RESEARCH PAPER | Medical Science | Volume : 5 | Issue : 3 | March 2015 | ISSN - 2249-555X

A Rare Case of Isolated Primary Penile Lymphoedema

KEYWORDS: lymphedema, penile, primary.

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ABSTRACT *Lymphedema is the occurrence of chronically swollen extremities or rarely the genitals due to inadequate drainage of interstitial fluid by the lymphatics. We report a case of isolated primary penile lymphedema which is a rare entity (1:60,000 live births). Surgery is the line of treatment for this condition.*

Introduction

Lymphedema is the accumulation of protein-rich fluid due to inadequate drainage of interstitial fluid by the lymphatics. [1,2] Isolated penile lymphedema is a relatively rare entity(1:60,000 live births). The literature on the subject and its management is very less. There is no demonstrable infective cause, it is persistent and there is no documented effective treatment .It differs from other forms of penile edema in several ways.[3,4,5] We report a rare case of isolated primary penile lymphedema.

Case report

We report a case of isolated primary penile lymphoedema in a 34-year old male. The patient presented with swollen penis of 4 months duration. The patient had right direct inguinal hernia. He did not have any history of trauma, infection, or any other cause of secondary lymphoedema. Clinically, he did not have lymphadenopathy. General and systemic examinations were essentially normal. Genital examination revealed enlarged, non-tender, large-curved penis measuring 5.5inches in length and 4 inches in circumference (figure 1). The scrotum and lower limbs were normal with no signs of lymphoedema.

Urinalysis, hematological, and biochemical tests were normal. Ultrasound abdomen was normal. Serologic study was negative for filariasis. With the diagnosis of primary lymphoedema of penis , surgery was planned in which the involved tissue was resected and scrotoplasty was done by the use of skin flaps.

Grossly specimen consisted of 3 irregular soft tissue masses out of which larger two were skin covered. The largest mass measured 8.4X5.2X0.5 cm and the smallest mass measured 1X0.5X0.3 cm. External surface showed flattening of skin. Cut surface was unremarkable.

Histopathology showed thickened epidermis below which dense aggregates of lymphoid cells were seen in a collagenous background (figure 3).

Figure 1. Pre-operative photograph of penile lymphedema

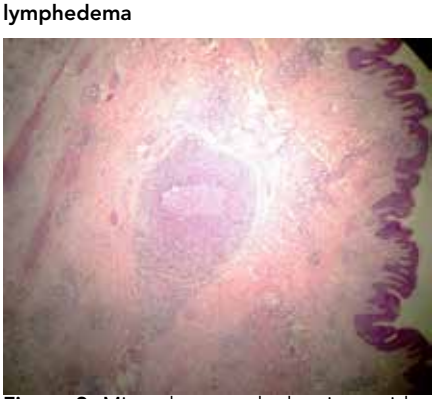

Figure 3. Microphotograph showing epidermis with underlying dermis showing lymphoid aggregates in collagenous background (H and E X10).

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Discussion

Lymphoedema results from the accumulation of protein-rich lymphatic fluid in the skin and subcutaneous tissues due to a dysfunction in the lymphatic system. In the chronic stage, lymphoedema is also characterized by the deposition of fat and fibrous tissue.[2] Lymphoedema is an abnormal collection of interstitial lymph fluid due to either congenital maldevelopment of the lymphatics or secondary obstruction. [4] Lymphedema is of two types: primary and secondary. Primary lymphedema is uncommon and has female predominance. It may be congenital or familial (Milroy's disease) or idiopathic appearing either at puberty (precox), or after 35 years of age (tardum).[5-7] Primary penile lymphoedema is a rare occurrence and usually involves the lower limbs, but rarely the genitalia.[4,8,9] Penile and scrotal lymphedema mostly is mostly due to infection or as a reaction to trauma. Idiopathic lymphedema is rarely seen and is caused by a primary obstruction of lymphatic vessels of scrotum. [5,10]

The obstruction of lymphatic vessels causes reduced lymphatic flow further causing enlargement of the genitalia. The patient suffers with severe discomfort, limitations of local hygiene and limitation of movement, progressive loss of urinary function, cosmetic and sexual problems which is caused by persistent penile lymphedema, skin excess and altered sensitivity over the distal penile shaft.[1,5,11,12] Congenital lymphedema of the genitalia has various effects over the growing child both physical as well as psychological. Genital elephantiasis is a functionally disabling and emotionally incapacitating entity.[5] There is no effective medical treatment; however prolonged course of fluoroquinolones may be helpful in patients with spotty occurrence of genital lymphedema.

All these functional disabilities caused due to any kind of lymphedema causes extreme emotional stress and since response to medical treatment is very poor surgical intervention becomes unavoidable. Extensive resection of this tissue and reconstruction by skin grafting offers a less than satisfactory cosmetic result. [1,5,13-15]

Satisfactory surgical methods mentioned in the literature are as follows:

Physiologic methods or lymphangioplasty through which lymphatic discharge from involved regions to new lymphatic channels is obtained.

Lymphangiectomy with reconstructive surgery.[5,16]

Histological examination reveals areas of epidermal thickening, smooth muscle hyperplasia and chronic inflammation cells.[1] Hemorrhage, hematoma, urethral injury, infection, painful erection, decrease of sensation, and scar in suture line are few Surgical complications of elephantiasis or genital lymphedema.[5,16,17]

Conclusion

Primary penile lymphedema is rare entity. Although primary hypoplastic lymphatic channels cause this condition, in our opinion it usually occurs as a result of recurrent unidentified infections and/or another concomitant penile dermatosis causing subsequent damage to the lymphatic vessels.[3] Surgery is the main line of treatment.

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