

# Unilateral vulval swelling: What is the diagnosis?

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A 17-year-old unmarried female presented with asymptomatic painless swelling over left side of vulva for three months. Initially small, it gradually progressed to the present size with no spontaneous bleeding or oozing. There were no constitutional symptoms. There was no history of tuberculosis, sexual contact, trauma or abdominal/genital surgery.

On examination, there was pedunculated, convoluted, flesh colored, smooth and non warty mass, with stalk length of approximately 10cm and globular mass of 3x3 cm, hanging from left side of labia majora. It was firm, non tender and local temperature over swelling was not raised. There was

serosanguinous discharge on puncturing it. There was no evidence of inguinal lymphadenopathy. There was no edema of lower extremities and vulva. No abnormality was detected in perianal region.

Routine investigations like hemogram, Chest X ray, USG abdomen, ELISA for HIV, VDRL, TPHA were normal. As there was no apparent cause of lesion, excision biopsy was done and it was subjected to histopathological examination.

## WHAT IS YOUR DIAGNOSIS?

[Figures 1-4]



Figure 1: Convoluted, pedunculated mass present over left side of labia majora



Figure 2: Convoluted, pedunculated mass present over left side of labia majora

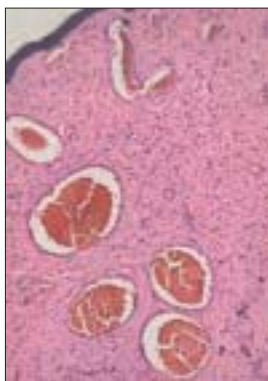


Figure 3: Histopathology- H and E staining (100x)

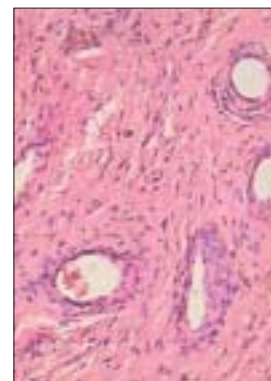


Figure 4: Histopathology- H and E staining (100x)

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## Diagnosis: Lymphangiectasia with Lymphedema (Acquired Lymphangioma)

On histopathological examination there was sparse superficial perivascular lymphocytic infiltrate with marked dilation of several lymphatics throughout the dermis. The reticular dermis showed several thickened and dilated larger lymphatics. Overlying epidermis showed mild spongiosis. The dermis showed abundant fibroplasia around dilated and thickened lymphatics.

These findings were consistent with diagnosis of Lymphangiectasia with lymphedema.

## DISCUSSION

Lymphangiectasia is a benign condition with multiple dilated lymph vessels in the dermis whereas Lymphangioma are distended lymphatics due to structural abnormality of tumor like nature.

Vulval Lymphangiectasia is a rare disease, usually acquired or secondary and not as a result of congenital lymphatic malformation. It is due to obstruction to drainage which leads to back pressure and dermal backflow with subsequent dilatation of surface lymphatics. It usually presents with circumscribed groups of tense, thin walled vesicles. It is reported following 7-15 years after ilioinguinal block dissection or pelvic surgery and radiotherapy for gynecological cancer.<sup>[1]</sup> In the case discussed, a 17-year-old girl presented with solitary, smooth, flesh colored, pedunculated, convoluted mass of three months duration only. The patient was investigated for secondary causes like TB, Crohn's disease and cervical or ovarian cancer. There was no history of genital/abdominal surgery or radiotherapy in this patient. There was no lymphedema of corresponding limb and no sign and symptoms of inflammation of vulva suggestive of recurrent infections and cellulitis which are common complications. The diagnosis was considered acquired lymphangioma on clinical and histopathological grounds but there was no obvious cause. Advanced investigation like lymphoscintigraphy and lymphangiography are reported to be useful for providing information regarding anatomy of lymphatics and any abnormal lymphatic flow pattern to identify the cause of lymphangiectasia.<sup>[2]</sup> However, they are not easily available.

Huilgol *et al.* reported that iatrogenic cases following surgery and radiotherapy for cancer appear to be most common, but many cases are idiopathic.<sup>[3]</sup>

Okur *et al.* reported a case of a 27-year-old woman with vulval lymphangiectasia with ipsilateral congenital

lower extremity lymphedema which appeared at age of 19 years and described that de novo acquired lesions without lymphatic disturbance are less frequently seen. However, the condition may arise without adjacent lymphedema.<sup>[1]</sup>

Buckley and Barnes reported a 35-year-old patient who had lymphedema since the age of 14 and underwent two cellulitis attacks. Thereafter, lymphangiectasia on the vulva appeared in the same year.<sup>[4]</sup>

## TREATMENT

The patient was referred to the gynecological department for excision of the mass. Stitches were removed after seven days and complete healing with reepithelialisation and resolution of postoperative pain and swelling occurred within one month. The patient was instructed to come back in case of any recurrence.

Excisional surgery and carbon dioxide laser are two major treatment modalities. Carbon dioxide laser vaporizes the lesion superficially, seals the underlying lymph vessels and diminish the recurrence. The vesicular and papular lymphangiectatic lesions and verrucous plaques are ablated to the level of visually and palpably normal dermis. The laser therapy can be repeated if any recurrences occur. Delayed wound healing, scars, and even keloid formation may be seen after laser therapy.<sup>[5]</sup>

Excisional surgery eliminates the abnormal subcutaneous lymph vessels and cisterns, and corrects the aesthetic appearance of the edematous vulva. It is the only plausible choice in the therapy of advanced disease. Cryotherapy, electrocoagulation, and sclerosing agent injection are other modalities of treatment.<sup>[6]</sup>

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