A Case Report of Eruptive Syringoma with Koebner Phenomenon

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ABSTRACT:

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Syringoma is considered to be benign neoplasm of the eccrine sweat ducts. The lesions usually symmetrical bilateral occurred in crops with flat topped, angulated, skin-colored to slight yellow, shiny papules, commonly asymptomatic; however, pruritus can sometimes occur. They were classically found in the periorbital region. However, the lesions with same morphology, but with prominent on ventral surface of the upper extremities and upper chest wall should raise the suspicious of another variant of syringoma including eruptive syringoma. Moreover, eruptive syringoma could be presented with the linear configuration which reflects Koebner phenomenon, hypothesized from trauma-induced process.

Key words: Syringoma, eruptive syringoma, Koebner phenomenon, syringoma with Koebner phenomenon

Case Presentation

We reported a case of 24-year-old Thai male presented with 10-years history of multiple small nodules on both arms and chest wall which was predilected on the ventral surface of the body (Figure 1). He did not notice the spreading pattern whether the lesions presented as successive crops. The patient did not have any itchy or painful symptom. None of the family members was affected by the similar skin lesions. He had no underlying disease such as Down's syndrome.

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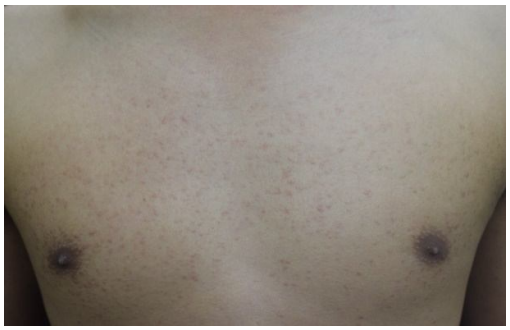

Figure 1A

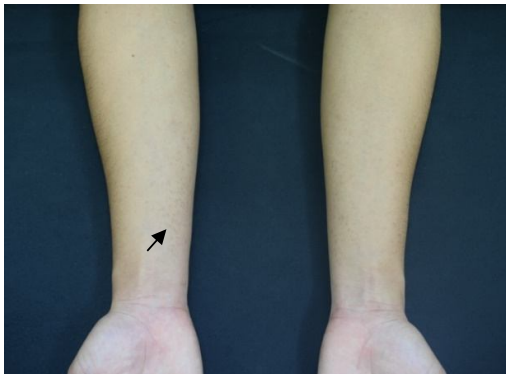

Figure 1B

Figure 1 A. Multiple ill-defined border, skin-colored, flat topped papules on upper chest. **B.** Small flat topped papules with linear configuration on both forearms.

The physical examination showed multiple ill-defined borders, skin-colored, monomorphic, flat topped papules with linear configuration on both forearms in flexor side and upper chest. Other physical examinations were unremarkable.

The histological findings from punch biopsy of the papule on right arm revealed the circumscribed nodular infiltration in upper dermis with strands of basaloid calls with duct formation in fibrous and sclerotic stroma which was compatible with typical findings of syringoma (Figure 2).

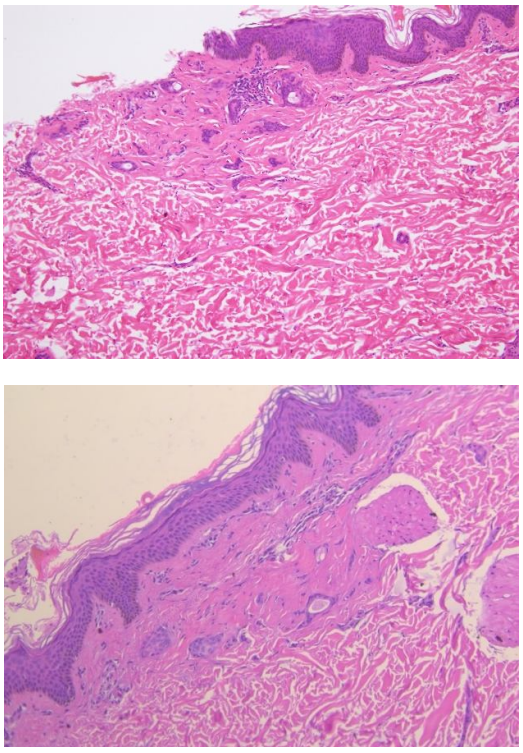

Figure 2 Circumscribed nodular infiltration in upper dermis with strands of basaloid calls with duct formation in fibrous and sclerotic stroma. The comma-like tails of epithelial cells contribute to the tadpole shape of the adnexal neoplasm. H&E X10

The treatment options were discussed and explained to the patient as he decided not to do

any treatment because of uncertain outcomes in many studies.

Discussion

Syringoma is a benign adnexal tumors, formed by well-differentiated ductal elements. The incidence was approximately 1% of population with female predominance[1,2]. It usually occured before or during puberty. Its association with Down's syndrome has been confirmed by the Down's syndrome-associated type of syringoma. The association of syringoma with milium, elevation of carcinoembryonic antigen, diabetes, psychiatric disorders, Ehler-Danlos syndrome, prurigo nodularis, melanocytic nevi, and sarcoidosis has also been reported[3,4,5].

The lesions usually symmetrical bilateral occured in crops with flat topped, angulated, skin-colored to slight yellow, shiny papules, commonly asymptomatic, but sometimes pruritus can occur[6]. They were classically found in the periorbital region. Other sites of involvement included neck, supraclavicular region, anterior and posterior aspect of trunk[4].

There were some studies providing hypothesis about the association between syringoma and hormonal influence because of higher incidence of syringoma in female and the lesions frequently developed before or during puberty. Moreover, it was reported that disease exacerbation during pregnancy was also found. Two studies also noted the strong expression of progesterone receptors by immunohistochemistry[7].

Syringomas was classified into 4 types based on location and association: localized, familial, generalized (multiple and eruptive), and Down's syndrome-associated[8].

Eruptive syringoma is a rare variant of syringoma, first described in 1887 by Jacquet and Darier, presented with the large numbers of successive crops occur during childhood or adolescent on ventral side of body. The lesions are usually found on anterior chest, neck, upper abdomen, axillae, periumbilical region with almost multiple in numbers. More are frequently seen in patient with Down's syndrome and Ehlers-Danlos[8].

Ibekwe P. also reported a 13-year old girl with multiple discrete asymptomatic dark-brown to reddish-brown dome-shaped firm papules that firstly erupted on her inner surface of the forearms. The diagnosis was established as eruptive syringoma which was confirmed by histological findings from the lesion[6].

The pathophysiology of eruptive syringoma was still not fully understood but it was proposed to be the previous cutaneous inflammatory process which led to hyperplastic reaction in eccrine duct. Many cases of syringoma were reported after contact dermatitis, shaving, hair removal laser, alopecia areata, radiation dermatitis, and neoplasms[3,9].

Histologically, syringoma were characterised by dilated, single-to double-layered cuboidal epithelial cell of cystic eccrine ducts. The cysts resembled small tails that looked like commas or tadpoles and surrounded by collagen bundles in the upper dermis[4,6].

In our patient, the morphology of the lesion was mimicking lichen nitidus, plane warts, lichen scrofulosorum, and syringoma, but the papules were prominent on both arms and upper chest wall which were not the typical location of syringoma. Therefore, other variant of syringoma including eruptive syringoma were in the differential diagnosis. The histopathology of the skin lesion revealed duct formation embedded in a fibrous stroma which was compatible with syringoma.

Moreover, we also noted the group of Koebner phenomenon of the lesions which can be found in another dermatoses includes psoriasis, vitiligo, lichen planus, Darier disease, warts, vesiculobullous diseases, and lichen nitidus. To the best of our knowledge, no study ever reported the Koebner phenomenon of eruptive syringoma. We hypothesised the trauma-induced process could be the etiology of Koebner phenomenon of the lesions in this patient.

Efficacy was limited in available treatment option because the tumors were located in the dermis and high risk of recurrence. Many modalities of treatment may lead to scarring which were often unsatisfactory. Some of therapeutic modalities including very light electrodessication, removal by shaving, and carbon dioxide laser (by pinhole method and fractional thermolysis) were reported as successful method. For larger lesions, surgical removal may be considered. Other treatment modalities included cryosurgery, chemical peeling, dermabrasion, and oral or topical retinoids. The treatment options were discussed and explained to the patient. Our patient decided not to do any treatment because of uncertain outcomes in many studies[10].

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