

Lymphangioma circumscriptum of the vulva and Klippel-Trenaunay syndrome

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Sir,

Lymphangioma circumscriptum is a rare vascular malformation affecting the lymphatic vessels of the papillary dermis [1]. Klippel-Trenaunay syndrome is a congenital vascular malformation characterized by venous varicosities, soft tissue and/or bony hypertrophy [2]. Here we report a case of association of Lymphangioma circumscriptum of the vulva and Klippel-Trenaunay syndrome.

A 42-year-old female, who has been diagnosed as Klippel-Trenaunay syndrome (KTS), presented with oozing and chronic lesions over the right labium majus that has been present for more than 6 years diagnosed initially as genital warts. The dermatological examination noted the presence of a group of multiple red-purple papules and verrucous vesicles over the right swollen labium majus (Fig. 1). Dermoscopy showed multiple lacunae surrounded by pale septa (Fig. 2). Meanwhile, her right limbs were covered by a large geographic telangiectasia (Figs. 3a and 3b). Histopathology of labium majus lesion revealed large, irregular and dilated thin-walled lymphatic channels and endothelial vasculatures in the papillary and reticular dermis without dysplasia or signs of Human Papillomavirus infection. Finally, a diagnosis of LC associated with KTS was confirmed. Our patient was treated by surgical excision with good improvement. Our case is quite particular by demonstrating the interest of dermoscopy in differentiating lymphangioma circumscriptum from genital warts, angiokeratomas and molluscum contagiosum. The presence of lacunae defined as roundish or oval clustered, well-demarcated, yellowish, reddish or dark-coloured structures which



Figure 1: Lesions of lymphangioma circumscriptum.



Figure 2: Dermoscopy [non polarized, 10X] revealed multiple lacunae surrounded by pale septa.

orient us to lymphatic malformation [3]. In their study of 45 cases, Zaballos and al demonstrated that lacunae

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Figure 3: (a and b) Large port-wine stain on most of the right lower et upper limb.

are the most specific dermoscopic features [4]. Genital warts are also characterized by vascular structures but they are red loops or dots surrounded by a white halo [5]. Here we report a case of association of vulvar lymphangioma circumscriptum and Klippel-Trenaunay syndrome oriented by dermoscopy.

Consent

The examination of the patient was conducted according to the Declaration of Helsinki principles.

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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