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Congenital vulvar Lymphangioma Circumscriptum: an Unusual Benign Pathology in Young Female

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Received: September 9, 2015; Accepted: September 14, 2015; Published: September 20, 2015

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Abstract

A female patient, aged 17 years, presented with papule-like condyloma of the labia majora. After major labiaectomy was performed, symptoms improved. Histology revealed lymphangioma circumscriptum of the vulva. Simple vulvectomy is an effective therapy for vulvar lymphangioma circumscriptum. It is a rare benign pathology that should be kept in mind in any vulval condylomatous lesion.

Introduction

The lymphangioma circumscriptum is a benign disorder of lymphatic origin that can be either congenital or acquired, involving the skin and subcutaneous tissues. Acquired vulvar lymphangioma circumscriptum is a rare disease and known to occur after radical hysterectomy with or without adjuvant radiation therapy for cervical cancer [1-3]. It is caused by damage to previously normal lymphatic channels, due to cancer treatment [4].

We report a case of congenital vulvar lymphangioma circumscriptum with surgical management.

Case Report

17 years old, virgin, regularly menstruating, non-Circumcised, complaining of vulval swelling and pruritus vulve since 7 months. Examination showed a left labial hyperkeratotic, non-pigmented and non-tender, plaque like lesion, about 5 cm with diffuse borders. Punch biopsy from the vulva revealed Lymphangioma Circumscriptum (LC). Ultrasound was Unremarkable and CT showed moderately enhancing vulval lesion. Simple vulvectomy, with 1 cm safety margin and 1ry closure. Pathology confirmed the preoperative diagnosis of LC with no malignancy. Surgical resection margins were free of lesion changes (Figures 1 and 2).



Figure 1: Left labial swelling



Figure 2: a: Simple vulvectomy, b: The excised specimen, c: Postoperative picture after 2 weeks with proper healing

Discussion

The vulvar lymphangioma circumscriptum is benign but may cause secondary infections with symptoms as edema and papules of the labia majora and oozing lymph fluid. There is no standard management. The treatments range from conservative treatment as manual lymph drainage and compression, to abrasive therapy as sclero-therapy, electrocoagulation, laser-therapy with CO₂, and surgical excision [5,6]. While lesion recurrence is frequent, surgical excisions were preferred in 19 of the 37 cases [7,8]. The recurrence rate after surgical management was 23.1%. The recurrence rate is twice in lymphangioma circumscriptum without surgical management [5]. Browse et al. reported that the recurrence rate after radical excision was high, when the initial lesions were greater than 7 cm diameter as compared with lesions less than 7 cm diameter for which a local excision was performed [9].

We propose that simple vulvectomy is an effective therapy for vulvar lymphangioma circumscriptum. To obtain a successful result, it is important to completely excise all the papules with a sufficiently deep margin of subcutaneous tissue. In an extensive vulvar lymphangioma circumscriptum, it is impossible to remove the lesion without a skin flap.

Recently, laser therapy with CO₂ has been selected as an effective treatment. This treatment is good for small, superficial lesions. We suggest that the indication of major labiaectomy is 1) the large diameter and deep lesions of vulvar lymphangioma circumscriptum, 2) severe symptoms such as more papules and vesicles, a serious lymph oozing, greater edema, pain, fever with infection, 3) therapeutic failure after conservative treatment or CO₂-laser therapy [10].

Conclusion

In conclusion, simple vulvectomy seems to be effective for vulvar lymphangioma circumscriptum and beneficial for the patients' quality of life.

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Citation:

Ahmed Samy EL-Agwany and Ahmed El-habashy (2015) Congenital vulvar Lymphangioma Circumscriptum: an Unusual Benign Pathology in Young Female. *Med Clin Res* 1: 1-2.