

Case Report

A case report of lymphangioma circumscriptum of scrotum along with milky urethral discharge

Naeem Ahmed Soomro, Rakhi Kumari, Irfan Ahmed Shaikh, Kaiser Khan, Dileep Kumar Ahuja, Sirajuddin Ahmed*

Department of Dermatology Shaheed Mohtarma Benazir Bhutto Medical University Larkana

* Department of Pediatric Surgery Shaheed Mohtarma Benazir Bhutto Medical University Larkana

Abstract Lymphangioma Circumscriptum is a lymphatic malformation localized to skin, subcutaneous tissue and rarely muscles. It may present at any age but usually noted at birth or during childhood. It is due to malformation of lymphatic channels. We present a case of 9 years old patient of lymphangioma circumscriptum of scrotum along with milky urethral discharge. Patient was treated with oral sildenafil and CO₂ Laser.

Key words

Lymphangioma Circumscriptum, Sildenafil, CO₂ Laser.

Introduction

Lymphangioma Circumscriptum presents as small clusters of blisters filled with lymph fluid. These may be translucent when the overlying epidermis is thin or varying in color from red to blue when contains blood. Common sites are axillary folds, shoulders, neck, flanks, proximal parts of limbs, tongue and buccal mucus membrane.^{1,3}

Case report

A 9-years old male patient presented with clusters of blister formation on scrotum and milky urethral discharge for 7 years. According to the patient's father, it first appeared at the age of 2 years and was associated with the swelling of scrotum. There was history of trauma. It was not associated with fever or burning micturition. Family history was not significant.

Address for correspondence

Dr. Irfan Ahmed Shaikh, Associate Professor
Department of Dermatology, Shaheed Mohtarma
Benazir Bhutto Skin Complex, Larkana
Ph: 03332309649
Email: shaikhirfan913@gmail.com

On examination, multiple clusters of lymph filled vesicles of variable size ranging from 1 to 4 mm scattered throughout the swollen scrotum along with milky urethral discharge (**Figure 1**). It was not associated with lymphadenopathy or lymphedema. Ultrasound scrotum suggested thick walled scrotum. Biopsy of scrotal skin showed hyperkeratosis and acanthosis of epidermis with dilated lymphatic channels containing eosinophilic proteinous material (**Figure 2**). Patient was treated with sildenafil 75mg in three divided doses and CO₂ laser.



Figure 1 Swollen scrotum with clusters of skin colored vesicles

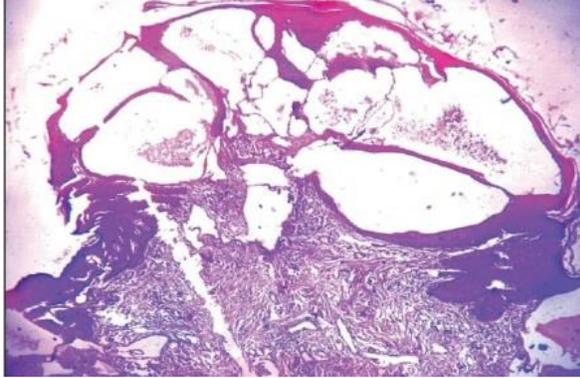


Figure 2 Histopathology of scrotal skin showing hyperkeratosis, acanthosis with dilated lymphatic channels

Discussion

Lymphangiomas can occur anywhere in the skin and mucus membrane.¹⁻² Common sites are axillary folds, shoulders, neck, proximal part of the limbs, tongue, and buccal mucus membrane, the scrotum is the rarest site. Although the disease is congenital, rarely there is occurrence of acquired cases in the scrotum³⁻⁵ and vulva.⁶ The disease is characterized by translucent vesicles of varying sizes scattered or grouped like frog spawn, containing clear lymph fluid. These vesicles are often associated with verrucous changes, which give them the warty appearance. The basic pathologic process in the congenital cases which are found at birth or early childhood are collection of lymphatic cistern in the deep subcutaneous plane that are separated from the normal network of lymphatic vessels. Acquired lymphangioma circumscriptum develops later, probably due to injury or damage from the deep collecting channels in the tissue, leading to stasis of lymph with backflow resulting in subsequent dilation of upper dermal lymphatics causing the lesion. These acquired cases are mostly due to infections like filariasis, lymphogranuloma venereum, tuberculosis, donovanosis, following

trauma, surgery, or radiotherapy.³⁻⁶ The treatment options include surgical excision, sclerotherapy, electrocoagulation, liquid nitrogen therapy and carbon dioxide laser therapy and oral sildenafil⁸ a selective phosphodiesterase. Our patient is treated with weekly CO₂ laser and 50 mg sildenafil, marked improvement was noted with decreased urethral discharge after two months.

References

1. Fox T, Fox TC. On the case of lymphangiectodes with an account of the histology of the growth. *Trans Path Soc, London* 1879; 30:470-6.
2. Morris M. Lymphangioma circumscriptum. *International Atlas of rare Skin Diseases*. In: Unna PG, Morris M, Duhring LA, Leloir H, editors. London: Lewis; 1889. p. 1-4.
3. Mohanty S, Gandhi V, Sing. Lymphangioma circumscriptum of the scrotum of late onset. *Indian J Dermatol Venerol Leprol* 1998; 64: 289-90.
4. Sheu JY, Chung HJ, Chen KK, Lin AT, Chang YH, Wu HH, et al. Lymphangioma of male exogenous organs. *J Chin Med Assoc* 2004; 64: 204-6.
5. Hagiwara K, Toyama K, Miyazato H, Nonaka S. A case of acquired lymphangioma due to suspected old filariasis and a review of literature. *J Dermatol* 1994; 21: 358-62.
6. Amouri M, Masmoudi A, Boudaya S, Amouri A, Ben Ali I, Bouassida S, et al. Acquired lymphangioma circumscriptum of the vulva. *Dermatol Online J* 2007; 13:10.
7. Treharne L J, Murison MS: CO₂ laser ablation of lymphangioma circumscriptum of the scrotum. *Lymphat Res Biol* 2006; 4: 101-103.
8. Swetman GL, Berk DR, Vasanawala SS, Feinstein JA, Lane AT, Bruckner AL. Sildenafil for severe lymphatic malformations. *N Engl J Med* 2012; 366: 384-386.