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Reconstructive

Reconstruction Approach to a Rare Case of Acquired Scrotal Giant Muscular Hamartoma

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Summary: Acquired scrotal giant muscular hamartoma is an uncommon benign lesion with fewer than 10 documented cases all over the world. It is characterized by a proliferation of dermal smooth muscle bundles of scrotum dartos fascia. The authors report a rare case of acquired scrotal giant muscular hamartoma, which occurred in a 70-year-old severely obese and diabetic man presenting with a progressive scrotal enlargement and swelling in the last year, causing marked reduction in quality of life and cosmetic problems. The patient underwent a wide excision of the hamartomatous lesion, and then, a reductive scrotoplasty and autologous skin grafting of penis were performed. Anatomopathological examination showed an acquired scrotal giant muscular hamartoma arising from muscular fascia of dartos. This surgical technique is a valid, safe, effective, and minimally invasive option to treat this pathology, achieving both excellent functional and aesthetic results, with a marked improvement of the patient's quality of life. *(Plast Reconstr Surg Glob Open 2016;4:e857; doi: 10.1097/GOX.0000000000000828; Published online 7 September 2016.)*

The hamartoma is a benign neoplastic lesion that combines cellular elements of various origins. There exist a congenital smooth muscle hamartoma and an acquired smooth muscle hamartoma (ASMH) form.[1–6]

The ASMH is rare, with fewer than 20 cases reported all over the world, and it often occurs in patients aged between 14 and 70 years. The anatomical regions most commonly affected are the forearm, the neck, the breast, and the genital region.[7,8]

The scrotal location is considered an exceptional event, with a very few cases reported in the literature (<10): ASMH can easily be confused with many other skin conditions such as a massive lymphedema, and benign or malignant tumor processes. Furthermore, it can be complicated by a different severity of lymphedema.[9–12] Histologically, it presents a local increase of large unorganized bundles of fusiform smooth muscle cells, well differentiated, with a characteristic fibrillar vacuolated cytoplasm and a cigar-shaped nucleus. The α-smooth muscle actin and desmin immunohistochemical positivity confirm the muscular histological origin.[13] The hyperproliferative muscle bundles originating from the dartos fascia are arranged randomly in the dermis but can potentially expand to the subcutaneous tissue and to the testicles.[14,15] In the literature, there are no reports of systemic involvement or malign neoplastic degeneration.[16]

A patient presenting an acquired scrotal giant muscular hamartoma who underwent a reductive scrotoplasty and autologous skin grafting of penis is described in this case report.

CASE REPORT

On January 2015, a 70-year-old white man, severely obese (body mass index > 50 kg/m^2), diabetic, and with hypertension, came to our attention, reporting an increase of the scrotum volume. The symptom appeared about 1 year earlier with progressive worsening.

The framework of "giant scrotal lymphedema" with multiple erythematous and ulcerated skin areas measuring about 20 × 32 cm was evident at physical examination. The scrotum, in supine position, came up to the level of the femorotibial joint. The skin appeared rough, tense, and inelastic with presence of multiple fixed papules of about 0.3 to 2 cm in diameter, located over the whole scrotum. The penis was completely swollen, slightly identifiable (Fig. 1). Computed tomography scan of lower abdo-

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men described a case of marked corpusculated hydrocele without any inguinal–scrotal hernias. The symptomatology was poor (eg, impossibility to maintain the orthostatic position for extended periods), added to the marked reduction of quality of life.

A reductive scrotoplasty with contextual autologous skin grafting of the penis was planned for this patient. We performed an inverted V cutaneous incision of the scrotum; the testicles were identified dipped in a whitish, lardaceous, edematous, and hypervascularized tissue, which surrounded the penis too. The testicles were isolated from this tissue, and the penis was degloved through a coronal sulcus incision. After that, we proceeded to remove the hamartomatous mass, maintaining a sufficient portion of local skin (Fig. 2). The portion of the scrotum removed weighed about 6 kg (Fig. 3). Two lateral random skin flaps were used for the reconstruction of the scrotum. These flaps, after orchidopexy, were sutured along the median raphe and circumferentially at the base of the penis. The penis, without skin mantle, was then covered with an autologous partial-thickness skin graft taken using an electrical dermatome from the anterolateral thigh area. The graft was meshed and immobilized by moulage. The donator area was medicated with calcium alginate. A scrotal suction drainage was inserted.

The drainage was removed after 3 days. Antibiotic prophylaxis with levofloxacin was administered. During the follow-up, we noticed a tiny area of dehiscence on the scrotum. A wound dressing with silver and hydrofiber was used for complete healing in 15 days.

The moulage on penis was kept in place for 5 days. We achieved a good skin graft take. The sutures were removed progressively over a period of 15 days. The donor site on the thigh was medicated intraoperatively with calcium alginate followed by application of a nonadherent gauze, changed every 2 to 3 days, until complete re-epithelialization was achieved. The patient was discharged on the sixth day with good clinical conditions.

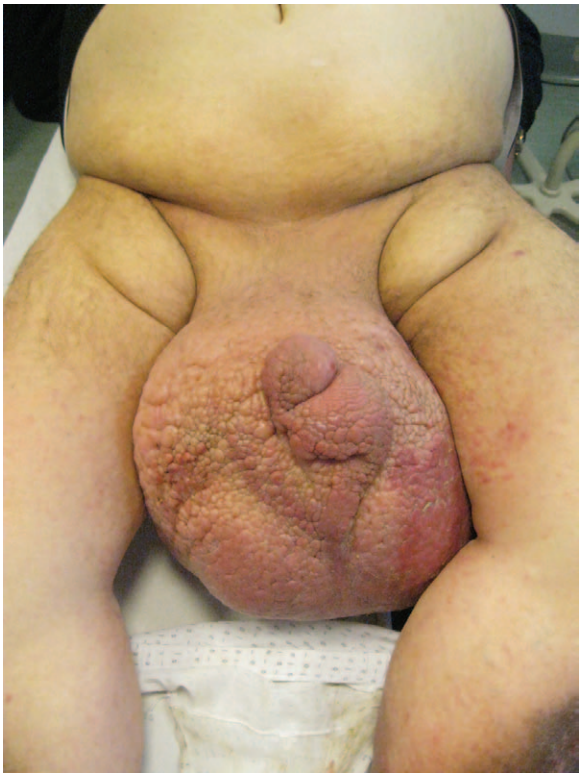

Fig. 1. Physical examination of the patient presenting giant scrotal lymphedema with multiple erythematous and ulcerated skin areas measuring about 20 × 32 cm.

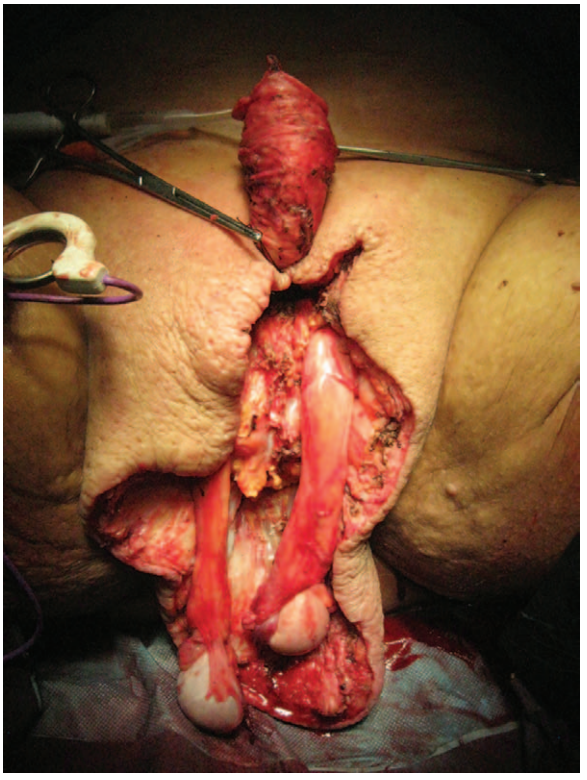

Fig. 2. The isolated testicles after the mass removal.

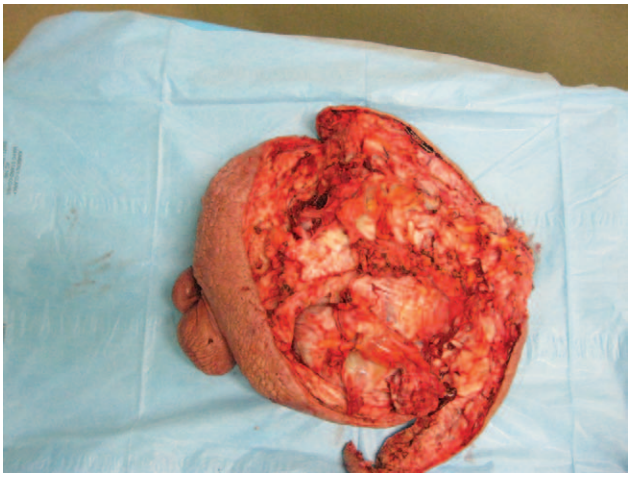

Fig. 3. The hamartomatous mass removed weighing around 6 kg.

Anatomopathological assessment revealed a scrotal giant muscular hamartoma arising from muscular fascia of the dartos.

After 2 months of follow-up, the patient presented normotrophic scars all over the genital region and the donor site of the skin graft. The skin graft applied on the penis resulted trophic (Fig. 4). The patient reported a marked improvement in quality of life with improvement of personal autonomy (walking, upright position, personal hygiene) and aesthetical functionality of the genital area.

DISCUSSION

The first reported reductive scrotoplasty with local random skin flaps and skin grafting of the penis was performed in 1820.[17] Because ASMH is considered a disease with exceptional incidence, only a few studies are present in the literature. However, it is certain that this disease may have a recurrence because of a nonradical resection or a local excessive spreading. Brotherhood et al[18] successfully applied the reductive scrotoplasty with skin grafting of the penis to treat a case of idiopathic scrotal elephantiasis. van Kooten et al[3] treated 2 cases of chronic scrotal lymphedema outcomes only by dermoepidermic grafts. Instead, Chen et al and Semerci et al were among the first to treat 2 cases of giant acquired scrotal muscular hamartoma using the technique above proposed by Brotherhood et al. Even in these cases, they did not observe any postoperative complications, resolving the disease in a radical way and with excellent functional results.[1,11]

After analysis of the results reported in the literature, we decided to use the technique proposed by Chen et al and Semerci et al. The aim of this surgery is a radical removal of hamartomatous lesion, maintaining the functionality of the genital tract and recreating the scrotal and penis skin mantle. We used 2 perineal random skin flaps sculpted on the healthy surrounding scrotal skin that were then medially sutured. The biggest risk was the possibility of a postoperative wound dehiscence. Special attention must be paid to hemostasis to prevent a blood subcutaneous mass formation that can compromise the vascularization of the flaps. The penis, instead, once degloved and leaked out through a new orifice created along the median suture, was covered with an autologous Ollier–Thiersch skin graft. The penis immobilization was necessary for the optimal skin graft take. For this purpose, the insertion of a bladder catheter for the first 5 postoperative days is essential.

Expecting a difficult engraftment, Stokes et al[19] applied over the skin graft a negative pressure dressing (−75 mm Hg), which was removed on fifth day to treat a case of scrotal-penis elephantiasis.[19]

We also agree that in cases with a high probability of skin graft failure, negative pressure can be used and can be a very useful tool.

In case of insufficient skin to create local healthy flaps, it is possible to insert scrotal skin expanders as already described by Chen et al.[1]

In our opinion, the technique of Chen et al and Semerci et al is the best surgical solution to treat scrotal giant muscular hamartoma. We do not think that there exist other alternatives except the skin grafting for penis reconstruction. The possible use of a local skin flap for penis coverage gives very poor results in terms of cutaneous thickness and shape.

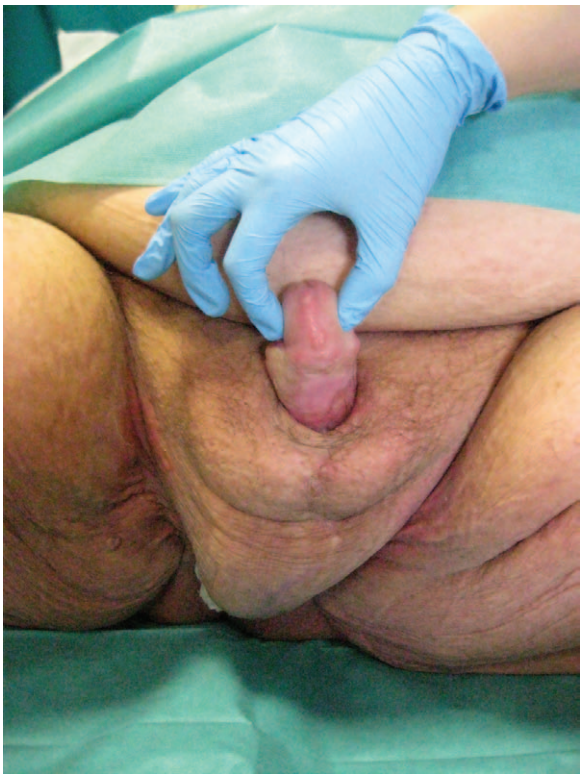

Fig. 4. Follow-up after 2 months.

CONCLUSIONS

It is evident that further studies should be conducted to better define this rare clinical condition and consequently identify the best therapeutic approach. The use of 2 perineal random skin flaps and skin grafting of the penis is a valid, safe, effective, and minimally invasive technique. It also allows excellent functional and aesthetic results, with a marked improvement of the life quality of the patient.

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The study was approved by the ethics committee of our institution and was performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki and its later amendments.

PATIENT CONSENT

Patient provided written consent before his inclusion in the study.

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