Poster n°7

A port-wine-stain mimicking secondary Raynaud's phenomenon in a phototype V patient: a clinical and videocapillaroscopic evaluation

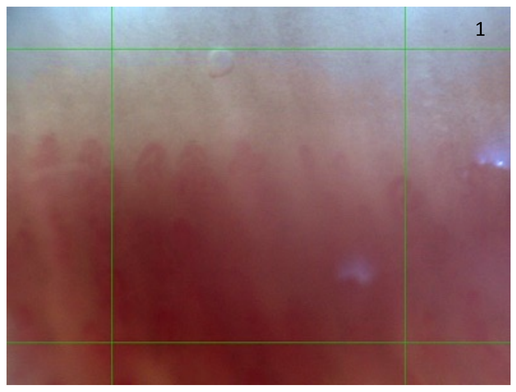
C. Gilmant[1], M. Verheyden[1], V. Badot[2], B. Richert[1], Y. Mostmans[1,3]

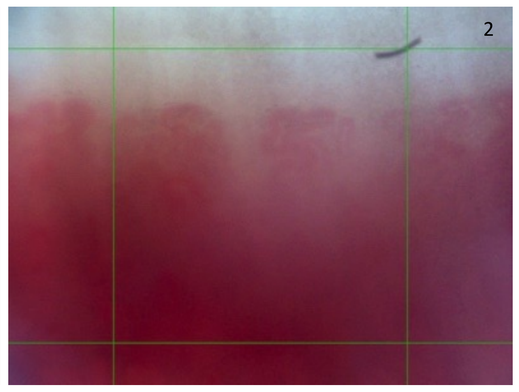
[1]Dept. Dermatology, [2]Dept. Rheumatology, [3]Dept. Immunology, CHU Brugmann, ULB, Brussels, Belgium

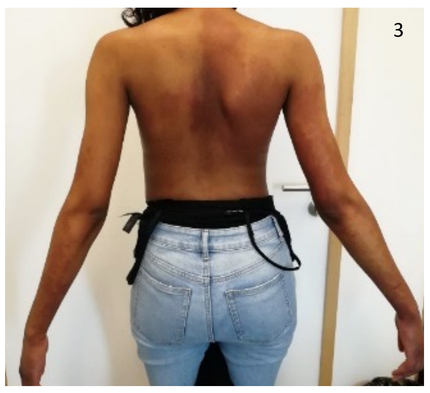
Purpose

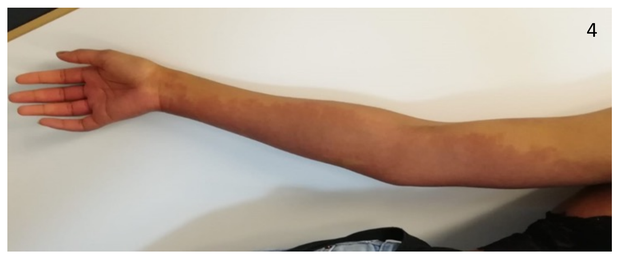
A 33-year-old woman was referred by her general physician to the Immunology Department to be assessed by nailfold videocapillaroscopy (NVC) for complaints of non-painful, intermittent and biphasic (blue and red) Raynaud's phenomenon of the fingers which appeared a few years ago and was induced by cold. The patient had no specific medical history and had no neurological or ophthalmological complaints. The patient was examined and sent for a nailfold videocapillaroscopy.

Results









The nailfold videocapillaroscopy showed: A loss of capillary density, some abnormal shaped capillaries (ramifications), multiple dilated capillaries and a giant one (63 µm) (Fig.1,2). There were no signs of cutaneous sclerosis with a modified Rodnan skin score of 0/51. Nevertheless, physical examination revealed a Fitzpatrick skintype V with extensive discretely violaceous and sharply delineated erythema located in the back and the chest, extending over the entire length of the arms and continuing towards the palmar sides of both hands, which confirms the diagnosis of a Port-wine-stain (PWS, naevus flammeus) (Fig. 3,4). Laboratory testing showed no abnormalities (normal hematology, renal/kidney function) and the antinuclear antibodies were negative.

Discussion

Vascular malformations are a large entity that can affect capillaries, veins, arteries and/or lymphatic vessels. Generally, they are classified as slow flow malformations and fast flow malformations. A PWS is a slow flow capillary malformation in the form of a mosaic disposition of pink to port wine macules. PWS malformations are known to have defective sympathetic innervation. This leads to an abnormal vascular tonus and a decreased perivascular innervation. This can also be seen in the vasospastic changes in Raynaud's phenomenon. These vasospastic changes are also seen in the systemic sclerosis, explaining their similar symptoms and similar impaired autonomic nerve system. There is a lack of capillaroscopy data on slow flow vascular malformations. Our case is the first to show capillaroscopy findings of PWS affecting the chest and the hands with symptoms imitating Raynaud's phenomenon.

Conclusion

In conclusion, non-specific capillary abnormalities on NVC can be seen in PWS extending to the hands, mimicking secondary Raynaud's phenomenon and an early scleroderma pattern. This could be explained by a dysfunctional autonomic nervous system regulation. Although NVC is an easy-to-use and non-invasive visualizing tool for the detection of scleroderma spectrum rheumatic diseases, this case underlines the necessity of a rigorous clinical dermatological examination on all patients suffering from Raynaud's phenomenon.