Case 21

A 45-year-old woman from Ubonrajathani

Chief complaint: Asymptomatic hyperpigmented papules on the face

Present illness: The patient gradually developed asymptomatic hyperpigmented papules on her face for 2-3 years. The papules increased in number and size. She had no systemic symptoms.

Past history: Gastritis, anemia, no history of drug allergy.

Personal history: 10 years PTA she had similar skin lesion and biopsy showed sebaceous gland hyperplasia. The lesions did not improved with topical medication. After that she went to a private clinic for laser treatment of the lesions but with recurrence.

Family history: Not remarkable

Physical examination:

HEENT: not pale, no jaundice, no parotid gland enlargement

LN: no lymphadenopathy

Heart and Lung: WNL

Abdomen: no superficial vein dilatation, no distension, liver and spleen are not palpable

Extremities: no edema, no palmar erythema

Skin examination:

Confluent small shiny flat top papules at face, pinna, forearm, lower leg.

Laboratory investigation: (13/6/08)

CBC: Hb10.9 g/dl Hct 33.9% WBC 4270 (N 44, L43,M10,E2)

Lipid profile : TG 75 mg/dl, Chol 180 mg/dl, HDL 53 mg/dl, LDL 113mg/dl

TFT: FT3 2.68pg/ml, FT4 1.1 ng/dl,TSH 1.26 uIU/ml

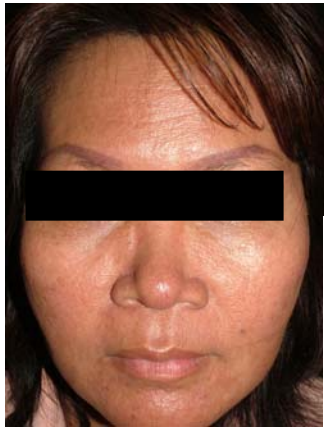

Fig. 21.1

Fig. 21.2

Fig. 21.3

Fig. 21.4

Histopathology (S08-11025) (Fig 21.5, 21.6)

- Nests, cords and ducts lined by ductal cubal cells, in dense collagenous stroma

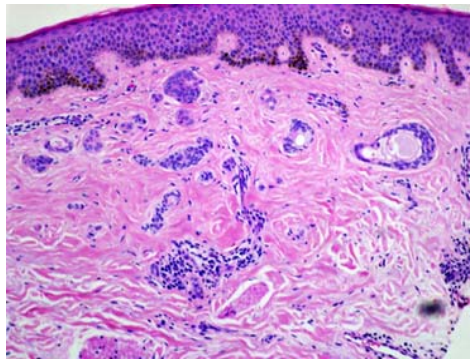

Fig. 21.5

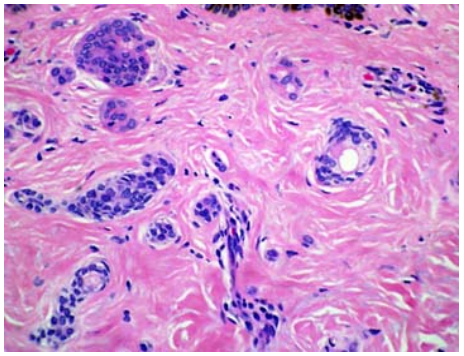

Fig. 21.6

Diagnosis: Eruptive syringoma
Presenter: Waraphorn Apasrawirote
Consultant: Penpun Wattanakrai

Discussion:

Syringoma is a benign adnexal tumors derived from the intraepidermal portion of eccrine sweat ducts. The prevalence is about 0.6% of general population, more common in females[1,2], Japanese people, and patients with Down syndrome. It appears during puberty or the third or fourth decade[3,4]. Clinically, they usually presents in asymptomatic multiple symmetrically distributed, soft, small skin-colored or slightly yellow papules ranging from 1-3 mm in diameter on lower eyelids[2,5]. Clinical classification of syringoma consists of four groups: a localized form, a familial form, a from associated with Down syndrome, and a generalized form that encompasses multiple and eruptive syringoma[6]. Differential diagnosis from clinical presentation are closed comedones, sebaceous gland hyperplasia, milia, lichen planus, eruptive xanthoma, urticaria pigmentosa, or hidrocystoma[7].

Definitive diagnosis can be made on histological examination, because syringoma demonstrate distinctive histopathological features. The dermis shown numerous, small ducts lined with a double row of flattened epithelial cells. Often outer layer extends into the surrounding stroma, forming a comma-like projection called “tad pole” appearance. Ductal lamina are filled with an amorphous, periodic acid-Schiff-positive material. Histochemical and electron microscopic findings have confirmed that syringomas represent adenomas of eccrine sweat ducts[7]. Patients with diabetes mellitus may present with a histological variant known as clear-cell syringoma[8].

Eruptive syringoma is a rare variant of syringoma describe by Jacquet and Darier in 1987[9], which occur in large numbers and in successive crops on the anterior aspect of body’s surface including chest, neck, upper abdomen, axillae, and periumbilical region. Sparing plantar foot and mucosa[10,11]. Unusual locations include the vulva[12], penis[13], and scalp that associated with diffuse thinning of the hair[14]. Unilateral, linear arrangement of syringoma is rarely[15]. Acral syringomas, located on distal extremities, are extremely rare, with only five cases reported in the dermatological literature[5]

The pathogenesis of eruptive syringoma is not yet completely clarified[1]. Recent studies suggest a reactive hyperplastic process in the eccrine duct, resulting from a prior cutaneous inflammatory process caused by another etiology rather than a true adnexal neoplasm[2,5,11].

Treatment of syringoma is cosmetic. There is no standard treatment for widespread syringomas. Most of the literature suggests using carbon dioxide laser[16,17]. Other treatment modalities include excision, electrodessication, curettage, dermabrasion, chemical peeling and oral/topical retinoids[18]. A recent report suggests the use of topical atropine to alleviate the pruritus in symptomatic eruptive syringoma. Other lasers such as argon laser, erbium-YAG laser, Q-switch alexandrite[5] have been tried with some success. Rarely, tumors may regress spontaneously in adulthood[18].

References:

1. Guitart J, Rosenbaum MM, Requena L. 'Eruptive syringoma': a misnomer for a reactive eccrine gland ductal proliferation?. J Cutan Pathol. 2003; 30: 202-5.
2. Seirafi HH, Akhyani M, Naraghi ZS, Mansoori P, Dehkordi HS, Taheri A, Ehsani AH, Kavusi S, Farnaghi F. Eruptive syringomas. Dermatol Online J. 2005; 11: 13.
3. Patrizi A, Neri I, Marzaduri S, Varotti E, Passarini B. Syringoma: a review of twenty-nine cases. Acta Derm Venereol. 1998; 78: 460-2.
4. Soler-Carrillo J, Estrach T, Mascaró JM. Eruptive syringoma: 27 new cases and review of the literature. J Eur Acad Dermatol Venereol. 2001; 15: 242-6.
5. Silva CS, Souza MB, Lima KA, de Almeida MA, Criado PR. [Generalized eruptive syringoma: case report]. Dermatol Online J. 2009; 15: 7.
6. Friedman SJ, Butler DF. Syringoma presenting as milia. J Am Acad Dermatol. 1987; 16: 310-4.
7. Goyal S, Martins CR. Multiple syringomas on the abdomen, thighs and groin. Cutis. 2000; 66: 259-262.
8. Ambrojo P. Requena Caballero L, Aguilar Martinez L,et al. Clear-cell syringoma: microscopy study. Dermatologica.1989; 178: 164-6.
9. Jacquet L, Darier J. Hiydradénomes éruptifs, épithéliomes adénoides des glandes sudoripares ou adénomes sudoripares. Ann Dermatol Syph. 1987; 8: 317-323.
10. Tsunemi Y, Ihn H, Saeki H, Tamaki K. Generalized eruptive syringoma. Pediatr Dermatol. 2005; 22(5): 492-3.
11. Draznin M. Hereditary syringomas: a case report. Dermatol Online J. 2004; 10: 19.
12. Belardi MG, Maglione MA, Vighi S, Di Paola GR. Syringoma of the vulva. A case report. J Reprod Med. 1994; 39: 957-9.
13. Lo JS, Dijkstra JW, Bergfeld WF. Syringomas on the penis. Int J Dermatol. 1990 ;29: 309-10.
14. Dupre A, Bonafe JL, Christol B. Syringomas as a causative factor for cicatricial alopecia. Arch Dermatol. 1981; 117: 315.
15. Yung CW, Soltani K, Bernstein JE, Lorincz AL. Unilateral linear nevoidal syringoma. J Am Acad Dermatol. 1981; 4: 412-6.
16. Metze D, Wigbels B, Hildebrand A. [Familial syringoma: a rare clinical variant] Hautarzt. 2001; 52: 1045-8.
17. Wang JI, Roenigk HH Jr. Treatment of multiple facial syringomas with the carbon dioxide (CO2) laser. Dermatol Surg. 1999; 25: 136-9.
18. Teixeira M, Ferreira M, Machado S, Alves R, Selores M. Eruptive syringomas. Dermatol Online J. 2005; 11: 34.