Generalized Eruptive Syringoma in Three Patients; Can Generalized Eruptive Syringoma be a Reactive Dermatosis?

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Abstract

Syringoma mostly presents with skin-colored or slightly yellow-brown colored papules on the periorbital region. Generalized eruptive syringoma is a very rare variant. We report three cases of generalized eruptive syringoma in patients with mycosis fungoides and intraductal papilloma.

Introduction

Syringoma appears as skin-colored or yellow colored papules mostly on periorbital region in adolescent females.1 Eruptive syringoma is a very rare variant of syringoma. The pathophysiology is poorly understood. They are considered benign adnexal tumors of eccrine sweat ducts or hyperplastic response of the eccrine ducts to an inflammatory reaction.1,2 Eruptive syringoma can present multiple papules on the neck, anterior trunk, upper and lower extremities or pubic area. It’s more common in women, and it can be seen in the prepubertal age as well as in the postpubertal age.3 Eruptive syringoma may associate diabetes mellitus, Down syndrome, Costello syndrome and Marfan syndrome.4,5,6,7,8 To our knowledge, no eruptive syringoma with mycosis fungoides and with other primary cutaneous lymphomas has been reported so far. We present here three generalized eruptive syringoma cases.

Case 1: 44 year-old female patient who was diagnosed as mycosis fungoides stage 1A with clinical and histopathological findings (Fig. 1) in January 2010 presented with wide-spread yellowish pruritic papules (Fig. 2) in June 2021 at Bezmialem Hospital.

She was treated with topical therapy for mycosis fungoides with good response and no progress was observed in the meantime. A new punch biopsy was performed from a lesion on the left arm because those lesions did not resemble mycosis fungoides. Characteristic features consistent with syringoma was displayed as dilated, cystic, comma-shaped eccrine ducts resembling “tadpoles” lined by two rows of flattened epithelial cells with a central lumina lined by compact eosinophilic cuticle (Fig. 2).

A diagnosis of generalized eruptive syringoma was confirmed based on clinical and histologic findings.

Case 2: 48 year-old female patient admitted with mildly pruritic skin-colored papules on her face and forearms to the out-patient clinic of Bezmialem Vakif University Hospital (Fig. 3). She had a story of two years with these lesions. She had an mammalian intraductal papilloma history also and it was removed surgically a year ago. A punch biopsy was performed on the left cheek. Characteristic features consistent with syringoma was observed as dilated,
**Figure 1:** a) Mycosis fungoides lesion on patient’s hip. b) Histology of punch biopsy taken from patient’s hip; Fibrosis in papillary dermis and lymphoid cells with dense chromatin (HEX400). c) In the epidermis, lymphoid cells with small dense chromatin surrounded by a halo (HEX400).

**Figure 2:** Yellow-brownish papules on left side of the trunk (a) and forearms (b). Dilated, cystic, comma-shaped eccrine ducts resembling “tadpoles” lined by two rows of flattened epithelial cells with a central lumina lined by compact eosinophilic cuticle (HEX400) (c). Nests and ducts with fibrotic stroma in the superficial epidermis (HEX40) (d).
Figure 3: Skin-colored papules on patient’s face (a) and forearms (b). Comma-shaped eccrine ducts resembling “tadpoles” lined by two rows of flattened epithelial cells with a central lumina lined by compact eosinophilic cuticle (HEX400) (c).

Figure 4: Non-follicular, papular lesions on the anterior and lateral trunk and arms (a, b, c). Ductus-like structures covered with small round-nucleated uniform cells and eosinophilic secretion in the lumens (HEX400) (d, e).
cystic, comma-shaped eccrine ducts resembling “tadpoles” lined by two rows of flattened epithelial cells with a central lumina lined by compact eosinophilic cuticle (Fig. 3).

A diagnosis of generalized eruptive syringoma was confirmed based on clinical and histologic findings.

**Case 3:** 43 year-old woman presented with pigmented eruptions on her body and arms to State Hospital of Tokat (a city in mid Anatolia). Personal and family history of the patient was unremarkable. In physical examination, pigmented, non-follicular, papular lesions that do not tend to coalesce were observed on the anterior and lateral trunk and arms (Fig. 4). Histopathological examination revealed a tumoral area consisting of ductus-like structures covered with small round-nucleated uniform cells and eosinophilic secretion in the lumens were observed in the dermis (Fig. 4). Patient has been diagnosed with generalized eruptive syringoma with clinical and histologic findings and the patient preferred follow-up without treatment.

**Discussion**

Syringomas are considered as benign neoplasms of intraepidermal eccrine duct origin. Four different types of syringomas have been described. Classification criteria proposed by Friedman and Butler divide syringomas into four variants based on clinical features: localized, familial, Down syndrome-associated, and generalized, inclusive of the eruptive type. Syringomas typically present as multiple, firm, skin-colored to yellow-brown papules, 2 mm to 4 mm in diameter, often in a symmetric distribution in the periorbital region. Syringomas occur most commonly in women during adolescence and in early adulthood. Unusual locations are also reported such as limited to scalp, vulva and penis. Syringomas have been reported in association with systemic diseases. The incidence of syringoma has been reported in up to 40% of patients with Down syndrome, particularly in females. Diabetes mellitus is associated with clear cell syringoma, consisting of nests of clear cells containing glycogen as the result of aberrations in glucose metabolism. Syringomas have been observed rarely to associate tumors and tumor-related syndromes such as Brooke-Spiegler syndrome, Nicolau-Balus syndrome and Costello syndrome. Syringoma presentation also reported in Marfan syndrome and Ehler-Danlos syndrome.

The pathophysiology of syringoma remains unlightened. Although syringoma may be considered as a neoplasm of eccrine or apocrine origin, in fact syringoma is a benign adnexal tumor derived from the intraepidermal portion of eccrine ducts. Staining with anti-keratin antibodies EKH4 and EKH6 supports this consideration. Some authors suggest that syringomas develop after reactive eccrine hyperplasia and therefore syringomas can not be regarded as true tumors. Syringomas arising after eczematous eruptions and within “waxed” regions of the pubic area support this theory. Incel et al reported an adult-onset eruptive syringoma in a 53-year-old man and suggested the possibility of its association with renal cell carcinoma as a paraneoplastic phenomenon. We here presented cases of eruptive generalized syringoma associated with primary cutaneous lymphoma mycosis fungoides stage 1a and intraductal papilloma. Although it is not possible to attribute the development of eruptive syringoma to mycosis fungoides or intraductal papilloma, but this association may suggest the possibility of generalized eruptive syringoma being as a reactive dermatosis.

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The patients in this manuscript have given written informed consent to publication of their case details.

**References**