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LETTERS TO THE EDITOR

Generalized Syringoma: A Case Study

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To the Editor:

Syringomas are benign adnexal tumors of eccrine ductal origin, characterized by small, firm, rounded, flesh-colored or yellowish papules located on the eyelids, anterior region of the neck, trunk, or abdomen. The highly characteristic histology consists of an epithelial growth in the form of tubules or nests made up of 2 flat cell layers; these structures develop a "tadpole-shaped" tail. The most widely used treatment is ultrapulsed CO_2 laser, although no studies have confirmed its efficacy. These lesions are rarely self-limiting, and recurrences are common.

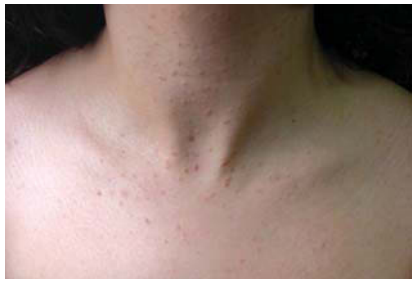

Figure 1. Multiple asymptomatic pink papules at the neckline and anterior region of the neck.

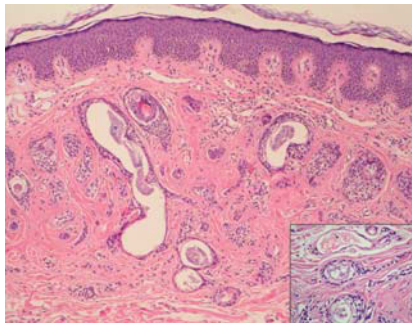

Figure 2. Diffuse dermal neoplasm with cystic structures and epithelial nests in slightly sclerotic stroma (hematoxylin-eosin, ×10). Insert: detail of a cystic ductal structure lined with an eosinophilic cuticle and cells containing a pale cytoplasm (hematoxylin-eosin, ×40).

We describe a 25-year-old woman with no relevant history who consulted for asymptomatic, flesh-colored papules of 1-2 mm, distributed on the anterior area of the neck and neckline (Figure 1). The symptoms started 10 years earlier on the lower eyelids, gradually spreading to the abdomen, neck, and neckline, with outbreaks after periods of sun exposure.

Histological study with hematoxylin-eosin showed an epithelial growth in the superficial and middle dermis, composed of cells with a pale eosinophilic cytoplasm and rounded, monomorphous nuclei. The cells were arranged in tubules and solid nests, some with a tadpole shape. The neoplasm showed a sclerotic stroma (Figure 2).

The diagnosis based on clinical presentation and pathology was generalized syringomas. The patient was informed of the possible therapeutic options, initially choosing to refrain from treatment.

Syringomas are uncommon neoplasms, described mainly in women before and during puberty, probably in relation to the presence of hormone receptors and the fact that dermatology consultation for cosmetic purposes is more common among women. They manifest as firm, rounded, pale pink or yellow-colored papules of 1-3 mm diameter. Syringomas have traditionally been classified according to site and form of onset on eyelids, the most common, and eruptive.[1] In 1987, Friedman and Butler[2] attempted to classify the multiple sites and described 4 clinical forms (Table): localized, familial, Down syndrome-associated, and generalized. Generalized syringomas included a multifocal form and another, more common, eruptive form. The eruptive form, described by Jaquet and Darier in 1887 as "eruptive hidradenoma," is characterized by outbreaks of papular elements in the anterior and lateral region of the neck, trunk, axillae, abdomen, genital area, and limbs between 4 and 10 years of age. It has been related to heat stimuli.[3]

Eruptive syringomas have been associated with Down syndrome, palpebral syringomas with Marfan and Ehlers-Danlos syndromes, and syringomas presenting with milia and atrophoderma vermiculata have been associated with Nicolau Balus syndrome. Cases of familial inheritance and others related to diabetes, alopecia,[4] and tumors, such as carcinoids,[5] have also been reported.

There are atypical presentations such as unilateral forms,[6] urticaria pigmentosa, or milia.

Table. Clinical Variants of Syringomas

Localized
Solitary
Multiple-single foci
– Papular
• Infraocular
• Genital
• Acral
• Unilateral
• Frontal
– Occult
• Scalp: alopecia
– Simulating lichen planus
• Genital
– Simulating milia
• Infraocular
• Perianal
In unilateral plaque
Generalized
Multifocal
Eruptive
– Simulating lichen planus
– Simulating urticaria pigmentosa
– Simulating milia
Down syndrome
Familial

Adapted from Friedman and Butler.[2]

These tumors show a normal analytical profile, although 1 incidental case with elevated carcinoembryonic antigen has been published.[7]

Pathology studies have shown an epithelial growth in the upper half of the reticular dermis consisting of cells with a pale eosinophilic cytoplasm arranged in nests or tubules and surrounded by a sclerotic stroma. The tubular areas contain basophilic granular cells and ductal differentiation with central lumens lined with a compact eosinophilic cuticle. Epithelial growths in the form of a "tadpole's tail" or "comma" is characteristic.[8] The variant known as clear-cell syringoma is more common among diabetics and is characterized by glycogen-laden cells.

Immunohistochemical studies of this tumor show positivity for EKH-6, which would support the eccrine ductal origin. The description of eczematous lesions that leave eruptive syringomas as sequelae would raise the hypothesis that the classic eruptive form is actually a reactive hyperplasia against inflammatory processes in the sweat gland ducts.[9]

The histological differential diagnosis should be done with milia, microcystic adnexal carcinoma, and desmoplastic trichoepithelioma.

These tumors may benefit from physical treatments such as superficial cryotherapy, fulguration, and electrodesiccation, or from chemical agents such as isotretinoin, tretinoin, adapalene, or a 1% aqueous topical solution of atropine. At present, the best treatment is considered to be ablation with ultrapulsed CO_2 laser and preliminary treatment with trichloroacetic acid to minimize scarring.[10] None of these treatments are considered satisfactory or prevent recurrences.

Because of the age at onset, the fact that the condition did not always appear in outbreaks, and that it affected various skin areas, including the eyelid, we considered our patient to present a form of multifocal generalized syringomas that started on the eyelid.

Acknowledgements

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Psoriasis at the Site of Healed Herpes Zoster: Wolf's Isotopic Response

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To the Editor:

A wide variety of dermatological processes can occur at the site of healed herpes zoster, mainly granulomatous processes, lymphomas, pseudolymphomas, and primary skin tumors or metastasis.[1] These conditions occasionally appear in immunosuppressed patients with neoplasms or human immunodeficiency virus infection, but in other patients there may be no underlying disease. The interval between viral infection and second disease is extremely variable, from days to years.[2] We describe a patient with paroxysmal nocturnal hemoglobinuria who developed guttate psoriasis lesions on the site of previous herpes zoster.

A 41-year-old man who had undergone allogeneic transplantation of bone marrow for paroxysmal nocturnal hemoglobinuria and received