Familial eruptive syringoma in a Thai woman: Case report and literature review

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ABSTRACT:

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Syringoma is a benign, adnexal tumor of eccrine sweat duct origin, for which familial eruptive forms have rarely been reported. We present the case of a 20-year-old Thai female who had a history of moyamoya disease and who developed numerous, widespread, skin-colored to hyperpigmented papules over the face, trunk, both axillae, and the groins. A skin biopsy confirmed syringoma. Her mother was also affected with the same lesions on her face. In addition, we conducted a literature review of cases of familial eruptive syringoma reported worldwide.

Key words: Familial eruptive syringoma, Hereditary syringoma, Generalized syringoma

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Introduction

Syringomas are benign, eccrine sweat gland tumors. They usually affect adult females, but they can occur in both sexes[1]. The lesions are usually multiple with a bilateral and symmetrical distribution, and they may present in a great number. Clinically, they appear as small, firm, smooth, skin-colored or slightly yellowish, 1–4 mm papules that commonly occur in the periorbital area, particularly around the lower eyelid[2,3].

The four clinical types of syringomas are classified according to their location and association: localized; familial/hereditary; a form associated with Down syndrome; and a generalized/eruptive form[4]. Eruptive syringoma is a rare variant that was first described by Jacquet and Darier in 1887[5]. Familial eruptive syringoma is a very rare clinical variant that combines the familial and generalized eruptive forms[2]. To date, only a few cases have been described in world literature (Table 1)[2,6–19].

Table 1 Case Reports of Familial Eruptive Syringomas

Report	No. of cases	Authors	Year of publication	Nation	Patient age (yrs) and sex	Age of onset	Site of lesions	Affected family members (area of involvement)
1	5	Woringer and Eichler[6]	1951	N/A	N/A, F	N/A	N/A	Father
					3 F	N/A	N/A	Sister
					N/A, F	N/A	N/A	Mother
					N/A, F	N/A	N/A	Grandfather
					N/A, F	N/A	N/A	Brother
2	2 (siblings)	Yesudian and Thambiah[7]	1975	India	19 M	18	Eyelids, neck	Brother
					21 M	N/A	Eyelids, neck	Brother
3	1	Hashimoto et al. [8]	1985	Iran	55 M	5–10	Eyelids, chest, back	Father, sister, two daughters (anterior neck, chest)
4	1	Crespo Erchiga et al. [9]	1987	N/A	9 F	N/A	Neck	Twin sister
5	1	Patrone and Patrizi [10]	1988	N/A	16 F	N/A	Anterior neck, chest	Mother (anterior neck, chest) Brother (anterior neck, axilla)
6	6	Patrizi et al. [11]	1998	N/A	N/A	N/A	N/A	N/A
7	1	Metze et al. [12]	2001	N/A	52 F	N/A	Face, neck, truck, extremities	Mother, daughter

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Report	No. of cases	Authors	Year of publication	Nation	Patient age (yrs) and sex	Age of onset	Site of lesions	Affected family members (area of involvement)
8	1	Smith and Skelton [13]	2001	N/A	19 M	Adolescence	Anterior truck (eruptive)	Mother
9	2	Soler-Carrilo et al. [14]	2001	Spain	33 F 16 F	10 12	Neck, eyelid Neck, trunk	Father, brother Father
10	1	Bautista et al. [15]	2003	N/A	32 M	N/A	Face, neck, anterior chest	Mother
11	5 (siblings)	Elsayed and Assaf [16]	2009	Egypt	23 F 27 F 36 F 22 M 32 M	N/A	Neck, axilla, abdomen Forearm, chest, axilla Forearm, chest, axilla Axilla, chest, neck, abdomen Axilla, chest, neck, abdomen	Father (eruptive), two sisters, two brothers Sister of the 1st case Sister of the 1st case Brother of the 1st case Brother of the 1st case
12	2	Marzano et al. [17]	2009	USA USA	36 F 17 M	N/A	Neck, chest, arms Truck, extremities	Son, mother, two brothers, two sisters Mother (son of the 1st case)
13	1	Lau and Haber [18]	2013	Asia	16 F	13	Axilla, abdomen	Mother, brother (arm, axillae, abdomen, groin)
14	1	Ibekwe[2]	2016	Nigeria	13 F	11	Periorbital, forearm, chest	Mother (local)
15	2 (siblings)	Yaldiz et al. [19]	2018	N/A	20 F 25 M	16 15	Neck, supraclavicular Upper chest, back	Brother Sister

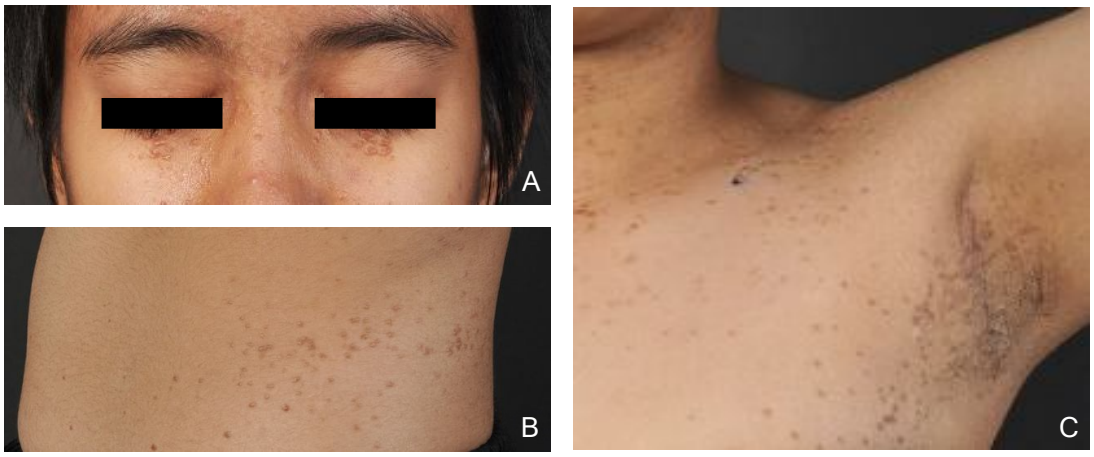

Figure 1 A-C Familial eruptive syringoma.

A) Multiple skin-colored papules on face predominated at both lower eyelids.

B) Multiple brownish soft papules on her trunk.

C) Multiple discrete brownish papules on her neck, upper chest, and axillae.

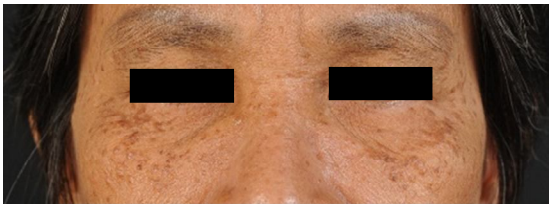

Figure 2 Familial eruptive syringoma; cluster of small, firm, brownish, flat-topped papules on the face of the patient's mother

Case Presentation

A 20-year-old Thai female patient presented to the dermatology department of Siriraj Hospital with a 2-year history of diffuse, numerous, skin-colored to hyperpigmented papules over the face, trunk, both axillae, and the groins. The majority of the lesions were present at the face. Although the lesions had initially developed over both lower eyelids, they subsequently gradually spread over the face, neck, trunk, both axillae, and the groins over 2 years, with rapid progress in the last 4 months of that period. The lesions then remained stable without progression elsewhere on the body. They were neither tender nor itchy, and they were not associated with local or systemic symptoms. The patient denied any history of trauma, insect bites, or chemical contact on the affected areas. The lesions were not precipitated by heat, sweat, or sunlight. They had previously been treated with a mild. potent, topical corticosteroid, but without improvement. The patient's underlying disease, moyamoya, was diagnosed after the onset of the rash. There was

no history of consanguinity. Although there was a family history of similar lesions in her mother, they were restricted to the periorbital areas.

A dermatological examination revealed multiple, discrete, well demarcated, firm, skin-colored to brownish, flat topped papules of variable size (1–4 mm in diameter); they had smooth surfaces and were noncrusted. They were also distributed bilaterally and symmetrically on both lower eyelids, the forehead, neck, chest, back, axillae, and groins (Fig. 1). The lesions did not show scaling, vesiculation, or erosion. The examination of the mucous membranes, palms and soles, hair, and nails was normal. Physical examination of the other body regions was unremarkable.

The mother had a cluster of small, firm, brownish papules on both cheeks and lower eyelids (Fig. 2).

The clinical differential diagnosis included eruptive syringoma, xanthoma disseminatum, generalized eruptive histiocytosis, and tuberous sclerosis.

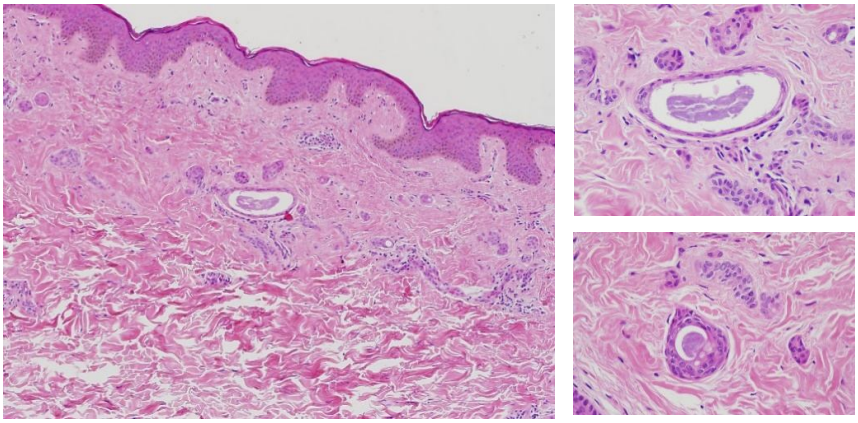

Figure 3 Histopathology showing numerous tubular structures lined by two layers of flattened cuboid epithelial cells which formed ducts and small nests with cords and stands and some filled with eosinophilic material in the dermis, embedded in an area of dense collagenous stoma. Some of the ducts show small, tadpole-like structures.

Laboratory investigations (comprising a routine hematological examination, and liver, renal and thyroid function tests) were all within normal limits.

A skin biopsy was performed from a lesion on the patient's left clavicle. A histopathological examination of the biopsy with haematoxylin and eosin staining revealed a normal epidermis and

dense collagenous stoma of the upper dermis, which presented multiple ducts and small nests with cord and stands formed by two layers of flattened cuboidal epithelial cells and fills with amorphous eosinophilic material. Some of the ducts showed small tails resembling commas or tadpoles (Fig. 3).

Based on the clinical and histopathological appearance of the lesions, a diagnosis of familial eruptive syringoma was made. After being informed that the lesions were benign, the patient and her mother decided to have no further intervention.

Discussion

The word syringoma is derived from the Greek word "syrinx", meaning pipe or tube[20]. Syringoma is a benign, adnexal tumor of intraepidermal eccrine sweat duct origin, which affects about 1% of the general population[17,21]. It usually affects adult females, but it can occur in both sexes[1]. While cases of syringoma usually first appear at puberty, there have been some reported occurrences in the elderly[22]. Syringomas are usually asymptomatic, but in rare cases, individuals may develop pruritus, especially during perspiration[17]. Clinically, the lesions are characterized as multiple, discrete papules or as clusters of small, firm, skin-colored or slightly yellow, papules, each 1–4 mm in diameter[2,3].

There are four clinical types of syringomas, classified based on their location and association:[4] localized; familial/hereditary; a form associated with Down syndrome; and a generalized variants including multiple that incorporates multiple body involvement and eruptive syringoma. The familial forms are those cases with a family history of syringoma[18]. Our case are familial eruptive syringoma which is a combination of the familial and generalized eruptive forms. Eruptive syringoma is a rare variant and was first described by Jacquet and Darier in 1887[5]. The peri-orbital region, particularly around the lower eyelid, is the most common site of involvement[3]. The eruptive nature of syringomas is almost always multiple and most frequently occur on the eyelids and upper cheeks[19]. The other surfaces that are commonly involved are the neck, axillae, upper chest, abdomen, periumbilical area, and genitalia, but the extremities (including the palms and soles) may also be involved[3,18,22]. There are some differences between prepubertal and postpubertal distribution patterns. The neck and anterior trunk are the most frequent sites before the age of 15 years followed by apocrine localizations: axillae or pubis. After 15 years of age, the apocrine localizations are rarely found[14]. The pathogenesis of eruptive syringomas remains unclear, but it is currently believed to be a neoplastic process[22]. The inheritance pattern is believed to be autosomal dominant, and the involved gene has been linked to chromosome

16q22[23].

The definitive diagnosis of syringoma can be made via histological examination. Histologically, numerous small ducts are embedded in a fibrous stoma, the walls of which are lined usually by two rows of epithelial cells. In most substances, these cells are flat, but occasionally, the cells of the inner row appear vacuolated. The lumina of the ducts contain amorphous debris. Some of the ducts possess small, comma-like tails of epithelial cells, giving them the appearance of tadpoles[2,3].

Eruptive syringoma can be associated with Down syndrome, Marfan syndrome, Nicolau–Balus syndrome (syringomas, milia, and atrophoderma vermiculata), Ehlers–Danlos syndrome, hyperthyroid, and diabetes mellitus[14,25,26]. Moyamoya disease can be associated with hyperthyroid[27]. However, no association with genodermatosis, abnormal thyroid function test or fasting blood glucose was found in our patient

Due to the fact that syringoma is a benign neoplasm and typically asymptomatic, no further intervention is needed[3,25]. Spontaneous resolution of the lesions is very uncommon[2]. Treatment of syringoma is performed for cosmetics reasons, but the outcomes are generally unsatisfactory. This is because the tumors are located in the dermis, so complete removal is often impossible, and the recurrence rate is high[2,19,22]. Based on our systematic review of the literature, both medical and surgical treatment modalities have had variable success[25]. The surgical interventions have included the use of electrodessication or shave removal, carbon dioxide laser, fractional thermolysis, cryotherapy, chemical peeling, and dermabrasion[19,25]. However, scarring and dyspigmentation are common complications of destructive interventions[25]. The medical treatments have largely involved the administration of oral topical retinoids; there have also been some reports of the successful treatment of symptomatic eruptive syringomas with 1% topical atopine[25]. In our case, the patient decided to receive no intervention.

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