

B. García-Bracamonte,* L. Fuertes, R. Llamas,
and F. Vanaclocha

*Servicio de Dermatología, Hospital Universitario 12
de Octubre, Madrid, Spain*

*Corresponding author.

E-mail address: beagarcia50@hotmail.com

(B. García-Bracamonte).

Smooth Muscle Hamartoma Associated with Acquired Blaschkoid Nevus Spilus

Hamartoma del músculo liso asociado a nevo *spilus* blaschkoide adquirido

To the Editor:

Smooth muscle hamartoma (SMH), described by Stokes in 1923, is a proliferation of smooth muscle bundles. It can be single or multiple and may be congenital and acquired.¹ The most frequent presentation is the localized congenital form, which is characterized by a plaque with a variable degree of hyperpigmentation, hypertrichosis, and induration. It is usually less than 10 cm in diameter and appears on the trunk or proximal areas of the limbs. Rubbing the affected area can lead to pseudo-Darier sign, which consists of elevation, induration, or transient piloerection.²⁻⁴ We report the case of a patient who recently attended our hospital with localized SMH within an acquired blaschkoid nevus spilus.

The patient was a 32-year-old man with no past history of interest who was seen for an asymptomatic pigmented lesion that appeared on the right shoulder when he was 6 years old. The patient and his mother described the gradual onset of darker spots within the lesion and, aged 13, the appearance of a hairy and slightly pruritic area.

Physical examination showed a homogenous, light brown, macular lesion with well-circumscribed irregular margins. It

had a unilateral, segmental distribution that followed the lines of Blaschko in a wide band across the upper area of the back and right shoulder and was sharply interrupted at the midline. Multiple blackish or dark brown spots were scattered over the lesion, most of which were elevated and less than 1 cm in diameter (Figure 1A). Near to the right external border there was a light brown plaque, 2 cm in diameter, firmer to the touch, and with long thick dark hair (Figure 1B); rubbing the lesion elicited transient piloerection.

Histopathology of the biopsy taken from the hairy area showed lentiginous melanocytic hyperplasia in the epidermis and disorganized, irregularly shaped smooth muscle fascicles in the dermis that were not associated with the pilosebaceous unit. These fascicles were surrounded by a clear space that separated them from the surrounding collagen (Figure 2). Immunohistochemical staining for muscle-specific actin highlighted the irregular distribution and organization of the smooth muscle fascicles in the dermis (Figure 3). A diagnosis was made of blaschkoid nevus spilus associated with SMH.

Smooth muscle hamartoma is sometimes associated with other skin conditions. Becker nevus is an abnormality that usually appears at the onset of puberty, presenting as a hyperpigmented area that can develop hypertrichosis. Histopathology shows a degree of acanthosis, elongated rete ridges, and hyperpigmentation of the basal layer. It is not unusual to observe smooth muscle hyperplasia in the dermis. For this reason, some authors suggest that SMH and Becker nevus represent opposite ends of a spectrum

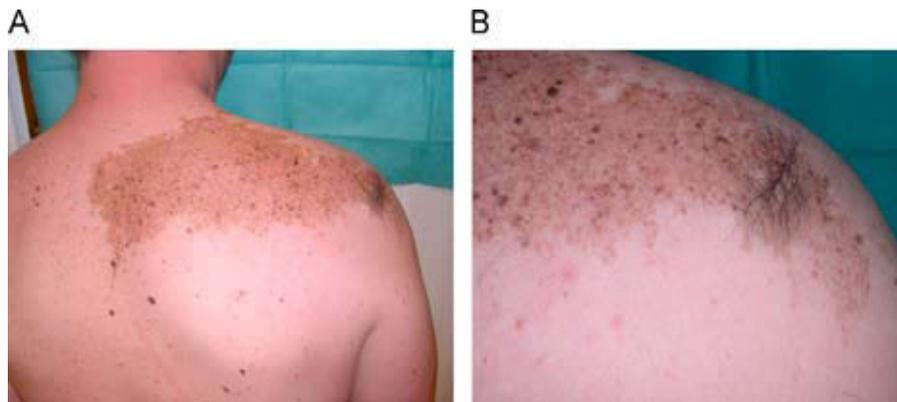


Figure 1 A, Upper back and right shoulder with a homogeneous clear brown macular lesion containing multiple blackish lesions and a hypertrichotic area. B, The area near the right external edge of the nevus spilus shown in greater detail. Light brown plaque, 2×2 cm diameter, containing thick, long, dark hair.

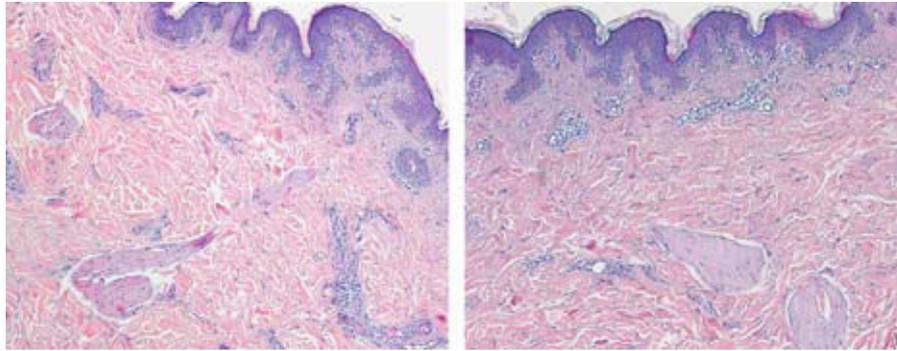


Figure 2 Lentiginous melanocytic hyperplasia in the epidermis and disorganized smooth muscle fascicles surrounded by a collagen retraction artifact in the dermis (hematoxylin-eosin, $\times 40$).

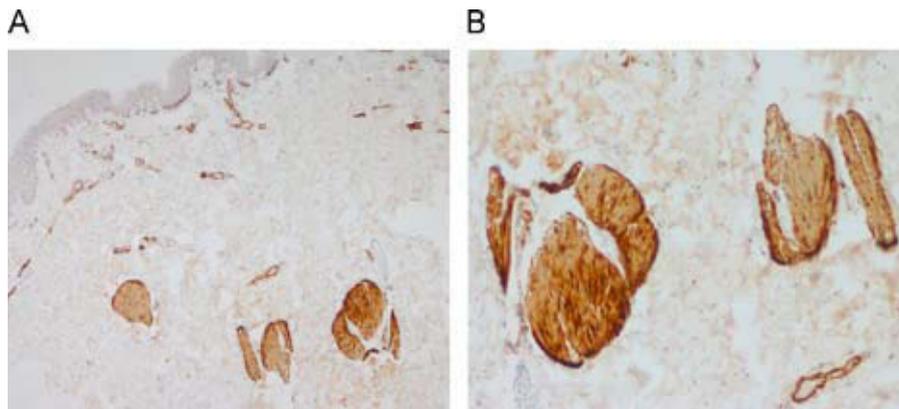


Figure 3 Immunohistochemistry. A, Irregular distribution and organization of smooth muscle fascicles in the dermis (muscle-specific actin, original magnification $\times 40$). B, The same area in greater detail (muscle-specific actin, original magnification $\times 100$).

of hamartomatous lesions that combine smooth muscle hyperplasia with hyperpigmentation and hypertrichosis.^{1,2} In 2007, Patrizi et al⁵ reported a case of Becker nevus associated with acquired melanocytic nevus and SMH on which basal cell carcinoma developed. Recently, Zarineh et al⁶ described a lesion with combined features of melanocytic nevus and SMH.

Nevus spilus, also known as speckled lentiginous nevus, is a speckled hyperpigmented lesion that appears in childhood and follows the lines of Blaschko in more than 50% of cases. Histology of the light-brown area shows lentiginous features and the darker foci are junctional or compound nevi.^{7,8} A literature search detected no similar cases of SMH associated with nevus spilus, except for 1 case with a plaque-type blue nevus associated with nevus spilus and smooth muscle hyperplasia, described by Park et al in 1999.⁹

In such cases, the differential diagnosis should include conditions that present: hyperpigmented lesions that may follow the lines of Blaschko, such as agminated melanocytic nevi and partial unilateral lentiginosis,⁷⁻¹¹ and hyperpigmented lesions that can be associated with hypertrichosis, such as congenital melanocytic nevi and Becker nevus.^{1,2}

References

- Holst VA, Junkins-Hopkins JM, Elenitsas R. Cutaneous smooth muscle neoplasms: clinical features, histologic findings, and treatment options. *J Am Acad Dermatol.* 2002;46:477-90.
- Requena L, Sanguenza OP, Requena C, Torrelo A. Hamartoma de músculo liso. *Monogr Dermatol.* 2002;15:33-7.
- Clemente-Ruiz de Almirón A, Corbalán-Vélez R, Martínez-Barba E, Frias-Iniesta JF. Hamartoma de músculo liso simulando hidrocistomas ecinos múltiples. *Actas Dermosifiliogr.* 2008;99:493-4.
- González-Beato Merino MJ, Zazo Hernández V, Hernández Hermosa JM, Lecona Echevarría M, Lázaro Ochaíta P. Hamartoma congénito de músculo liso: a propósito de un caso. *Actas Dermosifiliogr.* 1999;90:258-60.
- Patrizi A, Medri M, Neri I, Fanti PA. Becker naevus associated with basal cell carcinoma, melanocytic naevus and smooth-muscle hamartoma. *J Eur Acad Dermatol Venereol.* 2007;21:130-2.
- Zarineh A, Kozovska ME, Brown WG, Elder DE, Rabkin MS. Smooth muscle hamartoma associated with a congenital pattern melanocytic nevus, a case report and review of the literature. *J Cutan Pathol.* 2008;35(Suppl. 1):83-6.
- Hanayama H, Terashi H, Kashikawa K, Tahara S. Congenital melanocytic nevi and nevus spilus have a tendency to follow the lines of Blaschko: an examination of 200 cases. *J Dermatol.* 2007;34:159-63.

8. Torrelo A, Baselga E, Nagore E, Zambrano A, Happle R. Delineation of the various shapes and patterns of nevi. *Eur J Dermatol.* 2005;15:439-50.
9. Park YM, Kang H, Cho BK. Plaque-type blue nevus combined with nevus spilus and smooth muscle hyperplasia. *Int J Dermatol.* 1999;38:775-7.
10. Cabanillas González M, Monteagudo B, de las Heras C, Cacharrón JM. Unilateral agminated melanocytic nevi. *Dermatology.* 2009;218:387-8.
11. Monteagudo B, Cabanillas M, Suárez-Amor O, Pérez-Valcárcel J, de la Cruz-Mera A. Lentiginosis unilateral parcial siguiendo las líneas de Blaschko. *Piel.* 2009;24:459-60.

B. Monteagudo,^{a,*} A. Ramírez-Santos,^a M. Cabanillas,^a O. Suárez-Amor,^a and J. Pérez-Valcárcel^b

^a*Servicio de Dermatología, Complejo Hospitalario Arquitecto Marcide-Novoa Santos, Ferrol, A Coruña, Spain*

^b*Servicio de Anatomía Patológica, Complejo Hospitalario Arquitecto Marcide-Novoa Santos, Ferrol, A Coruña, Spain*

*Corresponding author.

E-mail address: benims@hotmail.com (B. Monteagudo).

Oral Venous Malformation Treated with Pulsed-Dye and Neodymium:Yttrium-Aluminum-Garnet Sequential Laser

Malformación venosa oral tratada con aplicación secuencial de láser de colorante pulsado y Nd:YAG

To the Editor:

Venous malformations are slow-flow vascular malformations present at birth, although they occasionally do not become clinically evident until several years later. They manifest clinically as soft, nonpulsatile masses of blue or violaceous color that compress easily on palpation. Their presence within the mouth is not uncommon and can be associated with bleeding, ulceration, pain, difficulty swallowing, airway obstruction, and facial deformity.^{1,2}

The classic approaches to these lesions include sclerotherapy and surgery although this can occasionally result in significant deformity, prolonged pain, skin necrosis, nerve damage, or systemic toxicity. This has led to the use of various lasers—carbon dioxide, argon or diode—sometimes in combination with radiofrequency current³ and, more recently, alexandrite² or long-pulsed Neodymium:Yttrium-Aluminum-Garnet (Nd:YAG) lasers.^{4,5} The Nd:YAG laser is considered the treatment of choice for this condition.

We present the case of a 16-year-old man with a soft, compressible bluish tumor on the mucosa of the cheek, lower lip, and half of the tongue on the right side (Figure 1). The patient reported the lesion had been present since birth and had increased progressively in size over the years. Doppler ultrasound confirmed the tumor to be a slow-flow vascular malformation. Sequential treatment with pulsed dye laser (PDL) and Nd:YAG laser (Cynergy with Multiplex®, Cynosure, Westford, MA, USA) was recommended in view of the diagnosis of venous malformation, the patient's difficulty swallowing, and the marked deformity.

The patient received 7 sessions of treatment over a 10-month period, using PDL (595 nm wavelength) followed

by Nd:YAG (1064 nm wavelength) with a 1-second delay. The following parameters were employed: 7 mm spot size; 10 ms pulses with a fluence of 10 J/cm² with PDL, and 15 ms pulses with a fluence of 70 J/cm² with the Nd:YAG laser. Throughout the treatment a pre-cooled airflow skin cooling system (Cryo5®, Zimmer Medizinsysteme GmbH, Neu-Ulm, Germany) was used at its highest setting.

Treatment was applied in a slow and progressive manner in order to avoid causing lingual edema that could compromise the airway, and 20 mg to 40 mg of intramuscular methylprednisolone was administered in the initial sessions. Anesthetic infiltration of the lesion with mepivacaine 2% was required prior to laser treatment but in the 2 last sessions no anesthesia was needed. Moderate edema was observed in the treated areas but there was no respiratory compromise and no other significant adverse reactions were seen. Follow-up 8 months after treatment showed a marked reduction in the size of the venous malformation, with complete resolution in some areas (Figures 2 and 3).



Figure 1 Soft bluish tumor on the mucosa of the right cheek and tongue.