nosis is essential, as it provides key prognostic information and guides management. It also enables clinicians to offer genetic counseling to patients and parents of affected individuals.[2,4]

Treatment is symptomatic and multidisciplinary, and several protein-, cell-, and gene-based therapies are currently under development for DEB.[1,2,4,9]

In conclusion, the prevalence of AD in our health care setting is similar to that described in previous studies.[10] Most of the patients were female and had been diagnosed at birth. The clinical manifestations were variable, but the most common findings were blisters, nail dystrophy, and pruritus. The main complication was anemia, possibly because of the higher prevalence of DEB. Treatment was largely symptomatic.

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Conflicts of interest

The authors declare that they have no conflicts of interest.

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S. Guillen-Climent[a,*], L. Fernández García[b], A. García-Vázquez[a], J.M. Martín[a,c]
[a] *Servicio de Dermatología, Hospital Clínico Universitario, Valencia, Spain*
[b] *Facultad de Medicina, Universidad de Valencia, Valencia, Spain*
[c] *Departamento de Medicina, Facultad de Medicina, Universidad de Valencia, Valencia, Spain*

* Corresponding author.
E-mail address: santigc85@gmail.com (S. Guillen-Climent).

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Diagnostic Value of Cutaneous Ultrasound in Verrucous Venous Malformation☆

Utilidad de la ecografía cutánea para el diagnóstico de la malformación venosa verrucosa

To the Editor,

Verrucous venous malformation (VVM) is a rare vascular lesion currently classified as a vascular malformation by the International Society for the Study of Vascular Anomalies (ISSVA). Diagnosis is based on the integration of clinical and pathologic findings. In this article, we describe the ultrasound features of 3 VVMs and propose ultrasound as a useful, noninvasive diagnostic and follow-up tool that can also be used to guide and optimize treatment.

We selected 3 patients with histologically confirmed VVM from the database in our department. Their lesions were assessed and compared using 22-MHz cutaneous ultrasound.

Patient 1 was a 10-year-old boy with a VVM on his left ankle that had been present since birth (Fig. 1A). Ultrasound showed a thickened, hyperechoic epidermis with hypoechoic vascular channels in the dermis and subcutaneous tissue and no color Doppler signal (Fig. 2A).

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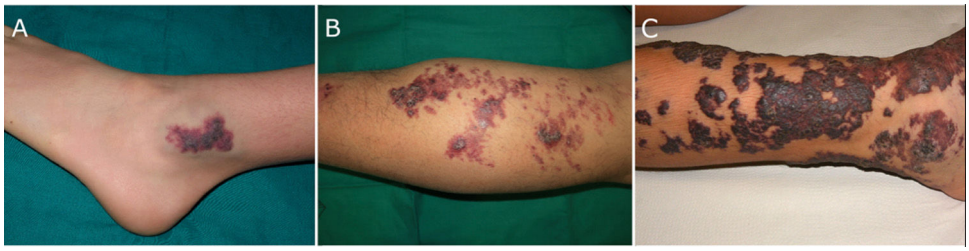

Figure 1 Clinical images of verrucous venous malformations. A, Patient 1. Verrucous erythematous violaceous plaque on the left ankle. B, Patient 2. Erythematous, violaceous, hyperkeratotic plaques with satellite lesions and hemorrhagic superficial vesicles on the back of the right leg. C, Patient 3. Erythematous, violaceous plaques with a verrucous surface comprising hyperkeratotic areas and crusts involving the circumference of the distal region of the left leg.

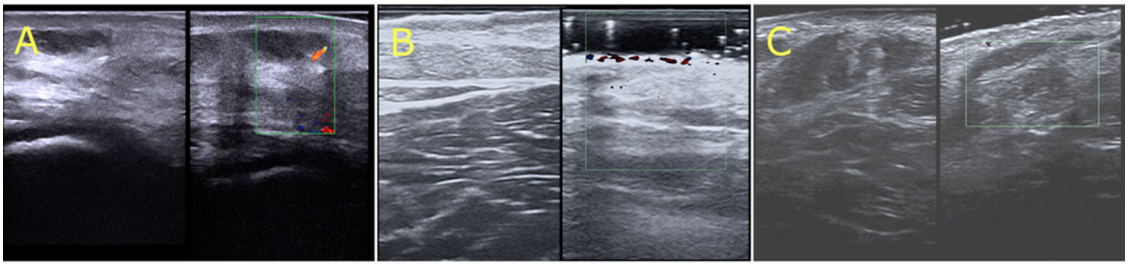

Figure 2 Ultrasound images of verrucous venous malformations (VVMs). A, Cutaneous ultrasound (22 MHz) of the VVM in patient 1. Thickened hyperechoic epidermis, hypoechoic dermis, hypoechoic vascular channels in the dermis and subcutaneous tissue, and absence of color Doppler signal. B, Cutaneous ultrasound (22 MHz) of VVM in patient 2. Thickened, hyperechoic epidermis with vascular channels in the superficial and deep dermis; no color Doppler signal. C, Cutaneous ultrasound (22 MHz) showing hyperechoic epidermis, decreased dermal echogenicity, and a loss of definition between the dermis and hypodermis. Note the thickened, heteroechoic subcutaneous tissue and absence of a color Doppler signal.

Patient 2 was a 25-year-old man with a VMM on the back of his right leg that had been present since birth (Fig. 1B). The patient had experienced recurring episodes of ulceration, pain, and bleeding over the previous 4 years. Cutaneous ultrasound showed a thickened, hyperechoic epidermis with vascular channels in the superficial and deep dermis. Again, there was no color Doppler signal (Fig. 2B).

Patient 3 was a 12-year-old boy with a circumferential VVM in the distal region of his left leg that had been present since birth (Fig. 1C). In recent years, the lesion had gradually gained in volume and become increasingly hyperkeratotic; it had also shown ulceration and bleeding. Cutaneous ultrasound showed a thickened, hyperechoic epidermis, decreased echogenicity in the dermis, a loss of definition between the dermis and hypodermis, hypodermal thickening and heterogeneity, and no color Doppler signal (Fig. 2C).

VVM is uncommon and is included in the group of vascular malformations in the 2018 ISSVA classification.[1] It has been linked to a somatic mutation in the *MAPK3* gene.[2]

VVM consists of a proliferation of venous blood vessels in the dermis and subcutaneous tissue, with sparing of the fascia and muscle tissue. It is congenital or develops in early childhood and mainly affects the lower extremities. Lesions are typically unilateral. Complications include bleeding, pain, and ulceration, especially in larger, more hyperkeratotic lesions. VVM is not associated with tissue hypertrophy or other developmental abnormalities.[3]

Diagnosis is usually established by correlating clinical and pathologic findings, although characteristic clinical findings typically help guide diagnosis. Histology shows epidermal acanthosis and papillomatosis, with compact hyperkeratosis and dilated venous vessels in the papillary and reticular dermis and subcutaneous tissue. Endothelial cells are positive for *GLUT-1* and *WT-1* and negative for D2-40 on immunostaining.[4]

Cutaneous ultrasound is a useful alternative to invasive diagnostic tests in the setting of vascular lesions. The ultrasound characterization of VVM is not well established, but based on our findings, we propose a description that could help diagnose this entity. Ultrasound features of VVM include

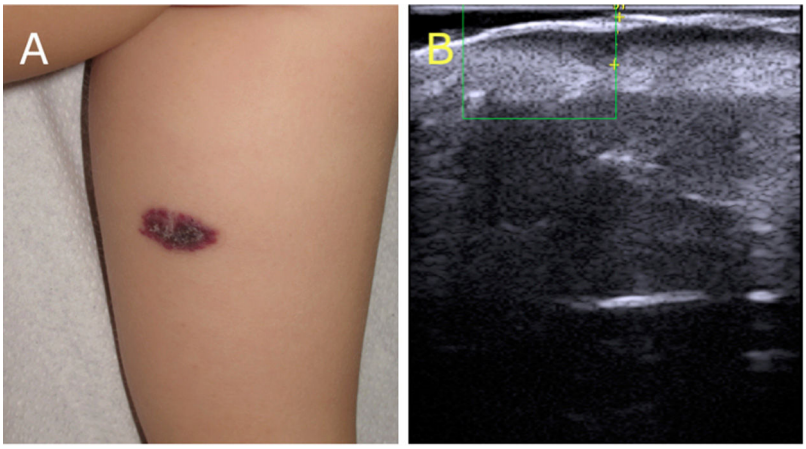

Figure 3 Clinical and ultrasound image of an angiokeratoma. A, Angiokeratoma circumscriptum. Erythematous plaque with a hyperkeratotic surface, clinically indistinguishable from verrucous venous malformation. B, Cutaneous ultrasound (22 MHz). Epidermal hyperechogenicity with vascular channels limited to the papillary dermis; no color Doppler signal.

a thickened, hyperechoic epidermis, a loss of definition between the dermis and epidermis, hyperechoic channels in the dermis and subcutaneous tissue, and absence of a color Doppler signal (Fig. 2).

Angiokeratoma circumscriptum is the main entity to be considered in the differential diagnosis (Fig. 3A)[5]. Its histologic features include hyperkeratotic acanthosis and dilated capillaries in the papillary dermis, with no involvement of the deep dermis or subcutaneous tissue. Contrary to VVM, angiokeratoma circumscriptum shows negative endothelial expression of GLUT-1 and WT-1 and positive expression of D2-40 on immunostaining.[6] Given the differences in the depth of involvement, ultrasound could help distinguish between the 2 entities without the need for invasive tests. Ultrasound features of angiokeratoma include epidermal thickening and hyperechogenicity and a hypoechoic lesion limited to the papillary dermis. As with VVM, there is no Doppler signal (Fig. 3B).[7]

The treatment of VVM is largely surgical, but surgery can sometimes be complex. No treatment is an option in the absence of cosmetic concerns or symptoms.[8] Pulsed dye, multiplex, and carbon dioxide laser therapy have been used to treat complications such as ulceration and bleeding and to achieve better cosmetic outcomes.[9] It should be noted that laser therapy does not resolve deep lesions and is associated with a risk of recurrence. Good results with topical and oral sirolimus have been described.[10]

Ultrasound could also be useful for monitoring the treatment of VVM as it identifies deeper areas of involvement, which can be treated surgically, and more superficial areas, which can be treated with laser therapy or topical sirolimus. A combination of approaches may improve outcomes.

In conclusion, cutaneous ultrasound appears to be a useful tool for diagnosing VVM and differentiating it from angiokeratoma. It may also be of value for planning and monitoring treatment.

Conflicts of interest

The authors declare that they have no conflicts of interest.

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V. Amat-Samaranch*, C. Morales-Múnera, L. Puig, E. Roé

Servicio de Dermatología, Hospital de la Santa Creu i Sant Pau, Barcelona, Spain

* Corresponding author.
E-mail address: vamat@santpau.cat (V. Amat-Samaranch).

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Sclerotherapy With Polidocanol for Digital Myxoid Cysts: A Series of 15 Cases☆

Tratamiento de los quistes mixoides digitales con escleroterapia con polidocanol: serie de 15 casos

To the Editor,

Digital myxoid cysts are the most common benign tumors of the fingers and toes. They present as single, generally translucent, nodules that typically affect the dorsal or lateral aspect of the distal interphalangeal joints or the proximal nail fold.[1] They tend to be asymptomatic, although they can cause pain.[2] The mechanism underlying their development is unclear. In most cases, degenerative changes involving the fibrous capsule of the joint are present. Multiple treatments exist, but none has proven ideal.

Polidocanol solution is a liquid sclerosing detergent that has been used to treat varicose veins, venous malformations, and other vascular anomalies.[1] The aim of this study was to describe our experience with the use of polidocanol sclerotherapy in the treatment of digital myxoid cysts at 2 university hospitals in Spain.

We performed a descriptive study of 15 digital myxoid cysts treated with polidocanol at Hospital Universitario Germans Trias i Pujol and Hospital Universitario Virgen de las Nieves. The procedure consisted of making an incision in the cyst and draining the fluid after application of cryoanesthesia. Polidocanol (Etoxisclerol 20/mL) was injected through the drainage incision using an insulin needle. The area was then covered with a compression bandage for 1 week. The first follow-up visit to assess response to treatment was held after 6 weeks. Patients with persistent cysts were advised to receive a second injection and return in 6 weeks.

Thirteen patients with 15 myxoid cysts were treated with polidocanol sclerotherapy (Table 1). Most of the patients (11/13 [85%]) were women. Their median age was 49.5 years (range, 41-79 years) and the median time since onset was 12 months (range, 4-24 months). Pain (9/13) and nail dystrophy (4/13) were the most common symptoms. One-third of the patients (5/13) had undergone previous treatment with corticosteroid injections and surgery. All 15 myxoid cysts were located on the fingers and the most common location was the distal interphalangeal joint of the third right finger.

At the 6-week follow-up visit, 8 of the 15 cysts had resolved completely, 6 had improved, and 1 persisted. The 6 cysts that showed improvement were treated with a second injection and 5 of them resolved. After 12 weeks thus, 13 of the 15 cysts (86%) had resolved (Fig. 1). Half of the patients (7/13) described pain as the most common adverse effect, but this disappeared in a few days. Just 3 of the 15 myxoid cysts recurred over a follow-up period of 6 months.

Various treatments exist for symptomatic digital myxoid cysts, including drainage, injection therapy with corticosteroids or sclerosing agents, cryotherapy, carbon dioxide or Nd:YAG laser therapy, and surgical excision. No treatment guidelines are available and none of the treatments used to date has proven to be totally effective.[1] Recurrence and adverse effects are common.

Little has been published on the use of polidocanol sclerotherapy in the treatment of digital myxoid cysts. Sclerosing agents include detergents (polidocanol, sodium sulfate, and sodium diatrizoate), chemical agents (iodine, alcohol), and osmotics (salicylates, hypertonic saline). Polidocanol is the most widely used agent for myxoid cysts.[2] Sclerosing agents attack the cell membrane, damaging the endothelial lining and triggering occlusion of the blood vessels that feed the cyst.[2]

The authors of a systematic review proposed sclerotherapy as second-line treatment after surgery, as they found it was the nonsurgical technique with the highest cure rates.[2,4]

The response rate in our series is similar to that described in 2 previous series of 63 and 6 cases, which reported a cure rate of 80% after 1 or 2 sessions.[3]

Surgery has the highest success rate, with resolution in up to 95% of cases and recurrence rates ranging from 2% to 10%, depending on the approach.[2,4] It is, however, an invasive technique that can cause considerable adverse effects.[2] Cryotherapy has a cure rate of 61.1% and a recurrence rate of 10%.[2] Corticosteroid injection therapy combined with drainage and aspiration of cyst contents is associated with a cure rate of 50% to 64% and frequent recurrences.[2,4] Finally, carbon dioxide laser therapy is associated with a 66% cure rate and a 33% relapse rate.[2]

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