CASE REPORT

Congenital Lymphangioma Circumscriptum of the Vulva

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Manuscript received: August 29, 2007; Initial review completed: September 21, 2007; Revision accepted: May 19, 2008.

Lymphangioma circumscriptum of the vulva is a disorder of lymphatic channels involving deep dermal tissues. Most of these cases are confused with genital warts leading to improper diagnosis and treatment. We present a three years young female child who had multiple skin colored papular lesions over the genitals. Skin biopsy revealed features of lymphangioma circumscriptum.

Key words : *Child, Lymphangioma, Vulva.*

Lymphangioma circumscriptum is a rare benign disorder of no specific etiology involving the lymphatic channels in the deep dermal and subcutaneous layers(1). It can occur as either a congenital abnormality or as acquired damage to previously normal lymphatic channels(2). Axilla, adjacent chest-wall, oral cavity and tongue are common sites. The vulval presentation is uncommon(1-3). Lymphangioma circumscriptum is characterized by clustered or diffuse thin walled, translucent vesicles 1 to 5mm in diameter and filled with clear lymphatic fluid. The vesicles may develop on normal skin or on top of preexisting papules; hyperkeratosis may sometimes give them a verrucous appearance.

CASE REPORT

A three year old girl was referred for evaluation of some eruptions of multiple skin colored papular lesions on the genitalia since birth. There was neither any preceding history of local trauma nor any drainage of fluid from the site and of sexual assault too. The site of involvement had progressively become more extensive in the last four months. On examination, the right labia majora was swollen, hypertrophied and hyperpigmented. There were multiple verrucous, coalescent papules (***Fig.* 1**). Many of the papules had focal sites of dark red to purple discoloration (i.e. hemorrhage) within them. No excoriation, crushing or oozing was observed. The remaining genital and physical examination was normal. A nontreponemal test for syphilis was non-reactive. The Mantoux test was negative. Total and differential WBC counts were within normal limits. Erythrocyte sedimentation rate was 10mm/hr. (Westergren). There was no evidence of pulmonary tuberculosis in a chest radiograph. In view of clinical presentation possibility of genital warts, lymphangioma circumscriptum, cystic hygroma was kept. A skin biopsy of a representative lesion revealed the features of lymphangioma circumscriptum, namely, dilatation of the lymphatic channels in the papillary dermis with overlying epidermal acanthosis and hyperkeratosis.

DISCUSSION

Vulva is an uncommon site for the development of lymphangiomata. Only 32 cases have been described

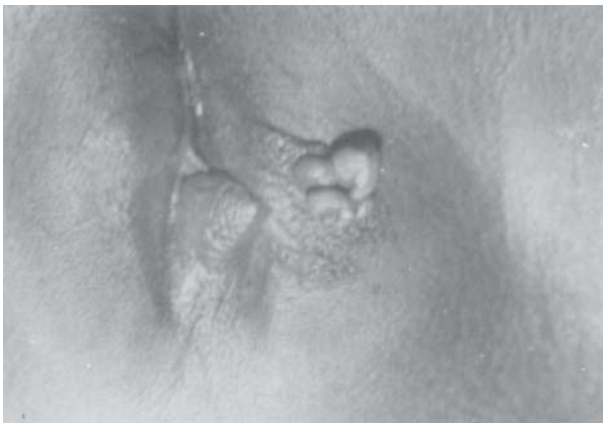

FIG.1 *Lymphangioma circumscriptum of the vulva showing verrucous, coalescent papules.*

previously (12 congenital and 20 acquired)(1,3). The youngest age of presentation of congenital lymphangioma circumscriptum of the vulva, till date, had been 14 years(4). Our case is probably the youngest in terms of age of presentation of lymphangioma of the vulva. Frequent complications of lymphangioma circumscriptum include swelling of the vulva, pain, recurrent cellulitis caused by excoriation or spontaneous vesicles, and subsequent infection of the vulvar area(1). The added complication of psychosexual dysfunction may lead to cessation of sexual activity. A rare major complication is lymphangiosarcoma arising at the site of a preexisting lymphangioma circumscriptum(1).

Treatment modalities include sclerotherapy, electrocoagulation, liquid nitrogen therapy, carbon dioxide laser therapy, radiotherapy and surgical excision(2,3,5).

Lymphangioma circumscriptum of the vulva may be clinically indistinguishable from molluscum contagiosum, genital warts or tuberculosis verrucosa cutis. Biopsy is essential to confirm the diagnosis.

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