microfilariae, while 4 out of 8 who tested negative had taken the anthelmintic drug. The mean weight excised was 15.8±2.3 kg, mean pre-operative score on quality-of-life assessment questionnaire was 83.3±2.6 versus 9.3±0.8 post operatively. The mean follow up time was 1.4±0.6 years. 1 patient had a minor recurrence necessitating re excision. Mean Hb was 13.5±0.5 mg/dl preoperatively compared to 11.8±0.5 mg/dl post operatively, with none requiring transfusion. **Conclusion:** Single staged excision with split thickness skin grafting is an effective and safe way to treat patients with giant scrotal lymphedema. It's the single best way to address the quality of life of patients.

**Keywords:** Lymphedema; Penoscrotal; Reconstruction

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## INTRODUCTION

Giant peno-scrotal lymphedema is a rare condition. It is classified as either primary or secondary, depending on whether it is acquired or not.[1] Scrotal lymphoedema can result from infection, malignancy, radiotherapy, lymphadenectomy or in bedridden patients.[2,3] Contrary to other areas, giant penoscrotal lymphoedema is usually secondary and in southeast Asia, caused mostly due to filariasis and less commonly due to other causes.[2,3] It presents a physical, social and psychological dilemma for the patient and a challenge for the plastic surgeon in terms of planning, surgery and post-operative management. The associated problems associated with it are not well documented as so in the case of lower or upper limb lymphoedema due to rare presentation but is graded similarly to lymphedema of other regions. As it usually presents in the most advanced stage with fibrosis and severe keratosis, debulking surgery in any form is the only option that remains.[4] Either staged excision or single stage debulking with coverage using the native remaining skin is described in the literature.[5,6] The staged debulking surgery reduces the risk of blood loss generally associated with reductive procedures, while coverage with uninvolved local skin at the perineum is advocated to minimize the risk of graft loss.[7,8] But this results in the poor aesthetic outcome and early recurrence requiring redo surgery.[7,8] Thus to address these issues, we present a case series where we describe our single stage surgical technique resulting in minimal blood loss, with a good aesthetic outcome and a very less rate of redo surgery. As there has been no published tool to evaluate the quality of life of patients suffering from giant scrotal lymphedema, we also present a quality-of-life assessment questionnaire that we have made and present the documented improvement in our cases.

## MATERIAL AND METHODS

This descriptive case series was done in Jinnah Burn and Reconstructive Surgery Center Lahore, from July 2016 to October 2019. After informed

consent, patients were recruited for the study. We included all male patients who were referred with Campisi grade five giant scrotal lymphedema, whether they had any co-morbidity or were on any medication.[9] Careful history was taken to identify primary or secondary lymphedema and any cause or disease in the case of secondary lymphedema. Associated symptoms, frequency of lymphangitis and relevant history was also taken. Local, regional lymph node and systemic examinations were done. Contrast enhanced magnetic resonance imaging (MRI) was then done to see the extent of the disease, define a surgical road map and investigate inguinal lymphadenopathy. Additional relevant investigations were done to identify the cause of the lymphedema. Procedural detail was clearly recorded, and post op complete blood count was advised to guide blood transfusion. The weight of the excised specimen was also noted. The patient was followed post operatively on OPD bases to assess wound healing, recurrence and BMI. The scrotal lymphedema quality questionnaire was developed after noting the complaints of the patients and taking guidance from the SF-36 quality of life questionnaire. Patients were requested to fill out the forms on follow-up visit.

Under general anaesthesia, the patient was placed in the supine position. A pit over the swelling was identified through which the patient passed urine. Midline was marked over the swelling with reference to the abdomen extending from the base of the penis to the pit, and an elliptical marking was done around the root of the penis and scrotum. Infiltration at the marking site was done to control bleeding. Multiple small stab incisions were made through the thick keratotic skin along the marking and then infiltration was done with infiltrating liposuction cannula, introduced deep within the swelling with the help of a 60cc syringe. We used a solution of 1-litre ringer lactate solution with 1 ml epinephrine (1:1000) and 30 ml 1% xylocaine for the infiltration. After waiting for 7–10 minutes, a probe was inserted in the pit and an incision was made along the marking. Dissection was done with monopolar cautery and exploration was done using skin marking and a probe as a guide to locate the glans. Once the glans were identified, urinary catheterisation was done and the penile shaft was followed just over the deep (Buck's) fascia until the root of the penis was reached. The penile shaft was dissected circumferentially, and then spermatic cords were easily identified through lateral dissection at the level of the root of the penis. Dissection was then done in antegrade fashion until the testes were completely dissected from the swelling. The incision was made circumferentially to completely remove the swelling. Blood loss was minimized due to the use of cautery and excellent haemostasis, achieved with infiltration and selection of the plane of dissection. Near the junction of the perineum and the swelling, large veins were identified and tied as the excision was completed. The wound was narrowed, and split thickness skin graft was harvested from the thigh. An unmeshed sheet graft was sewn in an oblique fashion over the penile shaft and meshed skin graft was placed over the testes with a vertical seam over the position of the midline raphae after stitching the testes together. Foam dressing was done over the penile shaft to create a roll and testes were sandwiched between two separate foams to reduce shearing and improve graft take. The dressing was opened after 4 days; the urinary catheter was removed at 7$^{th}$ post-operative. Long term use of emollients was advised over the grafted area. The resected specimen was sent for histopathology to rule out malignancy, especially lymphangiosarcoma. Figure 1 shows a case with giant scrotal lymphedema, treated with excision of the keratotic and fibrosed penile and scrotal skin and coverage with skin graft and follow up status after 2 years with no recurrence.

Table-1 shows the questionnaire that was made after exploring the symptoms of the patients and taking guidance from the general SF-36 quality of life questionnaire.[10–12] Patients were requested to fill out the form by recalling their condition before and after the procedure during the follow-up period. Health, limitation of activity, health problems, social problems, physical problems and emotional health were assessed, and quality of life was graded on a score of 104.

## RESULTS

Twelve patients were operated on with giant scrotal lymphedema of the penis and scrotum. Six patients were from central Punjab, 3 from the south and 3 were from northern Punjab. All the patients were of middle age, as the average age was 33.8±5.4 years. The patients had an average history of 3.0±0.5 years since the onset of the swelling. None of the patients had any family history and 4 out of 12 patients tested positive for microfilariae. Four patients had taken an anthelmintic drug and tested negative for the test. All patients had lymphoedema limited to the penoscrotal region. The average weight of the excised lymphedematous swelling was 15.8±2.3

kg, with a maximum of 20 kg and a minimum of 13 kg. Figure-1 shows the outcome of a patient treated with our technique, with no recurrence after 1 year and 10 months. The average pre-operative score on quality-of-life assessment questionnaire (Table-1) was 83.3±2.6 as compared to 9.3±0.8 post operatively ($p$=0.01). Average follow up time was 1.4±0.6 years, with a maximum of 2 years 8 months and a minimum 1 year. One patient had minor recurrent swelling of the penile shaft necessitating excision and grafting of the penile and scrotal swelling (Figure-2). Average value of haemoglobin on complete blood count was 13.5±0.5 mg/dl pre operatively as compared to 11.8±0.5 mg/dl post operatively, thus none of the patients required blood transfusion. All the patients had sub centimetric enlargement of the inguinal lymph nodes on MRI and thus lymph node dissection was not done in any case. Graft take was good in all cases and wounds healed without any complication. Histopathology report revealed dense fibrosis and keratosis and no signs of malignancy were seen in any case. 4 cases who tested positive for microfilariae and 4 who tested negative were given albendazole post operatively.

## DISCUSSION

Giant scrotal lymphoedema is a rare condition, with most case reports originating from the subcontinent and Africa. Patients usually hide their problems for social reasons and seek medical attention when there is profound enlargement and fibrosis.[13] Peno scrotal lymphedema is reported both as a primary and secondary entity, but the giant peno scrotal lymphedema which is difficult to treat is usually of secondary variety.[8,13,14] In South East Asia, where filariasis is endemic, giant penoscrotal lymphedema usually results from filariasis.[15,16] Although Wuchereria bancrofti is endemic in Southeast Asia, recently it was not regarded as endemic in Pakistan.[17] Two of our patients had already taken anthelminthic drugs previously, but three out of five patients tested positive for microfilariae. Immunologic tests for circulating filarial antigen were not available at any laboratory in Pakistan, so we resorted to the detection of microfilariae on blood smear examination, in nocturnally drawn blood samples between 10 pm to 2 am. This test was attributed as quick but had less sensitivity and depended on the pathologist's experience as well.[18] For these reasons, we decided that all patients be given anthelminthic therapy to prevent the spread and further damage to the lymphatics. Diethylcarbimazole, albendazole, doxycycline and ivermectin are all described as treatment as either a single drug or in combination.[19,20] Diethylcarbimazole is effective for both adult worms and microfilariae, but ivermectin is effective against microfilariae and doxycycline against the adult worm. Although diethylcarbamazine offers a straight forward treatment, it was not available in Pakistan. Thus, we gave a combination of doxycycline and ivermectin to all patients.

Traditionally, reductive, or excisional procedure of the lymphedematous tissue results in blood loss.[7] But we found that none of our patients required any transfusion post-operatively. Although vessels within the fibrotic tissue didn't contract, infiltration at the root of the scrotum and penis, together with infiltration of the swelling along the incision line reduced blood loss. Dissection was also very easy and blood less if a relatively avascular plane of tunica albuginea was followed. Thus, we have described our surgical technique in detail as it is significantly safer for the patients suffering from giant scrotal lymphoedema.

In the published literature, skin grafting of the scrotum was avoided and local tissue at the perineum was principally used to cover the testis. It had been attributed that graft loss was profound over the scrotum which required repeated dressings to heal the wound.[21–23] On the contrary, what we found that meshed skin graft prevented haematoma, seroma or lymph underneath the graft. Together with an enveloping foam dressing, this resulted in good graft take and better aesthetic results when already oedematous skin of the perineum was used. With the seam of the graft vertically orientated at the midline, a natural looking frenum was created. Similarly, unmeshed sheet grafting, obliquely oriented over the penile shaft reconstructed a natural looking shaft and prevented contracture. Excision of as much tissue of the penile and groin area reduced the chances of recurrence and contributed to better aesthetic results. One of the patients who had undergone this procedure underwent re excision and grafting after a follow-up of 2 years. We didn't find atrophic testicles in any case, thus orchidectomy was not done.

Giant scrotal lymphoedema greatly disturbs the quality of life of the patient. As this is a rare entity, no tool to grade the quality of life of patients suffering from such a problem could be found in the literature.[24] Particularly in Western countries, where it is even rarer, no such questionnaire has been developed. We developed a questionnaire specifically directed towards this entity to assess the impact of disease on the patient's life and improvement after the surgery. As we have found, significant improvement with the surgery can be achieved in a single setting. Thus, excision and grafting act as the main step towards rehabilitation of the patient and returning him back to normal life.


**Figure-1: A 30 years old patient presented with giant peno-scrotal lymphedema. Single staged debulking and coverage with skin graft was done. Post op result after 1 year and 10 months.**

**Figure-2: A 40 years old patient presented with Giant Peno-Scrotal Lymphedema. Excision and grafting was done. 1 year post operatively, patient requested for debulking and grafting of lymphedematous tissue of the penile shaft.**

**Table-1: comprehensive penoscrotal lymphoedema quality of life assessment questionnaire**

PENO SCROTAL LYMPHEDEMA: QUALITY OF LIFE ASSESSMENT QUESTIONNAIRE

Name: Date: ID#: Age: Gender: M / F

| **GENERAL HEALTH:** | **GRADING** | | | | |
|---|---|---|---|---|---|
| Generally, how does your condition effect your health | 0 | 1 | 2 | 3 | 4 |
| **LIMITATIONS OF ACTIVITIES:** | | | | | |
| How much does you condition limit vigorous activities, e.g running, lifting heavy objects | 0 | 1 | 2 | 3 | 4 |
| How much does you condition limit moderate activities, such as moving a table: | 0 | 1 | 2 | 3 | 4 |
| How much does your condition effect following activities: | | | | | |
| Lifting or carrying groceries | 0 | 1 | 2 | 3 | 4 |
| Climbing flights of stairs | 0 | 1 | 2 | 3 | 4 |
| Bending, kneeling, or stooping | 0 | 1 | 2 | 3 | 4 |
| Walking several blocks | 0 | 1 | 2 | 3 | 4 |
| Walking one block | 0 | 1 | 2 | 3 | 4 |
| Difficulty in passing stool | 0 | 1 | 2 | 3 | 4 |
| Dressing yourself | 0 | 1 | 2 | 3 | 4 |
| Cleaning yourself | 0 | 1 | 2 | 3 | 4 |
| Sleeping | 0 | 1 | 2 | 3 | 4 |
| Difficulty in passing urine | 0 | 1 | 2 | 3 | 4 |
| **PHYSICAL HEALTH PROBLEMS (How much does your problem affect your work or other regular daily activities)** | | | | | |
| How much have you cut down the amount of time you spent on work or other activities | 0 | 1 | 2 | 3 | 4 |
| How much your condition has affected you to accomplish less than you would like | 0 | 1 | 2 | 3 | 4 |
| How much you are limited in your work or other activities due to your problem | 0 | 1 | 2 | 3 | 4 |
| How much pain do you have due to your condition | 0 | 1 | 2 | 3 | 4 |
| How difficult is it to maintain hygiene | 0 | 1 | 2 | 3 | 4 |
| | | | | | |
| **SOCIAL ACTIVITIES: How much does your condition affect you in:** | | | | | |
| Relationship with your spouse | 0 | 1 | 2 | 3 | 4 |
| Spending time with your family | 0 | 1 | 2 | 3 | 4 |
| Socialize with friends and relatives | 0 | 1 | 2 | 3 | 4 |
| Getting involved in social activities | 0 | 1 | 2 | 3 | 4 |
| **ENERGY AND EMOTIONS: How much does your condition affect you in the following ways**: | | | | | |
| Do work or other activities as carefully as usual | 0 | 1 | 2 | 3 | 4 |
| Feel anxious due to your condition? | 0 | 1 | 2 | 3 | 4 |
| Feel depressed & nothing cheers you up? | 0 | 1 | 2 | 3 | 4 |
| Loss of energy / tired? | 0 | 1 | 2 | 3 | 4 |
| | | | | | |
| | | | | | |
| SCORE: _____ / 104 | | | | | |
| | | | | | |

## CONCLUSION

Giant scrotal lymphedema is a physically and mentally debilitating condition. It is mostly associated with W Bancrofti infection where filariasis is endemic. Most people seek medical advice late in the course as it is often thought of as untreatable. A combination of anthelminthic drugs at a later stage can prevent the spread and further destruction of lymphatics only, but the mainstay of treatment is surgical. Excision and grafting with the technique that we have described results in easier dissection, decreased blood loss and favourable outcomes as shown by the questionnaire that we have developed.

## AUTHORS' CONTRIBUTION

AUH: Conceptualization, data collection, interpretation, case contribution. MOA: Literature search, conceptualization, data collection, analysis, write-up, case contribution. MNT: Conceptualization, case contribution, data analysis, proof reading.

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