

Non-Filarial, Non-Lymphatic Elephantiasis of Scrotum

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Abstract

Scrotal elephantiasis is a clinical diagnosis made for isolated massive lymphedema of the scrotum; most commonly due filarial infestation. However, there are increasing reports of non-parasitical lymphedema of scrotum and non-lymphatic elephantiasis of scrotum with a common histological feature of massive fibrosis of skin, subcutaneum and connective tissues. We present a case of idiopathic fibrous pseudotumor of scrotum managed effectively by surgical excision.

Keywords: *Elephantiasis; Lymphedema; Fibrous disease; Scrotum; Pseudotumor*

1. Introduction

Scrotal elephantiasis is a clinical diagnosis made for isolated massive lymphedema of the scrotum due to any cause, resulting in 'woody' and indurated thickening of the skin and subcutaneous tissues. In a vast majority of cases, chronic lymphatic obstruction at lymph vessel or lymph node level leading to lymphedema is the underlying pathology. Although filarial infection is the most important cause of lymphatic obstruction in India, resulting in lymphedema, other non-parasitical conditions of lymphedema have also been reported to result in elephantiasis. However, we present a unique case of non-parasitic, non-lymphatic case of scrotal elephantiasis, which was initially misdiagnosed as due to filarial lymphedema.

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2. Case Report

A 37-year-old man presented with progressive swelling of the scrotum since the last four years. There was no fever, skin ulceration or suppuration. Since the penis was partially buried inside the enlarging scrotum, he had progressive difficulty in urination. There was no swelling present on any other body area and there were no other medical comorbidities. The patient had been treated by multiple courses of anti-filarial medication in the past due to suspected filariasis, without having clinical improvement. On examination, patient was obese, with a body weight of 101 kilograms, height of 170 cm and BMI of 34.9 kg/m². The scrotum was hugely enlarged and measured about 30 cm × 40 cm × 40 cm in size (FIG. 1, Panel A). The penis was buried partially in the scrotal swelling. The overlying skin was normal without any signs of inflammation, exfoliation, ulceration, or sinuses. There was no significant skin induration. However, the subcutaneous tissue was grossly thickened and the normal testes or spermatic cords could not be felt separately. The regional inguinal nodes were not enlarged. Lower limbs were normal. System examination was normal. Complete blood count and serum biochemical parameters were normal. Erythrocyte sedimentation rate was 7 mm/1 hr. Peripheral smear examination did not show filarial parasite. Viral markers were negative for hepatitis, sexually transmitted diseases and Covid. Serum IgG4 level and IgG4/IgG ratio were normal. Due to the extreme size of scrotum, total scrotoectomy was done, with preservation of both testes. The subcutaneous tissue was found to be at least 15 cm thick though the testicular tunics were normal. The spermatic cord and the testes were found unaffected (FIG. 1, Panel B). Scrotoectomy was done after sparing the cords and testes. The testes were then re implanted on to medial part of thighs for preserving the hormonal function. The skin edges were primarily closed. After uneventful post-operative period, the patient was discharged on 5th post-operative day. The excised scrotum was sent for histopathological examination. The gross specimen weighed 11.8 Kilograms. Gross sectioning showed thick edematous subcutaneous tissue (FIG. 1, Panel C).

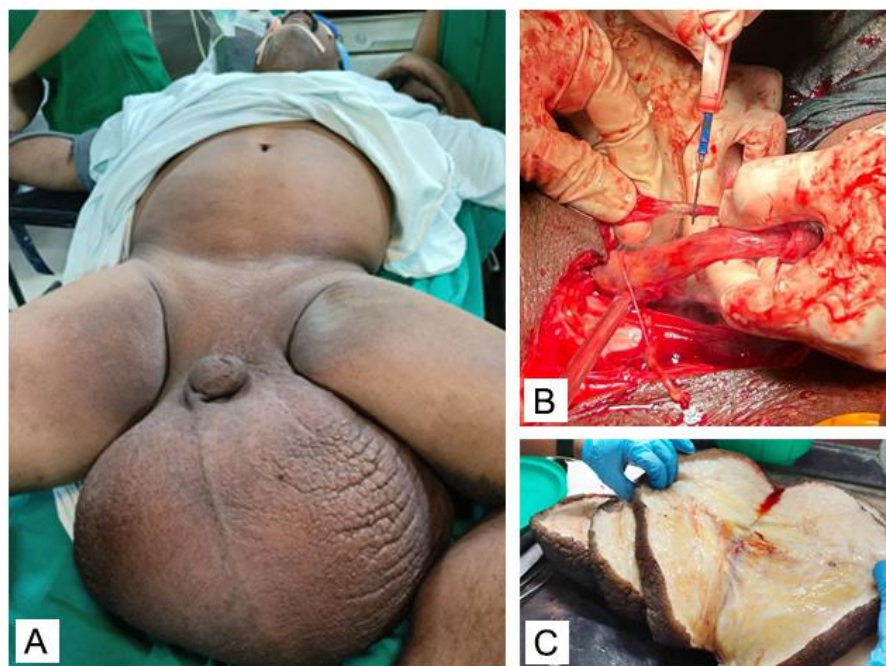


FIG. 1. Panel A-clinical photograph of the case. Panel B- cord and testis are unaffected and is being isolated during surgery. Panel C- Gross section of the specimen showing massive thickening of the subcutaneous tissue.

Histology showed normal skin with edematous connective tissue stroma composed of spindle cells with variable amounts of fibrosis and hyalinization. There was perivascular and interstitial infiltrate of lymphocytes, plasma cells, histiocytes and occasionally scattered eosinophils (FIG. 2, Panel A). Immunohistochemistry (IHC) was negative for SMA, S-100, ALK, B catenin, Calretinin, IgG4, and Ki-67 (FIG. 2, Panels B to G). The findings were consistent of fibrous pseudotumor of the scrotum. Three weeks post operatively, the wound healed completely, leaving mild penile edema which was conservatively managed (FIG. 3). Patient reported great improvement of quality of life including sexual aspects.

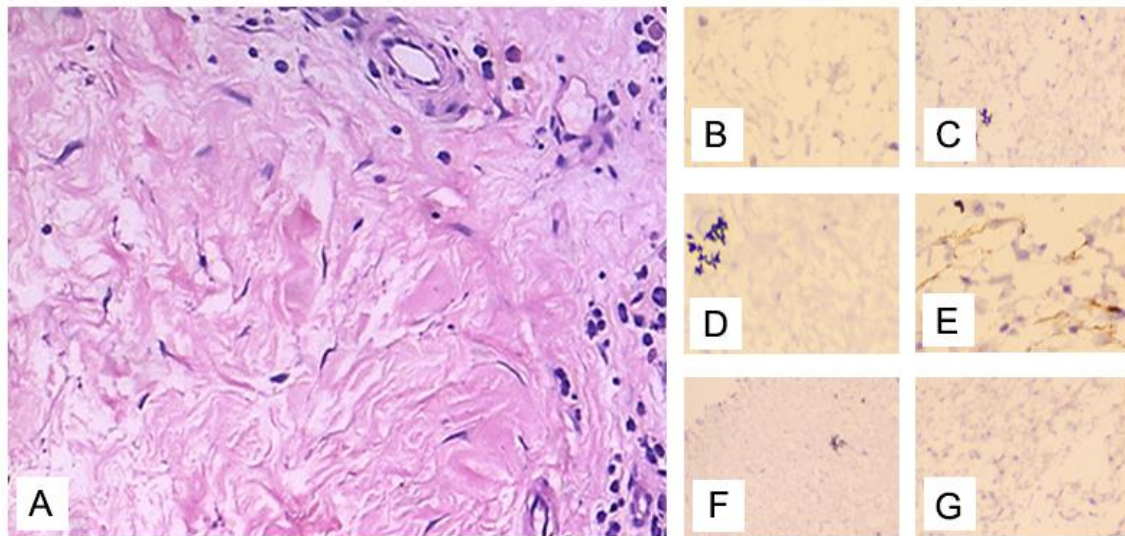


FIG. 2. Panel A- Histopathology showing fibrosis, hyalinisation, and inflammatory infiltrates in the subcutaneous tissue. Panel B, C, D, E, F, G – IHC showing markers negative for SMA, S-100, B catenin, Calretinin, ALK and Ki-67.



Fig. 3. Post scrotectomy wound healing seen at base of penis at 3 weeks after surgery.

3. Discussion

It is widely known that the commonest acquired cause of genital elephantiasis is infective lymphedema. In filarial-endemic regions, infestation by *Wuchereria bancrofti* or *Brugia malayi*, transmitted by infected mosquito enters human lymph vessels, blocks the lymph nodes and lymphatics resulting in lower extremity and scrotal elephantiasis. Histologically, identification of filarial parasite in the dilated lymph vessels is pathognomonic of this condition. Inguinal lymph node infection by sexually contracted *Chlamydia trachomatis* (Lymphogranuloma venereum- LGV) is another cause of infective scrotal lymphedema that can reach massive sizes similar to filarial scrotum. Massive localised lymphedema (MLL) is a non-parasitic lymphedema recognised in morbidly obese individuals in which lymphedema could affect isolated body areas such as abdomen, thigh, scrotum etc. [1]. When the histology reveals only fibrosis and inflammation without lymphatic dilatation, lymphedema cannot be attributed, making specific diagnosis difficult.

Histological features observed in such cases include epidermal thickening, dermal fibrosis, chronic inflammation with edema, and smooth muscle hyperplasia. Pathologist's diagnoses in these non-specific scrotal elephantiasis are 'nonspecific fibrosis', 'fibrous pseudotumor', 'fibrosing dermo epidermal hypertrophy', 'pseudo sarcoma' etc. [1-3]. Various Immunohistochemistry (IHC) markers help to rule out malignant changes. IgG4 is an important marker where positive cases benefit from steroid therapy. IgG4 positivity requires 10 or more IgG4 positive cells per high power field, IgG4+/IgG+ cell ratio greater than 40% and Serum IgG4 concentration greater than 135 mg/dL [4]. Reported cases of fibrous pseudotumor and IgG4 related diseases affects the testicular tunics sparing the skin and the subcutaneous tissues. In our case, IgG4 related disease was ruled out by serum studies and IHC. Due to the extensive fibrous elements seen, the final diagnosis in our case was made as Fibrous Pseudotumour. Treatment of idiopathic cases of scrotal elephantiasis is surgical excision [5]. Wound coverage is variably done with local flaps or skin grafts. Testes are either preserved in the residual scrotal flap or in the groin in selected cases particularly in the young patients.

4. Conclusion

Even though most common cause of scrotal elephantiasis is parasitic lymphedema, our patient had non-parasitic, non-lymphatic non-specific idiopathic scrotal elephantiasis disease, which was diagnosed as Fibrous Pseudotumour of scrotal wall. Surgical excision of scrotum with sparing of testis and primary wound closure was effectively done in this case with good results.

5. Disclaimer

We have obtained consent from the patient to use the clinical photographs, the gross specimen and histology images for publication. There are no conflicts of interests.

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