



Chronic Recalcitrant Lesions of an Enlarged Limb: Consider Parkes Weber Syndrome

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Editorial

Parkes Weber syndrome represents a very rare combined congenital vascular malformation that can lead to chronic leg ulcers in young age groups [1]. We report the case of a patient with dragging leg ulcerations treated as venous ulcers in adulthood.

This is a 56 year old patient, with no notable pathological history and no similar cases in the family, who presents since birth an enlarged right lower limb without pain or other associated signs, followed for 2 years for stagnant and recurrent ulcerations of the leg spreading poorly to treatments. The dermatological examination found an enlarged right lower limb with a reduced calibre at the ankle with sclerotic skin giving an inverted bottle appearance. At the perimalleolar level, there were 3 hollow rounded ulcerations with irregular edges and a fibrinous background topped by hemorrhagic crusts (Fig. 1). The echodoplasty was difficult to interpret due to the importance of the hypertrophy. The angioscanner showed a hypertrophic artery Tibialis anterior and arteria tibialis posterior of the right leg, both of which were responsible for feeding the underlying ulcer, with arteriovenous shunts confirming the diagnosis of Parkes Weber. Compression therapy with short stretch bandages was recommended for the patient. The patient was referred to vascular surgery for possible embolization of the arteriovenous shunt.

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In this case, we diagnosed the patient with Parkes Weber syndrome. This syndrome is considered by some experts as a sub-form of the Klippel-Trenaunay syndrome. Both syndromes are defined by congenital vascular malformations, which can affect the capillary system as well as the venous or lymphatic system [2].

PWS is a congenital angiodysplastic syndrome combining capillary, venous, lymphatic and arteriovenous shunts in the overgrown limb. It can affect the upper or lower limbs. It can affect the upper or lower limbs, including the pelvic vessels. Most of these patients are already diagnosed in childhood or at puberty [3]. In contrast, in our patient's case, the disease did not present until he was 36 years old. The abnormal increase in flow in the venous vasculature resulted in venous hypertension. As a result, the patient developed chronic venous insufficiency with increasing age. Simultaneously, the areas located distally around the AV shunts suffered from arterial under excitation. The combination of these two mechanisms resulted in decompensation of the microcirculation, which then led to local hypoxia that subsequently led to chronic leg ulcers that were difficult to treat. Hyperpigmentation, such as yellow ochre purpura, and hypopigmentation, such as white atrophy, developed as typical clinical signs [3,4].

The treatment of the ulcer consists of correcting the arterio-venous shunt and adopting venous compression therapy by short-stretch bandages in parallel with ulcer-specific dressings [5]. In addition, considerable attention to both arterial and venous components would be the best approach for Parkes Weber syndrome. Cases of complete regression after coil embolization of AV shunts have been reported [6].

In conclusion, Parkes Weber syndrome presents a rare cause of chronic wounds. Nevertheless it should always be considered, even in patients of an advanced age. During therapy, both arterial and venous components should be kept in mind and treated to achieve the best possible outcome.



Figure 1: An enlarged right lower limb with a reduced calibre at the ankle with sclerotic skin giving an inverted bottle appearance. Presence of ulcerations with irregular edges and a fibrinous background topped by hemorrhagic crusts.

Keywords

Vulvar Lichen Sclerosus; Dermoscope; Vascular Pattern; Non-Vascular Pattern

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Consent

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

Conflict of Interest

The authors declare that they have no conflict of interest.

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