extensive skin lesions, multiorgan involvement, and even death.[6]

It is important to recognize these cases because, given their greater severity, they should be managed and treated as TEN.

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Acquired Port-Wine Stain: Not a simple stain!☆

Mancha de vino de Oporto adquirida: ¡no es una simple mancha!

Dear editor,

we read with interest a case series of acquired port wine stain (PWS) in 3 otherwise healthy children (2 females and 1 male) by Millán-Cayetano et al.[1] published in Actas Dermo-Sifiliográficas journal. The authors stated ''acquired capillary malformation may be considered simply to be a late-onset capillary malformation with a variable latency period''. Actually, acquired PWS is not as ''simple'' as considered by the authors. The authors underestimated skin diseases masquerading as PWS.

Linear morphea is a form of morphea that can affect an entire extremity and follow the lines of Blaschko. Children are more likely than adults to have linear morphea on the face.[2] In many cases, the affected skin is initially erythematous and may resemble a PWS. Vascular damage, such as microvascular injury, and T-cell activation, with subsequent abnormal collagen production by fibroblasts, is thought to be involved in its pathomechanism.[3] Nihjawan et al.[4] reported four cases that had presented with erythematous vascular-appearing patches resembling PWS. Three lesions were located on the face and one was on the leg. The initial biopsies of two patients revealed telangiectatic dermal vessels, consistent with PWS. However, further biopsies revealed dermal fibrosis with patchy lymphocytic infiltrate, consistent with morphea. Diagnosis of morphea was made approximately 6 months to 3 years after the onset of the acquired PWS. On the other hand, perineural inflammation has rarely been reported to be an early histopathological feature of morphea.[5] Singh et al.[6] reported 2 cases of morphea with subtle sclerotic changes initially, presented with perineural and intraneural lymphoplasmacytic infiltration. According to Nihjawan et al.,[4] there was prominent perineural inflammation which prompted the diagnosis of early morphea. In other words, early inflammatory morphea can present initially with a vascular, nonindurated patch.[7] Biopsies of these lesions may not reveal the characteristic features of established morphea and the diagnosis has to be considered if perineural inflammation is seen.[4] Nihjawan et al.[4] recommended, in patients with acquired PWS, delaying PDL treatment until a diagnosis of early morphea can be excluded.[4] However, it is difficult to ascertain whether laser therapy to the initial lesions triggered the increase in fibrosis as some of the reported cases did not receive laser treatment.[7] Treatment of PWS using the PDL may reduce the skin erythema, but did not prevent subsequent sclerosis.

To sum up, acquired PWS is not a simple stain. Inflammatory morphea should be considered in the differential diagnosis whenever an acquired PWS has been identified, especially on the face.[2] Early stages of morphea are sometimes difficult to recognize, and histology may not helpful in early cases because there is overlap, leading to misdiagnosis. Clinicopathological correlation is of paramount importance in such cases. Morphea should be considered if perineural inflammation is seen in histopathology. Dermoscopy can assist in the early diagnosis of localized scleroderma (LS), with no need for invasive examinations.[8]

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Noteworthy, ultrasound used for LS has demonstrated clear differences from healthy skin and improvement after initiation of treatment.[9]

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Reply to «Acquired Port-Wine Stain: Not a Simple Stain»☆

Réplica a: «Mancha de vino de Oporto adquirida: ¡no es una simple mancha!»

Dear Editor:

We read and considered the letter by Abdelmaksoud and Vestita with interest and greatly appreciated their comments. It is true, as they point out, that morphea is one of the possible diagnoses that must be included in the differential diagnosis for an acquired capillary malformation. In the early stages, morphea may present with erythema and mild sclerosis but not atrophy and, in such cases, may mimic an acquired capillary malformation.[1] There are, however, many other conditions that can simulate an acquired capillary malformation at different stages of its evolution. For instance, such malformations must also be differentiated from diseases such as lupus.[2] The differential diagnosis can also include other vascular anomalies,[3] including arteriovenous and venous malformations, and abortive hemangiomas. Likewise, syndromes associated with vascular anomalies (in our cases capillary malformation-arteriovenous malformation syndrome and Sturge-Weber syndrome) must be ruled out.

Notwithstanding the above, acquired capillary malformation is an independent entity characterized by a specific course and prognosis. It is one thing to say that other conditions can mimic the clinical appearance of capillary malformations and another to say that such lesions are capillary malformations. In the 3 cases we presented, the lesions have remained stable over the course of clinical and ultrasound follow-up for between 3 and 5 years and no changes in appearance or texture have been observed during this period. Furthermore, as commented in our letter, in the first case, given the atypical clinical picture, we performed a diagnostic biopsy to rule out other possible diagnoses and the result was consistent with that of capillary malformation.[4] Our intention was to comment on the definitive diagnosis of the lesions rather than what they might appear to be.

Once we had established the diagnosis of capillary malformation (acquired in these cases), we considered—in line with the prevailing opinion in the literature—that the prognosis was the same as for a congenital capillary malformation. When we asserted that ''acquired capillary malformation may be considered simply to be a late-onset capillary malformation with a variable latency period'' we were not referring to the conditions that should be included in the differential diagnosis but rather to acquired capillary malformations that had been diagnosed as such and not to lesions mimicking the appearance of capillary malformations. In light of that, we stand by our assertion.

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