

# Acral Syringomas Presenting as a Photosensitive Papular Eruption

Rafael F. Martín-García, MD; Christine M. Muñoz, MD

*A 43-year-old woman presented with a recurrent and remitting eruption of reddish-brown papules in the sun-exposed areas of the arms and forearms that had been present for 2 years. Results of a histopathologic examination revealed syringoma. To the best of our knowledge, this is the first case report of syringomas confined to sun-exposed areas with a recurrent and remitting course over time. We also review the literature on acral syringomas.*

*Cutis. 2006;77:33-36.*

Syringomas are benign skin tumors of adnexal origin. Clinically, they usually present as single or grouped, soft, dome-shaped or flat-topped flesh-colored papules. Syringomas appear predominantly on the eyelids of pubertal or middle-aged women but also can present localized to the vulva, penis, buttocks, scalp, upper extremities, or trunk.

We present the case of a 43-year-old woman with a recurrent and remitting eruption of acral syringomas presenting clinically as a photosensitive papular dermatosis localized to the sun-exposed areas of the upper extremities.

## Case Report

A 43-year-old woman in otherwise good health presented to our clinic with a 2-year history of a skin eruption localized to her distal arms and forearms. According to the patient, sun exposure, heat, hot baths, and stress exacerbated the lesions. She also reported that the condition was characterized by periods of exacerbations and remissions, with complete disappearance of the lesions between episodes. Results of a physical examination

revealed evanescent reddish-brown papules on the sun-exposed areas of the distal arms and forearms. Our differential diagnosis included photodermatitis, polymorphous light eruption, and cutaneous lupus erythematosus. We recommended that the patient protect her skin from the sun and use sunscreen and desonide lotion. Laboratory tests were ordered. The patient was lost to follow-up for 5 months, after which she presented to our clinic without skin lesions. Results of laboratory tests for a hemogram, urinalysis, serum chemistries, and antinuclear antibodies were within reference range. It was recommended she continue with sun protection and return to the clinic as soon as new lesions appeared.

Three years later, the patient returned to our clinic with a similar skin eruption. Results of an examination again revealed numerous reddish-brown, confluent papules localized to the sun-exposed areas of the distal arms and forearms (Figures 1 and 2), with relative sparing of the sun-protected wristwatch area.

The results of a histopathologic examination of 2 different lesions revealed the presence of small aggregates of epithelial cells forming ductal structures in the dermis. Tadpolelike epithelial cords extended from some of the ductal structures into the surrounding stroma. The findings were consistent with a diagnosis of syringoma (Figures 3 and 4).

## Comment

Syringomas, initially described by Kaposi<sup>1</sup> in 1872 as *lymphangioma tuberosum multiplex*, are benign skin tumors of adnexal origin. They usually present as single or grouped, soft, dome-shaped or flat-topped papules ranging in size from one to several millimeters. Individual lesions may be flesh-colored or have varying shades of yellow, red, or brown. Lesions appear predominantly on the eyelids of pubertal or middle-aged women. Most of the cases are sporadic, but familial cases have been reported.<sup>2</sup> An increased incidence of syringomas has been reported in patients with Down syndrome.<sup>3</sup>

Accepted for publication August 13, 2004.

From the Department of Dermatology, University of Puerto Rico School of Medicine, San Juan.

The authors report no conflict of interest.

Reprints: Rafael F. Martín-García, MD, Department of Dermatology, University of Puerto Rico School of Medicine, PO Box 365067, San Juan, Puerto Rico 00936-5067 (e-mail: martinmubarak@aol.com).



**Figure 1.** Papular eruption in the sun-exposed areas of the left forearm.

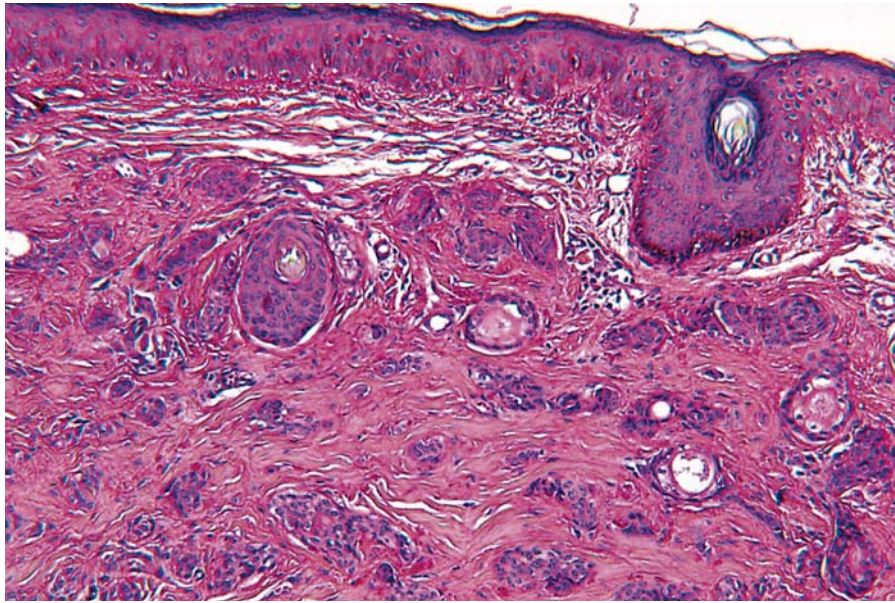


**Figure 2.** Reddish-brown papules localized to the dorsal aspect of the left forearm.

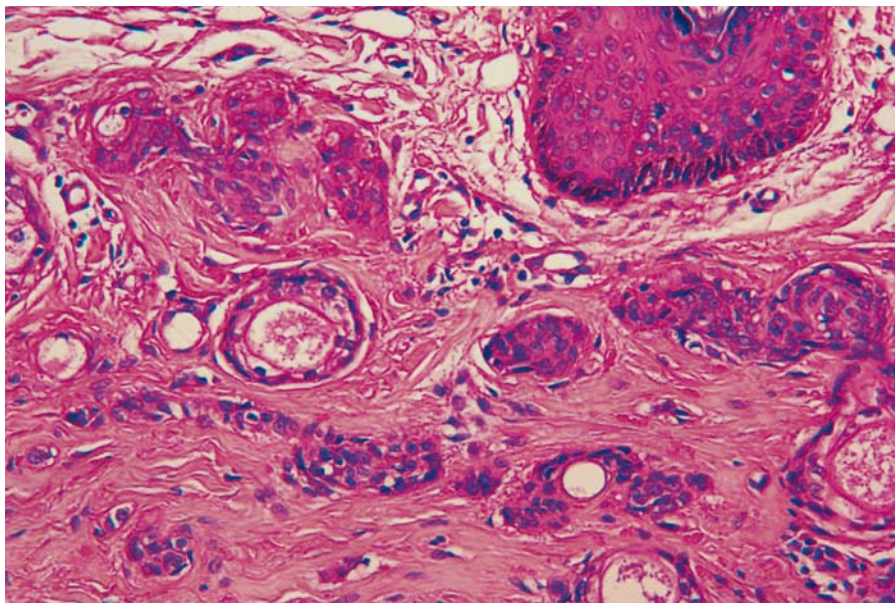
Histopathologically, syringomas are characterized by the presence of numerous small ductal structures and cords of cells surrounded by a fibrous stroma. The ductal structures usually have a double layer of epithelial cells and may contain an amorphous substance or keratin. Commalike or tadpole-like tails of epithelial cells extend from some of the ductal structures. A clear cell variant of syringoma with epithelial cells containing abundant glycogen has been described.<sup>4</sup> Although some controversy still exists, immunohistochemical and ultrastructural studies have confirmed that these lesions originate from the acrosyringal portion of the eccrine sweat gland.<sup>2</sup>

Friedman and Butler<sup>5</sup> classified syringomas into 4 clinical variants: a localized form, a familial form, a Down syndrome-associated form, and a generalized form that includes multiple and eruptive syringomas described by Jacquet and Darier<sup>6</sup> as *eruptive hidradenoma*. Unusual reported presentations of localized syringoma include syringomas with a unilateral nevoid pattern,<sup>7</sup> localized vulvar and penile syringomas,<sup>8,9</sup> syringoma of the scalp associated with nonscarring alopecia,<sup>10</sup> and acral syringoma.<sup>11</sup>

There are 4 reports of syringomas with predominant localization to the upper extremities. Hughes and Apisarnthanarax<sup>11</sup> described a 31-year-old man with multiple grouped and confluent syringomas



**Figure 3.** Ductal structures and epithelial strands scattered within a fibrous stroma (H&E, original magnification  $\times 10$ ).



**Figure 4.** Ductal lumina lined by 2 rows of epithelial cells with central amorphous eosinophilic debris. Note the commalike tails of epithelial cells extruding from ductal structures (H&E, original magnification  $\times 20$ ).

localized to the dorsal aspect of the proximal and middle phalanges of both hands. They coined the term *acral syringomas* for this presentation. Asai et al<sup>12</sup> presented a 35-year-old woman with multiple grouped syringomas localized to the dorsum of the proximal phalanges, some with annular configuration. van den Broek and Lundquist<sup>13</sup> reported a 52-year-old man who presented with multiple syringomas on the dorsal aspect of the wrists and distal forearms. A year later, new lesions had developed distally on the hands, proximally on the forearms, and on the lower eyelids. In a 1998 published review of 29 cases of syringoma, Patrizi et al<sup>14</sup> included 2 patients with lesions localized to the

forearms and wrists without further describing the exact distribution of the lesions within the affected areas. They suggested classifying cases of syringoma with exclusive localization to the wrists and forearms as acral syringoma.<sup>14</sup>

Our case is unique for several reasons. Even though syringomas with increased pruritus during hot weather<sup>15</sup> and increased erythema during the summer<sup>16</sup> have been reported, to the best of our knowledge, this is the first case report of syringomas localized to sun-exposed areas. This presentation mimics a photosensitive papular eruption such as photodermatitis, polymorphous light eruption, or cutaneous lupus erythematosus. The possibility

of a misdiagnosis due to inadequate tissue sampling was ruled out when the biopsy result of a similar lesion revealed identical histopathologic findings. Other histopathologic changes, such as inflammation, were lacking. It may be possible that precipitating factors such as sun exposure, heat, and stress contributed to the exacerbation of the lesions (previously clinically imperceptible) through stimulation of the sweat response. In addition, although one report comments on the rare possibility of the spontaneous disappearance of syringomas,<sup>17</sup> the exacerbating and remitting course of the condition in our patient has never been previously reported.

Acral syringoma should be included in the differential diagnosis of papular photodistributed eruptions of the upper extremities. We agree with Patrizi et al<sup>14</sup> in that syringomas with exclusive localization to the distal upper extremities should be classified as acral syringoma.

## REFERENCES

1. Kaposi M. Lymphangioma tuberosum multiplex. In: Fagge CH, trans-ed. *Hebra on Diseases of the Skin*. Vol. 3. London, England: New Sydenham Society; 1872:386.
2. Hashimoto K, Blum D, Fukaya T, et al. Familial syringoma: case history and application of anti-ecrine gland antibodies. *Arch Dermatol*. 1985;121:756-760.
3. Butterworth T, Streaun L, Beerman H, et al. Syringoma and mongolism. *Arch Dermatol*. 1964;90:483-487.
4. Headington JD, Koski J, Murphy PJ. Clear cell glycogenosis in multiple syringomas. description and enzyme histochemistry. *Arch Dermatol*. 1972;106:353-356.
5. Friedman SJ, Butler DF. Syringoma presenting as milia. *J Am Acad Dermatol*. 1987;16:310-314.
6. Jacquet L, Darier J. Hidradénomes éruptifs, I: épithéliomes adénoides des glandes sudoripares ou adénomes sudoripares. *Ann Dermatol Venereol*. 1887;8:317-323.
7. Yung CW, Soltani K, Bernstein JE, et al. Unilateral linear nevoidal syringoma. *J Am Acad Dermatol*. 1981;4:412-416.
8. Carneiro S, Gardner H, Knox J. Syringoma of the vulva. *Arch Dermatol*. 1971;103:494-496.
9. Zalla JA, Perry HO. An unusual case of syringoma. *Arch Dermatol*. 1971;103:215-217.
10. Neuman KM, Burnett JW. Alopecia associated with syringomas. *J Am Acad Dermatol*. 1985;13:528-529.
11. Hughes PS, Apisarnthanarax P. Acral syringoma. *Arch Dermatol*. 1977;113:1435-1436.
12. Asai Y, Ishii M, Hamada T. Acral syringoma: electron microscopic studies on its origin. *Acta Derm Venereol*. 1981;62:64-68.
13. van den Broek H, Lundquist CD. Syringomas of the upper extremities with onset in the sixth decade. *J Am Acad Dermatol*. 1982;6:534-536.
14. Patrizi A, Neri I, Marzaduri S, et al. Syringoma: a review of twenty-nine cases. *Acta Derm Venereol*. 1998;78:460-462.
15. Dyll-Smith DJ, Connors TJ, Scurry J. Generalised eruptive syringoma—a papular dermatosis. *Australas J Dermatol*. 1990;31:95-98.
16. Lee AY, Kawashima M, Nakagawa H, et al. Generalized eruptive syringoma. *J Am Acad Dermatol*. 1991;25:570-571.
17. Beers BB. An eruption of asymptomatic brown papules. eruptive disseminated syringomas. *Arch Dermatol*. 1990;126:954-955, 957-958.