

## Vulvar Elephantiasis

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### Abstract

Lymphangiectasia is a secondary phenomenon that results from obstruction of previously normal lymphatic systems leading to dilation of the lymphatic vessels. Its occurrence in women is very rare. We report the clinical case of vulvar lymphangiectasia observed in a patient seen in gynecological consultation. The symptomatology was dominated by the appearance of vulvar swelling that had been evolving for 6 months and gradually increased in volume; the patient benefited from an exploration assessment which came back in favor of vulvar lymphangiectasia and was referred to a dermatological consultation for further treatment.

**Keywords:** Lymphangiectasia, lymphatic systems, gynecological consultation.

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## INTRODUCTION

Abnormal lymphatic dilation can be either a primary structural (anatomical) abnormality, known as lymphangioma, or a secondary event following lymphatic obstruction caused by various processes, known as elephantiasis.

Vulvar skin, according to a review of the literature, is particularly sensitive to the formation of lymphangiectasia.

The diagnosis of vulvar lymphangiectasia is most often evoked clinically and confirmed by histological or radiological exploration.

The CO2 Laser remains the most effective therapeutic method according to several studies.

## CLINICAL CASE

This is Mrs. S.K aged 60, single; nulligest; menopausal for 5 years who consults for an oblong

swelling in the suprapubic region and the left labia majora. This training had been evolving for 6 months and had gradually increased in size over the last two months. Furthermore, the patient does not report the occurrence of infectious complications. She had no gynecological history of neoplastic or infectious diseases (notably tuberculosis), nor functional or inflammatory colopathy (Crohn's).

The somatic examination found a patient of normal build (BMI: 24) The skin examination revealed:

- the presence of a roughly oval erythematous swelling covering the entire left vulva of firm consistency, measuring approximately 10 CM; of major axis; painful when pinching with an orange peel appearance
- gynecological examination: not done; virgin patient
- presence of erythematous plaques in the submammary folds; inguinal folds and hypogastric folds



**Figure 1: clinical images of vulvar swelling**

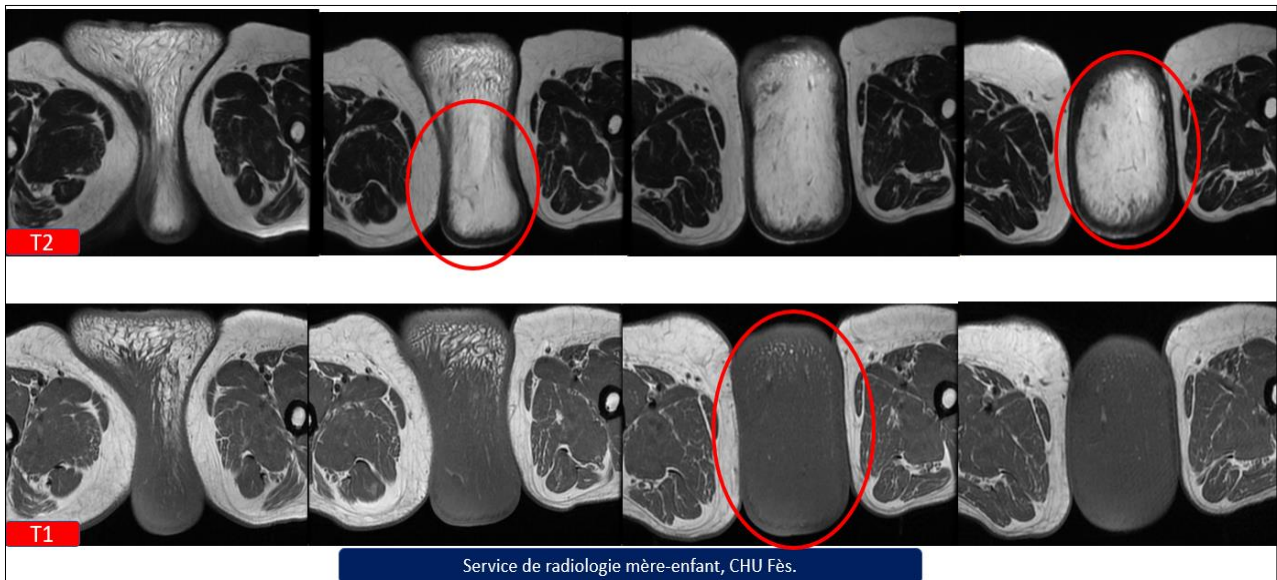
Diagnoses mentioned after dermatological opinion:

- Idiopathic acquired lymphangiectasia associated with candidal and dermatophytic infection
- Cutaneous manifestation of Crohn's disease associated with candidal and dermatophytic infection

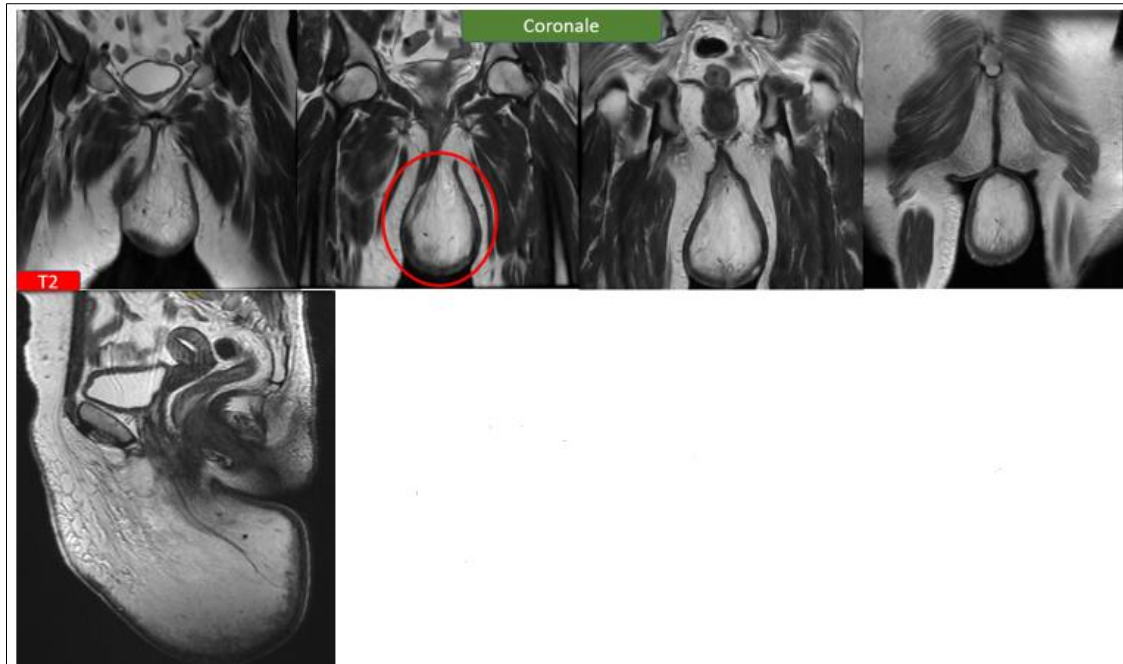
An infectious etiological assessment including a blood count, a search for Koch bacillus in the sputum

and urine, a chest x-ray and an abdominopelvic ultrasound looking for images suggestive of urogenital tuberculosis was negative.

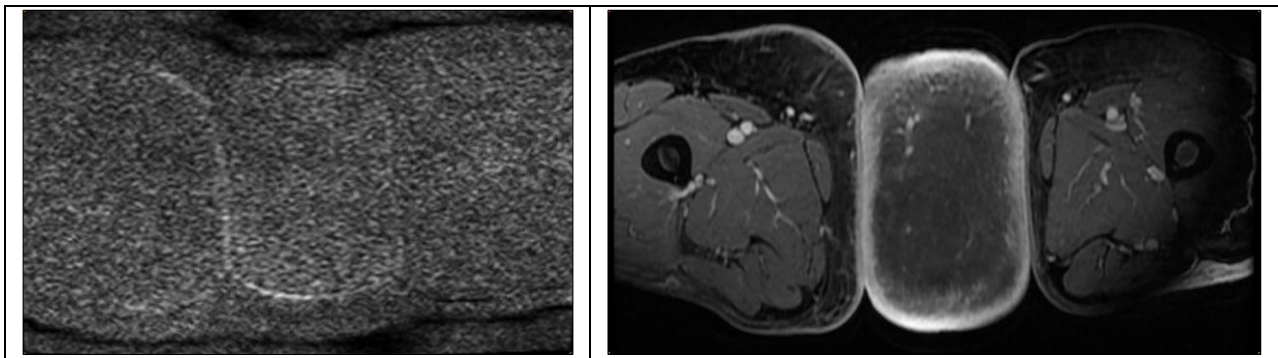
Furthermore, an assessment of the swelling was carried out using a pelvic MRI: which revealed significant hypertrophy and infiltration, first suggesting left perineal and vulvar lymphangiectasia without pathological enhancement of 15 x 13 x 6cm (figure 2,3,4).



**Figure 2: Axial pelvic MRI carried out weighted in T1, T2 showing significant hypertrophy and perineal infiltration and of the left right lip with exophytic development described in T1 hyposignal, frank T2 hypersignal**



**Figure 3: Coronal and sagittal sections of the T2-weighted pelvic MRI showing significant perineal hypertrophy and infiltration and the left right lip with exophytic development described in frank T2 hypersignal.**



**Figure 4: Diffusion sequences and after gadolinium injection, showing a perineal mass and the left right lip with exophytic development, non-restrictive in diffusion and discreetly enhanced after gadolinium injection.**

After diagnostic confirmation, the patient was referred to dermatology with the therapeutic decision of CO<sub>2</sub> laser sessions, associated with a strict dietary regime.

Note that the patient was put on treatment for her mycotic infection.

## DISCUSSION

Vulvar lymphangiectasias are lymphatic dilatations secondary to lymphatic stasis. They are rare, 27 cases have been recorded in the literature [1]. They affect women aged 22 to 75 years with an average age of 48 years [1], and sometimes produce a pseudocondylomatous appearance [1, 2].

Histological examination helps correct the diagnosis. These lymphangiectasias most often occur following therapeutic treatment (surgery, radiotherapy)

of squamous cell carcinoma of the cervix, or following inguinal tuberculosis or Crohn's disease [1-3].

Vulvar lymphangiectasias are often complicated by infectious episodes [1]. Malignant transformation of these lesions has never been observed [8].

Treatment of this disease is extremely difficult. Various therapies for vulvar lymphangiectasia have been proposed but none have demonstrated their effectiveness. In the case of lymphangioma (primary structural damage), it is emphasized that the disease recurs if the entire depth of the abnormal lymphatics is not removed. Secondary lymphangiectasias are more problematic because the obstruction is likely diffuse and complete surgical removal is probably impossible. Although laser vulvectomy was performed in some patients, but the authors were not optimistic about the long-term results.

John's Institute of Dermatology identified three women with vulvar lymphangiectasia and two women with vulvar lymphangioma circumscriptum treated with CO2 laser. Patient tolerance of the procedure was good. Healing was complete within one month and occurred without change in skin texture. All patients reported considerable improvement in their symptoms. Focal recurrence and a localized area of persistence were noted in both lymphangioma patients. Therefore, CO2 laser therapy for vulvar lymphangiectasia and circumscribed lymphangioma is effective and well tolerated; the latter may possibly be more resistant to treatment than the former [1-8].

## CONCLUSION

Vulvar lymphangiectasias are lymphatic dilations secondary to lymphatic stasis. It is a rare benign pathology whose malignant transformation has never been observed. The diagnosis is evoked clinically; and confirmed histologically or radiologically. Various therapies for vulvar lymphangiectasia have been proposed: electrocoagulation, application of liquid nitrogen, surgery and CO2 laser which represents the treatment of choice with good clinical progress.

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