Eruptive Syringoma: Clinicopathologic Analysis of Thirteen Cases

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Background **: Eruptive syringoma is a very rare variant of syringoma and arises most frequently on the anterior trunk of young people. Because eruptive syringoma represents clinically a very rare distinct entity, clinicopathologic studies rarely have been described in literature.**

Objective **: The aim of this study is to characterize clinical and histopathologic findings of eruptive syringomas which were diagnosed by histopathologic examination for a 10 year period in Asan medical center.**

Methods : The hospital records, clinical history, and biopsy slides of 13 eruptive syringoma were reviewed retrospectively.

Result **: There were 5 men(38%) and 8 women(62%) with the mean age of 31 years. The mean age of onset was 23 years and 3 patients(23%) presented the lesions before puberty. The most frequently involved sites were anterior trunk, axilla, upper extremities, neck, and face in descending order. The most common initial clinical diagnosis was verruca plana(38%). Most of the lesions did not show self-limiting course and several treatment modalities including oral isotretinoin or CO_2 laser were not so effective. All cases showed characteristic histopathologic features of syringoma and 76% of them revealed increased basal melanin pigment or pigmentary incontinence.**

Conclusion **: Eruptive syringoma should be included in the differential diagnosis of flat papular lesions by histopathologic examination and it may help to avoid inappropriate therapeutic approach. (Ann Dermatol 14(3) 143~148, 2002).**

Key Words : Eruptive syringoma

Syringoma is a common tumor of eccrine sweat gland origin, which usually appears on the eyelids of healthy middle-aged women[1]. On the other hand, the eruptive syringoma is a rare variety that arises in prepubescent children or young adults and distributed over the anterior surfaces of the body but may concentrate on the chest, neck, axillary fossa, abdomen and inner surfaces of extremities[1,2]. Because eruptive syringoma represents clinically a very rare distinct entity, clinicopathologic studies have not been reported in the Korean literature.

The aim of this study is to report our experience regarding eruptive syringoma by reviewing the clinical and histopathologic features of series of 13 patients with histologically diagnosed syringoma observed for a 10 year period.

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MATERIALS AND METHODS

Thirteen cases with a histopathological diagnosis and clinical history of eruptive syringoma ob-

Table 1. Epidemiologic and clinical characteristics of 13 patients with eruptive syringoma

No	Age/Sex	Onset age	Localization	Clinical diagnosis	pruritus	combined disease
1	48/F	42	FA,N,UE	Verruca plana	+	-
2	22/F	18	UE,A,LE	Verruca plana	-	-
3	30/F	25	FA,N,CA,AB,P,I	Darier's disease	-	-
4	26/F	24	N,A,P	Eruptive syringoma	-	Plantar warts
5	27/M	17	A,C,AB,UE,LE	Urticaria pigmentosa	+	Androgenic alopecia
6	18/F	14	N,A	Eruptive syringoma	-	-
7	28/M	18	C,AB,BA	Eruptive syringoma	+	-
8	19/F	9	UE,AB,G	Lichen nitidus	-	Plantar warts
9	25/M	15	C,AB	Verruca plana	-	-
10	28/F	26	AB,G	Verruca plana	-	Chronic superficial gastritis
11	27/F	23	A,C,AB	Eruptive syringoma	-	Alopecia areata
12	53/M	30	C,AB,B	Urticaria pigmentosa	+	-
13	47/F	39	FA,UE,C	Verruca plana	-	Hyperlipidemia, chronic colitis

M:male, F:female, FA:face, N:neck, UE:upper extremities, A:axilla, LE:lower extremities, AB:abdomen, P:pubic area, I:inguinal area, C:chest, G:genitalia, BA:lower back

served in the dermatologic department of Asan medical center between 1992 and 2001 were selected for this retrospective study. The hospital medical records and biopsy slides were reviewed. Clinical evaluations were performed regarding to age, sex, age of onset, familial history, characteristics of individual lesions, localization, symptoms, combined systemic or skin disease, and treatment.

RESULTS

Epidemiological and clinical features of the patients diagnosed with eruptive syringoma are summarized in Table 1.

1. Epidemiological description

There were 5 men(38%) and 8 women(62%) with the age from 18 to 53 years(mean 31 years). Age of onset of eruptive syringoma varied from 9 to 42 years(mean 23 years). Only three patients(23%) presented lesions before the age of 15 years(mean of puberty), in the remaining 10 patients(77%) the lesions appeared after this age and were considered as postpubertal. There was no patient with familial history of eruptive syringoma.

2. Clinical features

The most frequent lesions were round to oval, slightly raised, 1-5 mm sized, firm, brownish papules and some represented yellowish or erythematous papular lesions. The most frequently involved body sites were anterior trunk (including chest and abdomen), axilla, upper extremities, neck, and face in descending order(Fig. 1). The lower extremities, genitalia, inguinal area, and lower back were also involved in some patients(Table 2). Three patients(23%) had symptom of pruritus. The presumptive clinical diagnoses before the histopathologic examination are summarized in Table 1. Eruptive syringoma was considered as the initial diagnosis in only 4 patients(31%). The

Table 2. Localization of eruptive syringoma

Localization	No. of patients
Anterior Trunk	9
Axilla	6
Upper extremitie	5
Neck	4
Face	3
Lower extremities	2
Pubic area	2
Genitalia	2
Inguinal area	1
Lower back	1

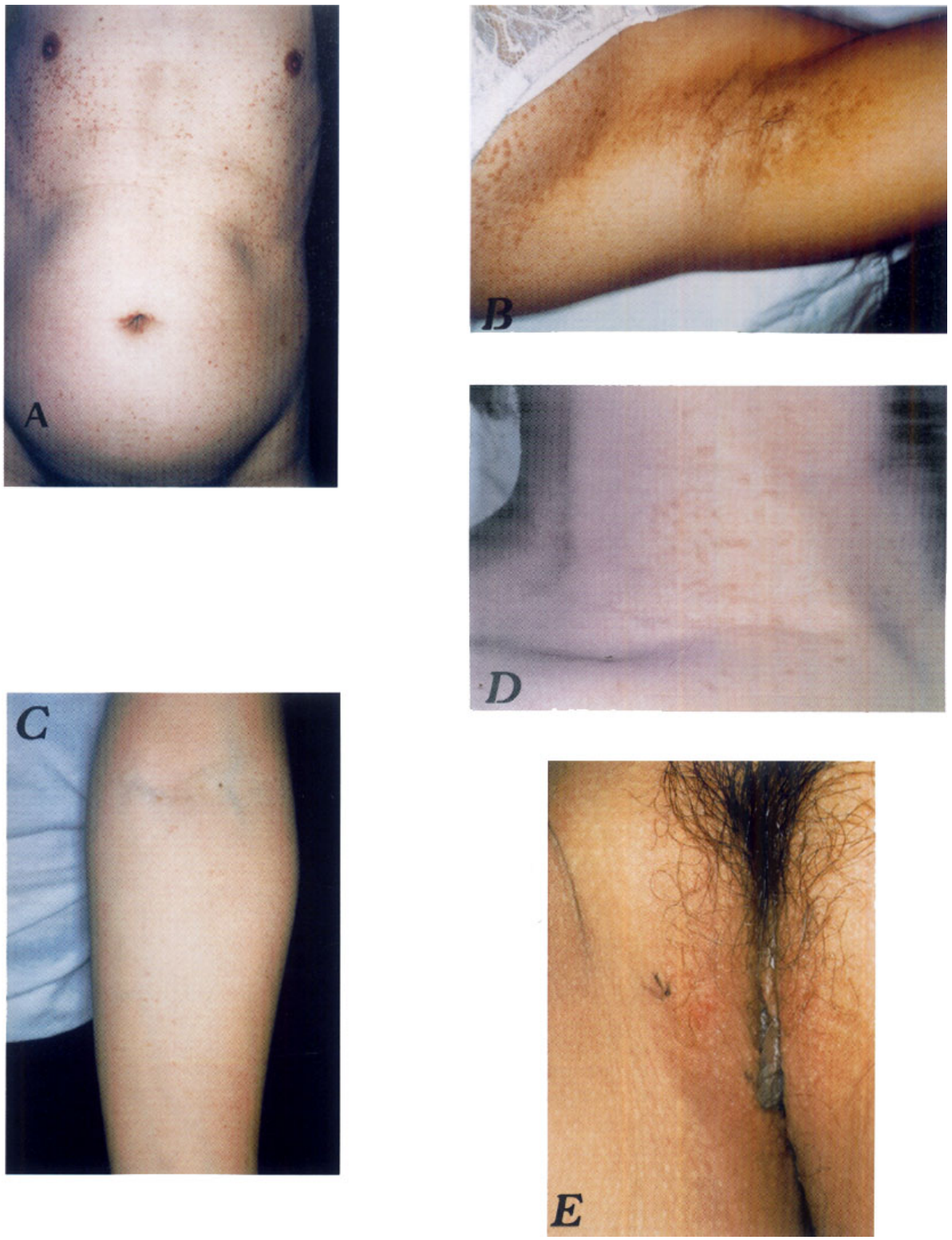

Fig. 1 Multiple, discrete, erythematous to brownish, flat-topped papules on the trunk (A,case12), axilla (B,case3), upper arm (C,case1), neck (D,case6), and vulva (E,case10).

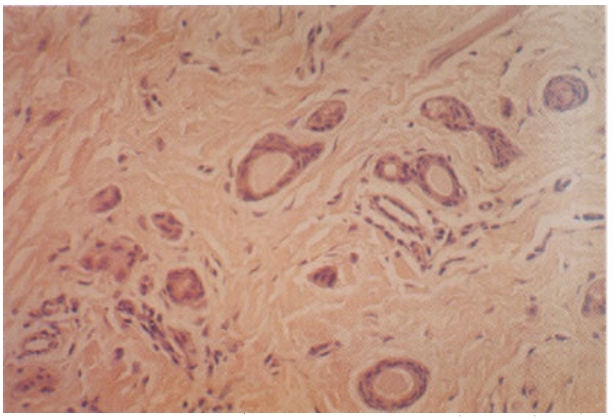

Fig. 2. Characteristic "tadpole" from eccring gucts in a collagenous stroma(H&E, × 100).

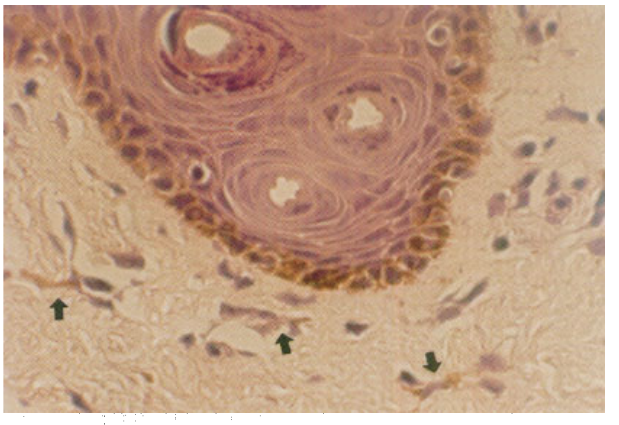

Fig. 3. Increased melanin pigment in basal layer with melanophage(arrow) in upper dermis(H&E, × 400).

most frequent clinical diagnosis was verruca plana(5 of 13, 38%). Three of them had been treated with diphencyprone(DPCP) before the histopathologic diagnosis without improvement. Two patients(15%) had had combined internal illness including chronic superficial gastritis and hyperlipidemia with chronic colitis. Other 4 patients had had other skin problems such as plantar warts(2 patients), alopecia areata(1 patient), and androgenetic alopecia(1 patient). Eruptive syringoma did not show self-limiting course in any case of our studies except one case with recurrent regression and reappearance in a 19-year-old male patient. Four patients had been treated with oral isotretinoin(Roaccutane^{R}), 30mg/day, for each one to three months without improvement. Other three ones had been treated with CO_2 laser on some lesions, but they complained of hyperpigmented scars and refused to continue the treatment.

3. Histopathologic features

All biopsy specimens showed characteristic histopathologic features of syringoma including upper and some mid-dermal proliferation of eccrine duct and solid epithelial cords in a collagenous stroma. Typical "comma" or "tadpole" forms were observed in all cases(Fig. 2). In 3 patients(23%), ductal proliferation was extended to the mid-dermis. One case was desmoplastic type. Increased melanin pigment of basal layer was found in 10 of 13 patients(76%). Six of them showed pigmentary incontinence or melanophages in upper dermis(Fig. 3).

DISCUSSION

Syringoma is a benign tumor which derives from intraepidermal eccrine ducts[1]. Although syringomas appear most commonly on the eyelids in middle-aged women, many other clinical variants differing in age of onset, location, and clinical aspect have been reported in the literatures[1-3]. Friedman and Butler[4] proposed a classification of syringoma according to clinical features and association. This consists of four principal clinical variants: a localized form, a familial form, a form associated with Down's syndrome, and a generalized form which includes eruptive syringoma[5]. The localized form is most commonly seen in the periocular region, but less frequently reported on the scalp, forehead, genitalia(penis or vulva), and acral sites such as hands[6-11]. This disorder is generally described as sporadic, but there have been some reports indicative of familial occurrence[12]. There have also been some case reports of eruptive syringoma associated with Down's syndrome[13,14]. Butlerworth et al[14] insisted that the incidence of syringoma within Down's syndrome is 18.5% and this is 30 times greater than normal population.

Eruptive eccrine hidradenoma or syringoma is a rare clinical variant and six case reports and one electronmicroscopic study of a syringoma have been described in Korean literature[15-21]. Eruptive syringoma is usually seen on the chest of young adults and the abdomen, axilla, neck, shoulder, pubic area, and face also can be noted[6]. The lower extremities or inguinal area can also be rarely in-

volved, but extending below the umbilicus is unusual[8]. Genital lesion is also known to be very rare[8]. In our study, the most frequently involved sites were anterior trunk, axilla, upper extremities, neck, and face in descending order. Unusual present sites such as genitalia(2 patients), inguinal area and lower back(each of one patient) were also involved in our study. The facial involvement with trunk lesion was observed in three patients. Besides these, eruptive syringoma with unilateral eruption, unilateral linear nevoid distribution, lichen planus pattern, milia-like pattern, and urticaria pigmentosa like type also have been described in literatures[1,6,22-25].

The syringoma present in 0.6% of the population but the eruptive form is rare[3]. It occurs more frequently among women. Eight of 13 cases(62%) in our series were women and this is slightly less than the previous reports(75-90%)[3]. In the previously reported cases, 33% to 50% of patients experienced the eruptive syringoma before the age of 15[3,26]. However, in our series, only three cases(23%) presented the lesions before the age of 15 years. Clinical presumptive diagnosis of eruptive syringoma was only considered in four patients(31%). Other initial diagnoses were verruca plana(5 patients), urticaria pigmentosa(2 patients), Darier's disease(1 patient), and lichen nitidus(1 patient). Verruca plana was the most frequent clinical diagnosis. Three of the patients who were diagnosed with verruca plana before histopathologic examination had been treated with diphencyprone(DPCP) without improvement. Therefore, even though the lesions which are considered as verruca plana or other flat papular diseases should be confirmed by histopathologic examination[27].

Characteristic histopathologic features of syringoma are upper dermal non-encapsulated proliferation of ducts and solid epithelial cords in a loose collagenous stroma. Typical 'comma' or 'tadpole' forms are evident[28]. The histopathologic feature of eruptive syringoma is not different from common syringoma. In our study, all biopsy specimens showed typical features of syringoma. Furthermore, 10 cases(76%) revealed increased melanin pigment in basal epidermis and six of them showed pigmentary incontinence with some melanophages in upper dermis. We think that this is the possible cause of brown color of most eruptive syringomas clinically.

The pathogenesis of eruptive syringoma is unclear[28]. An early theory by White[29] suggested an influence of high altitude on the sweat apparatus. More recently, some reports of eruptive syringomas restricted to the sites of urticaria pigmentosa lesions suggested a role for mast cell mediators[25]. However, the exact mechanism is still unknown. Some authors in the literature stressed the association of eruptive syringoma and other systemic diseases including malignant neoplasm or diabetes mellitus, and some genodermatoses such as Nicolau-Balus syndrome, Marfan syndrome, and Ehlers-Danlos syndrome[3,30-32]. In our patients, however, there was no association with genodermatoses or systemic diseases except two cases of combined chronic superficial gastritis and hyperlipidemia with chronic colitis.

There is no satisfactory treatment for eruptive syringoma[3]. Surgical methods including electrodissection, cryotherapy, CO_2 laser ablation, and excision may produce cosmetic problems of scars[3,33]. Chemical therapy such as topical or systemic retinoids have been used by some clinicians with some improvement[3,34-36]. The use of them, however, has become controversial. In our study, 4 patients were treated with oral isotretinoin and 3 other patients with CO_2 laser ablation. However, there was no marked clinical improvement and most patients had withdrawn from therapy. Some authors insisted that this benign appendageal tumor may regress spontaneously[28]. One patient in our series also experienced the spontaneous regression and reoccurrence of the lesions, but this is known to be a very unusual event and never proven.

In conclusion, eruptive syringoma is a unusual clinical entity and can be presented in young adults as brownish flat papular lesions, especially on the trunk, like verruca plana. We recommend that flat papular lesions should be diagnosed correctly by histopathologic examination. Exact diagnosis of eruptive syringoma is important since it may lead to unnecessary over-treatment.

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