

Lymphangioma circumscriptum of vulva

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Abstract

Lymphangioma circumscriptum in vulva is an uncommon encounter in Dermatology clinics. It presents as a blistering eruption in vulva. It can be easily mistaken for genital Herpes Simplex. When the lesions are infected and ulcerated it makes clinical diagnosis difficult.

We report a case of lymphangioma circumscriptum of vulva in a 48 year old female who presented with recurrent pain, oedema, redness and ulceration. She developed septicemia on one occasion needing ICU care.

Clinical and histological features of lymphangioma circumscriptum and treatment options are discussed below.

Introduction

Lymphangioma circumscriptum is a benign condition. It is usually seen in children and young adults commonly in trunk, axillary folds and proximal limbs. It is more commonly seen in females. Vulval lesions are uncommon. Vulval lesions clinically present as clear fluid filled vesicles arranged in a herpetiform pattern. Recurrent lymphorrhoea and secondary infections can occur.

Case report

Otherwise healthy 48 year old female presented to the Dermatology clinic CSTH with a history of blister like lesions in vulva for 20 years duration. The number of lesions gradually increased with time. She gives a history of on and off watery discharge.

She also complains of recurrent pain, redness of the lesions. During these episodes the vesicles have been ulcerated. On one occasion she needed ICU care following severe infection and septicemia. She developed edema of vulva with each episode of infection which gradually increased with time. She does not give a history of abdominal or pelvic surgeries.

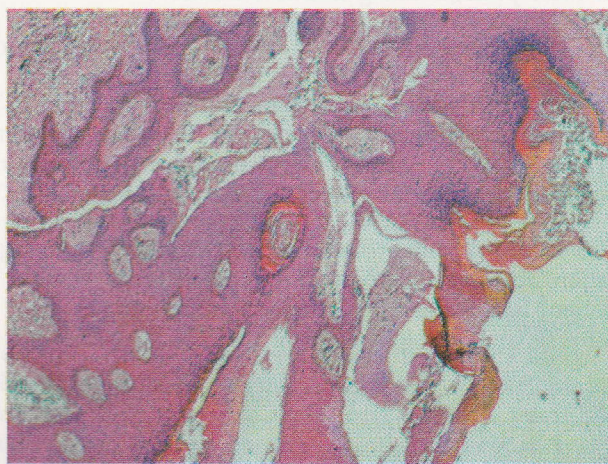
Examination of vulva revealed multiple clear vesicles in both sides of vulva. The vesicles are arranged in herpetiform pattern. They are firm and some vesicles are discrete. Vulval edema was noted. There was no ulceration.

Abdominal examination is clinically normal. No lower limb edema.



Multiple clear fluid filled vesicles in vulva with edema.

Histology shows cystically dilated lymph vessels lined by single layer of endothelial cells in upper and mid dermis. Some dilated lymph vessels appear enclosed in the mid dermis. No atypical cells were observed. Epidermis shows acanthosis, papillomatosis and hyperkeratosis.



Histology showing dilated lymph vessels.

She was started on prophylactic oral penicillin 500mg bd to prevent recurrent infections, and was referred for surgery.

Discussion

Lymphangioma circumscriptum results from superficial lymphatic malformations localized to skin subcutaneous tissues and muscles. It has been postulated that the original malformation arises from deep contractile vessels. Lymphatics draining to these deep channels dilate and form lymphatic cisterns. Rarely there can be extensive deeper involvement.

The dilated lymphatics clinically manifests as fluid filled clear vesicles which bulge on skin surface. The vesicles are usually well defined and discrete or may be grouped into structures resembling frog spawn. It can become warty with time resembling viral warts.

The common sites are axillary folds, shoulders, flanks and proximal limbs. Vulval lesions can occur following recurrent cellulites Crohn's disease and following radiation therapy and is accompanied by lymphoedema. It has also been reported in patients without previous lymphatic damage.

When limb swelling is present extensive underlying lymphatic abnormalities should be excluded.

Squamous cell carcinoma has been reported arising within lymphangioma circumscriptum.

Treatment of choice for lymphangioma circumscriptum is radical surgery. Wide excision assuring free lateral and deep margins. But recurrences are common especially when there are involved margins. Surgery will not be feasible on certain sites. Intralesional sclerosants can be used. Vaporization with carbon dioxide laser has shown good results. Superficial X rays has been used successfully.

References

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