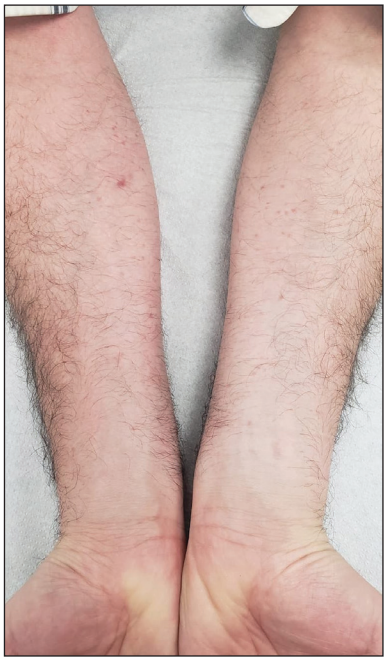
Eruptive Erythematous Papules on the Forearms

Diego Aragón-Caqueo, MD; Mark Gregory, MS; José Luis Gatica, MD; Ivo Sazunic, MD; Rodrigo Loubies, MD



A 44-year-old man presented to the dermatology department with multiple eruptive, nonconfluent, erythematous papules on the anterior forearms of 2 years' duration. The patient's medical history was notable for right-sided testicular cancer diagnosed in childhood and 3 excised basal cell carcinomas, the most recent of which was concurrent with the present case. The patient denied any recent pruritus, exposure to irritants, or use of over-the-counter medications. Physical examination was remarkable for numerous monomorphic, symmetric, nonconfluent, flesh-colored to slightly pigmented papules on the dorsal aspect of the forearms. No involvement of the fingers or lower extremities was observed. Two punch biopsies of representative lesions on the right and left forearms were taken. Histopathologic examination revealed eccrine ductal proliferations lined by cuboidal cells embedded within bundles of sclerotic collagen.

WHAT'S YOUR **DIAGNOSIS?**

a. acral eruptive syringoma
b. Darier disease
c. flat warts
d. lichen planus
e. urticaria pigmentosa

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Dr. Aragón-Caqueo (ORCID: 0000-0001-7233-960X) is from Escuela de Medicina, Universidad de Tarapacá, Arica, Chile. Mark Gregory is from the School of Medicine, Wayne State University, Detroit, Michigan. Drs. Gatica and Loubies are from Clínica Orlandi, and Facultad de Ciencias Médicas, Universidad de Santiago, Santiago, Chile. Dr. Sazunic is from Laboratorio Histodiagnóstico Málaga, Santiago.
The authors have no relevant financial disclosures to report.
Correspondence: Diego Aragón-Caqueo, MD, Universidad de Tarapacá, Avenida 18 de Septiembre, 2222, Arica, Chile (diegomarceloaragon@gmail.com).
Cutis. 2025 May;115(5):E15-E17. doi:10.12788/cutis.1231

THE **DIAGNOSIS:** Acral Eruptive Syringoma

Syringomas are small, benign, often asymptomatic eccrine tumors that originate in the intraepidermal portion of eccrine sweat ducts.[1] Clinically, they present as multiple symmetric white-to-yellow or discrete flesh-colored papules measuring 1 to 3 mm in diameter, often located on the face (most commonly on the eyelids), with a greater prevalence in middle-aged women. Occasionally, they manifest in other locations such as the cheeks, chest, axillae, abdomen, and groin.[2]

In 1987, Friedman and Butler[3] developed a classification system categorizing syringomas into 4 clinical subtypes: familial syringoma, localized syringoma, Down syndrome–related syringoma, and generalized syringoma. The fourth subtype includes the variant of eruptive syringoma,[3] a rare clinical manifestation that often develops before or during puberty with several flesh-colored or lightly pigmented papules on the neck, anterior chest, upper abdomen, axillae, periumbilical region, and/or genital region.[1,4,5] The etiology of eruptive syringomas is unclear, although it has been linked to abnormal proliferation of sweat glands due to an underlying local inflammatory process.[6]

Acral distribution of syringomas is a rare variant that can manifest as part of generalized eruptive syringoma with consequent involvement of the arms and other areas.[5,7] There are limited case reports on eruptive syringomas with predominant acral distribution.[8] Compared to classic syringomas, the acral variant is associated with an older age of onset as well as a similar prevalence between men and women.[9] Acral eruptive syringoma (AES) usually is isolated to the distal arms and legs. The most commonly affected region is the anterior surface of the forearms, although involvement of the dorsal hands, wrists, and feet also has been reported.[10-16]

The first known case of AES, which was reported in 1977, described eruptive syringomas on the dorsal hands of a healthy 31-year-old man.[17] Several cases have been reported since then, mostly in patients aged 30 to 60 years, with predominant involvement of the dorsal hands and forearms.[18-24] A review of Embase as well as PubMed articles indexed for MEDLINE using the search terms *syringoma OR eccrine ductal tumor* and *eruptive OR acral OR arms OR forearms OR extremities* identified 19 reported cases of AES between 1977 and 2023. For the reported AES cases, the mean (SD) age at diagnosis was 45.1 years (15.96 years), with patient ages ranging from 19 to 76 years. Notably, most cases occurred in individuals aged between 30 and 60 years, which deviates from the typical age of onset of localized syringomas, commonly seen during puberty or early adulthood.

Currently, AES is categorized within the clinical presentation of eruptive syringoma. Nevertheless, some authors have proposed classifying it as a distinct fifth clinical group due to specific features that distinguish it from generalized eruptive syringoma.[9] This reclassification has considerable implications for the differential diagnosis, particularly because exclusive acral involvement poses a substantial diagnostic challenge and often requires histologic confirmation.

As shown in the Figure, histopathologic examination revealed tubular structures in the upper dermis with characteristic comma-shaped extensions. Some of these structures were lined with cuboidal cells and contained eosinophilic material within the lumen. There was no

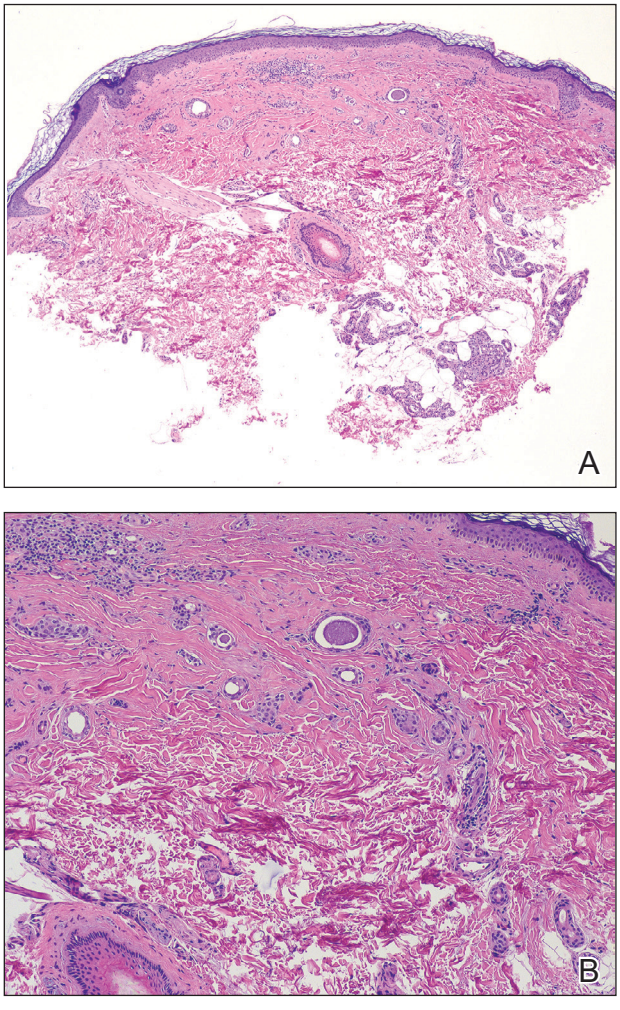

FIGURE. A, Histopathology revealed tubular structures within the upper dermis with no involvement of the epidermis or deeper dermis (H&E, original magnification ×4). B, Higher magnification revealed thick bundles of sclerotic collagen at the upper dermal level, comma-shaped prolongations, and an eosinophilic cuticle occupying the lumen of some of the tubular structures (H&E, original magnification ×10).

involvement of the epidermis or deeper dermis. The histologic features were consistent with syringoma, which is distinguished by its predominant involvement of the upper dermis and the presence of enlarged, dilated eccrine ducts, as observed in our case.

Treatment of syringomas often is challenging due to the high rate of recurrence and the risk for postinflammatory hyperpigmentation. Since the condition is benign, treatment typically is pursued for aesthetic reasons. Various therapeutic approaches have been reported, each with diverse response rates. The most common method involves surgical intervention, either with electrodesiccation or CO_2 laser—both of which have shown satisfactory resolution of lesions without recurrence at 1-year follow-up, with no major scarring reported.[25,26] Alternatively, topical management with retinoids daily over a 4-month period leads to flattening of the tumors with no further appearance of new lesions.[27] Despite the availability of numerous management options, establishing a first-line treatment remains controversial due to the high risk for recurrence and the variability in the number and location of lesions among individual patients. In our case, given the benign nature of syringomas, the asymptomatic nature of the lesions, the involvement of noncritical aesthetic areas, and the limited response to noninvasive therapeutic options, the patient was informed of the diagnosis, and no further pharmacologic or surgical intervention was pursued.

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