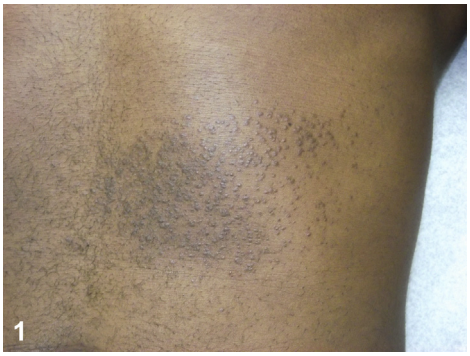
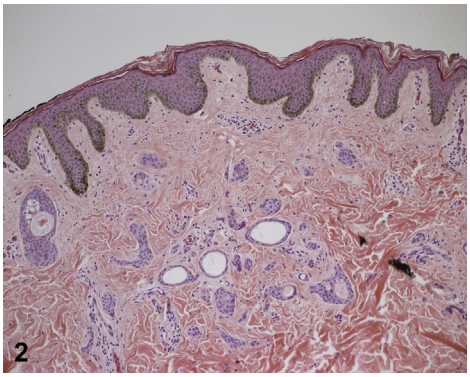
Papules in a segmental unilateral distribution.

Kevin P. Lee, BS,[a] David M. Perry, MD, PhD,[b,c] and Dirk M. Elston, MD[d]
Houston, Texas and Charleston, South Carolina

Key words: eccrine glands; eruptive syringomas; syringomas.





A 23-year-old black man presented with a two-year history of multiple asymptomatic small bumps on the left abdomen. The lesions appeared abruptly and had been stable in size during the past 2 years. The patient denied any prior procedure, itching, or other symptoms before the onset of lesions, and is otherwise healthy. His family history is negative for similar lesions. On examination, the left abdomen showed numerous monomorphic, small, 1-2-mm dome-shaped folliculocentric papules in a sharply demarcated area, which did not extend beyond the midline (Fig 1). A 3-mm punch biopsy was obtained (Fig 2).

Question 1: What is the most likely diagnosis?

A. Segmental angiofibromas

B. Segmental eruptive syringomas

C. Segmental lichen planus

D. Segmental folliculotropic mycosis fungoides

E. Segmental leiomyoma of Reed Syndrome

Answers:

A. Segmental angiofibromas — Incorrect. Angiofibromas are dome-shaped, skin-colored-to-reddish papules that may be a solitary lesion on the nose or may occur in a segmental pattern. On histopathology, fibroplasia and varying degrees of vascular proliferation are seen.[1] Segmental lesions commonly represent loss of heterozygosity (LOH) for tuberous sclerosis (TS) mutations. All patients with TS are born lacking one allele. LOH occurs when the remaining allele is lost during fetal development. As LOH leaves no remaining tumor suppressor gene, these lesions typically present as plaques, which expands the blaschkoid segment, such as the forehead plaque of TS.

B. Segmental eruptive syringomas — Correct. Syringomas are benign adnexal tumors. Segmental or unilateral eruptive syringomas are rare but have been reported. Histopathology shows a dense fibrous stroma with multiple dilated, cystic spaces lined by 2 layers of cuboidal cells. Some cysts have small comma-like tails.[1] Segmental lesions commonly follow the LOH seen in type 2 mosaicism described by Happle. However, segmental forms have also been postulated to represent type 1 mosaicism,

From the McGovern Medical School[a]; Perry Dermatology[b]; Ralph H. Johnson VA Medical Center[c]; Department of Dermatology and Dermatologic Surgery, Medical University of South Carolina.[d]

Funding sources: None.

Correspondence to: Kevin P. Lee, BS, McGovern Medical School, 6431 Fannin St, Houston, TX 77030. E-mail: kevin.p.lee@uth.tmc.edu.

JAAD Case Reports 2021;9:102-4.
2352-5126

https://doi.org/10.1016/j.jdcr.2021.01.027

reflecting heterozygosity for a postzygotic mutation usually seen in non-segmental forms.[2,3]

C. Segmental lichen planus – Incorrect. Segmental lichen planus is a variant of lichen planus also known as 'zosteriform'. The papules usually exibit 'the 5 Ps': planar, polygonal, pruritic, purple, and papular. Typical histopathology shows a band-like interface dermatitis with epidermal hyperplasia, orthokeratosis, and beaded hypergranulosis.[1]

D. Segmental folliculotropic mycosis fungoides – Incorrect. Folliculotropic mycosis fungoides can show papular lesions around hair follicles, sometimes in a segmental distribution. Histopathology reveals epidermotropism and a dense lymphocytic infiltrate around the perimeter of hair follicles. The infiltrate can also penetrate the hair follicle and follicular mucinosis can be seen.[4]

E. Segmental leiomyoma of Reed Syndrome – Incorrect. Reed syndrome is a hereditary genodermatosis consisting of cutaneous, painful leiomyomas, which are frequently distributed in a segmental or dermatomal pattern and often associated with uterine leiomyomas in women. There is a predisposition for renal cell carcinoma. Histopathology shows bundles of smooth muscle fibers.[1]

Question 2: All are clinical presentation types of the above patient's diagnosis, except:

A. Localized

B. Eruptive

C. Familial

D. Trisomy 21-associated

E. Oral

Answers:

A. Localized – Incorrect. Syringomas are associated with a localized presentation. The localized variant is the most common form of syringomas. They are usually seen in the periorbital region. Women are more affected than men.[5]

B. Eruptive – Incorrect. Syringomas are associated with an eruptive presentation. The eruptive presentation is a very rare variant usually seen in young patients. A sudden increase in the number of lesions on the neck, face, axillae, upper arms, and periumbilical areas would be seen.[5] It is unknown whether the lesions are truly neoplastic or are reactive in response to a prior inflammatory process, as they have been reported to occur after trauma (*i.e.* waxing of the suprapubic area).

C. Familial – Incorrect. Syringomas are associated with a familial distribution. Rarely, familial cases have been reported. Familial eruptive variants have also been reported.[6]

D. Trisomy 21-associated – Incorrect. Syringomas are associated with Trisomy 21. A total of 18% of women with Trisomy 21 have syringomas. Patients with Down syndrome are about 30 times more likely than patients with other syndromes to develop syringomas.[1]

E. Oral – Correct. Syringomas are not associated with an oral presentation.

Question 3: All are treatment options of the above patient's diagnosis, except:

A. CO_2 laser

B. Chemical peels

C. Acyclovir

D. Cryotherapy

E. Electrodessication & curettage

Answers:

A. CO_2 laser – Incorrect. Therapeutic intervention is based on patient cosmetic concern. CO_2 laser is a first-line therapy in the treatment of syringomas. It may be used solely or in conjunction with other treatments.[7]

B. Chemical peels – Incorrect. Trichloroacetic acid has been reported to be effective for some patients with syringomas.[7]

C. Acyclovir – Correct. Acyclovir is not indicated as a treatment for syringomas.[7]

D. Cryotherapy – Incorrect. Cryotherapy may also be beneficial in the treatment of syringomas.[7]

E. Electrodessication & curettage – Incorrect. Electrodessication & curettage has been reported as a technique used in the treatment of syringomas.[7]

Abbreviations used:

LOH: loss of heterozygosity
TS: tuberous sclerosis

Conflicts of interest

None disclosed.

REFERENCES

1. James WD, Berger TG, Elston DM, Neuaus IM, Andrews GC. *Andrews' Diseases of the Skin: Clinical Dermatology*. 12th ed. Philadelphia: Elsevier; 2016

2. Happle R. A rule concerning the segmental manifestation of autosomal dominant skin disorders. *Arch Dermatol*. 1997;133:1505-1509.
3. Ceulen RPM, Van Marion AMW, Steijlen PM, Frank J, Poblete-Gutiérrez P. Multiple unilateral skin tumors suggest type 1 segmental manifestation of familial syringoma. *Eur J Dermatol*. 2008;18:285-288.
4. Malveira MIB, Pascoal G, Gamonal SBL, Castañon MCMN. Folliculotropic mycosis fungoides: challenging clinical, histopathological and immunohistochemical diagnosis. *An Bras Dermatol*. 2017;92:73-75.
5. Fessa CK, Sharma R, Fernández-Peñas P. What is your diagnosis? Eruptive syringoma. *Cutis*. 2014;93:31-32.
6. Lau J, Haber RM. Familial eruptive syringomas: case report and review of the literature. *J Cutan Med Surg*. 2013;17:84-88.
7. Lebwohl MG, Heymann WR, Berth-Jones J, Coulson I. *Treatment of Skin Disease: Comprehensive Therapeutic Strategies*. 5th ed. Philadelphia: Elsevier; 2018