Giant Abdominal and Penoscrotal Idiopathic Lymphedema

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The lymphatic system produces and transports fluids, immune cells and fats throughout the body. Abnormal transport and accumulation of lymph fluid may cause swelling (lymphedema).

Lymphedema, occurring quite frequently in the medical practice, may appear in a variety of pathologies such as: allergic reactions, infectious diseases, localized infections, radiotherapy, insect bites, as post-surgery reaction, etc. In all cases, it represents a subsequent effect of the main illness [1].

There is very small number of cases where lymphedema occurs as sole affection, the so-called "primary or idiopathic lymphedema". It is considered a genetic disease resulting in agenesis or the insufficient development of the lymphatic system [2].

The FLT4 gene provides instructions for producing a protein called vascular endothelial growth factor receptor 3 (VEGFR-3), which regulates the development and maintenance of the lymphatic system. Mutations in the FLT4 gene interfere with the growth, movement, and survival of lymphatic cells. These mutations lead to the development of small or absent lymphatic vessels. If lymph fluid is not properly transported, it builds up in the body's tissues and causes lymphedema. It is not known how mutations in the FLT4 gene lead to the other features of this disorder, but many of these patients do not have a FLT 4 gene mutation. In these cases the cause of the swelling is unknown [1,3].

Depending on the time of occurrence, the lymphedema may be present at birth (Milroy syndrome) or may occur later in life – either during teenage (praecox form or Meige disease), or later on in life (the tardy form).

Milroy's Syndrome is an old term used to describe hereditary congenital lymphedema. It is a congenital familial primary lymphedema which results from vertical autosomal inheritance of a single gene. The condition usually presents itself at birth with the swelling of one or even both legs. If the condition is unilateral, the other leg may continue in the latency stage for years before expressing itself [2,4].

The other forms of idiopathic lymphedema, in either praecox or tardy form occur during teenage or during adulthood and include more or less extended areas of the body, in most cases being localized in the lower limbs and/or the genitalia. These forms are often reccurring, the recurrences appearing within years and even in parts of the body different from the original affected area [3].

Below we are presenting the case of the patient C.M., aged 43, coming from rural environment, hospitalized in our clinic on February 16, 2009, diagnosed with a fast growing tumor, which started developing 6 months ago, affecting the penis, the left hemiscrotum and the lower part of the abdominal wall. From the patient's medical record, we have noticed a history of idiopathic lymphedema of the right hemiscrotum, operated 20 years ago (at the age of 23).

During the physical exmination, we have observed a giant tumor comprising the penis, the left hemiscrotum and the lower abdominal wall, of solid consistency, painless, with uneven surface and numerous crypts with stagnant, fetid secretions. The teguments in the close vicinity of the tumor are normal-looking (Fig. 1 and 2).

Paraclinical investigations – laboratory and imagistic – do not show pathologic modifications.

After a general pre-operatory preparation with antibiotics and local preparation with antiseptic wash, the patient undergoes surgery on February 20, 2009, with our team performing a large disection and excision of the abdominal penoscrotal idiopathic lymphoedema, preserving both spermatic funiculi and both testicles, closing the remaining post-surgery wounds with club-shaped tegumentary strips, creating a neoscrotum, covering the post-excision wound of the penial head with split skin graft, thick type. Figures 3 and 4 illustrate intraoperatory aspects.

The postoperatory evolution was slow but favorable, dominated by abundant lymphoragia on the drain tubes and on the postoperatory wounds, which required even 2 dressings/day at times. Three weeks after surgery, a quasi-circular necrosis of the skin graft at the base of the penis occured, requiring another minor skin graft intervention. The erectile function of the penis has been fully preserved.

The aesthetic results observed during examination 10 weeks after the surgery are presented in Figure 5.

References

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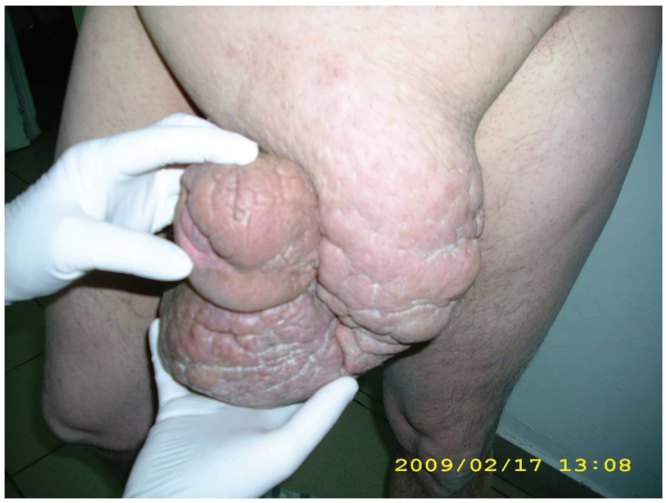

Fig. 1.

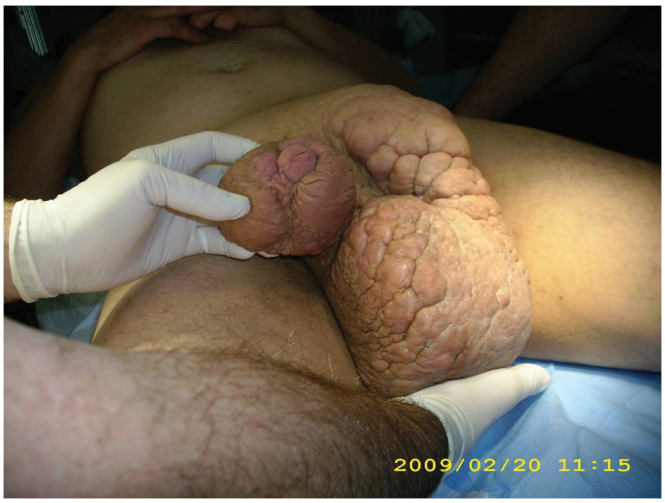

Fig. 2.

Fig. 3.

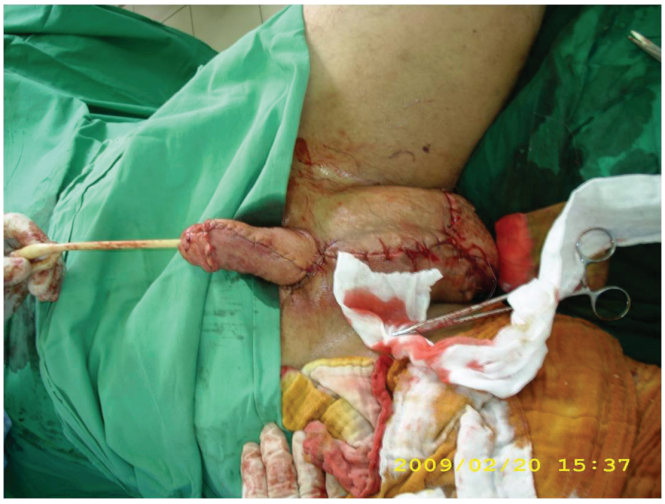

Fig. 4.

Fig. 5.