Lymphangioma circumscriptum of the vulva
Clinical picture and surgical management

Vulvar lymphangioma circumscriptum is a rare entity that may mimic many other diseases of the vulva. It presents with the non-specific symptoms of persistent vulvar itching and soreness, and the diagnosis is confirmed through vulvar biopsy. Surgical treatment has the lowest recurrence rates compared with other treatment modalities. The case is presented here of a woman diagnosed with lymphangioma circumscriptum of the vulva and its surgical management.

Lymphangioma circumscriptum (LC) is a benign disorder of the lymphatic system that involves the proliferation of both the deep dermal and subcutaneous lymphatic channels. Its most common sites are the axillae, chest and oral cavity, including the tongue. Vulvar LC is rare, with only 75 cases reported in the literature. The case is presented here of a woman with bilateral vulvar LC diagnosed on vulvar biopsy and subsequently treated with surgical excision of the lesions.

CASE PRESENTATION

A 63-year-old null gravida woman was referred to the University Hospital with symptomatic biopsy-proven LC of the vulva. The woman had a 6-month history of severe vulvar symptoms which she described as a sense of pricking, burning, and soreness that were made worse with exercise and walking. She also reported vulvar itching which she tried to relieve by scratching. A vulvar punch biopsy, taken by her primary health provider, confirmed the diagnosis of LC. After 6 months of expectant management she was referred to the vulvar diseases center for further management of the persistent and deteriorating symptoms.

Careful inspection of the vulva revealed an area of clusters on the right labium majus consisting of small, vesicular-looking, clear-filled, globular structures. A similar, smaller cluster was observed on the left labium majus (fig. 1). Cotton swab testing demonstrated pain within 1 cm of these areas. In view of her worsening symptoms, the patient was referred for surgical excision.
the patient consented to wide local excision of the two affected areas. The possibility of recurrence was addressed.

The patient underwent wide local excisions of the lesions with a 1 cm margin. Histopathology confirmed complete excision of the diseased areas. Post-operatively the patient’s symptoms were alleviated. She was seen again in our clinic three years following the wide local excision because of urine leakage. She is currently asymptomatic with significantly improvement in her quality of life, despite a small (1 cm) area of recurrence on her vulva.

DISCUSSION

LC is clinically characterized by translucent clear fluid filled vesicles measuring <5 mm in diameter that resemble frogspawn. The lesions may be totally asymptomatic or accompanied by pain and itching. The diagnosis is confirmed by histopathological examination which reveals dilated lymphatic channels in the dermis. Due to similarities in clinical appearance, LC may mimic many other diseases that need to be excluded, including vulvar tumours and infective diseases such as herpes infection, genital warts and molluscum contagiosum.

Although there is no consensus on the treatment of LC, surgical excision is usually considered as the first-line treatment. Because LC involves benign proliferation of the lymphatic channels within the deep dermis, there is a recurrence rate of 17% even after complete surgical excision. Other treatment modalities include laser ablation, with a reported recurrence rate of up to 60%, sclerotherapy, and radiofrequency ablation.

In the case reported here, the decision for surgical excision was taken because of the deteriorating symptoms. Although histopathological examination confirmed the complete excision of the lesions, there was a small recurrence of LC on the vulva, but the patient is symptom-free three years after surgical excision.

ΠΕΡΙΛΗΨΗ

Περιγεγραμμένο λεμφαγγείωμα αιδοίου: Κλινική εικόνα και χειρουργική αντιμετώπιση

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Περιγράφεται η περίπτωση γυναίκας ασθενούς η οποία διαγνώστηκε με περιγεγραμμένο λεμφαγγείωμα αιδοίου, καθώς και η χειρουργική της αντιμετώπιση.

Δέλτα ευρετηρίου: Αιδοιεκτομή, Αιδοίο, Ευρεία τοπική εκτομή, Περιγεγραμμένο λεμφαγγείωμα

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