

CASE REPORT

GENERALIZED SYRINGOMA: BENIGN, YET A SERIOUS COSMETIC PROBLEM SUCHANA MARAHATTA^{1*}, PARICHA UPADHAYAY², RAMGYAN YADAV¹

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ABSTRACT

Syringoma is a benign appendageal neoplasm of the eccrine ducts. The word "Syringoma" has been derived from greek word "Syrinx", meaning pipe of tube. Friedman & Butler has classified it into four principal clinical variants. Among them, generalized syringoma is a rare variant comprising two sub-variants: multiple syringoma and eruptive syringoma. It presents with multiple crops of skin colored to tan colored, flat topped papules of 1-3 mm diameter, distributed symmetrically over face, trunk and extremities. It is a benign disease with great cosmetic concern.

Key words: Appendageal tumor, eccrine duct, generalized syringoma.

INTRODUCTION

Syringoma is a benign appendageal tumor arising from ductal part of the eccrine glands. It is characterized by asymptomatic, small, firm, flesh-colored to yellow brown, flat papules. Mostly, there are multiple lesions distributed bilaterally in front of the chest, face and neck.¹ Friedman & Butler has classified it into four principal clinical variants: i) localized form ii) familial form iii) associated with Down's syndrome iv) generalized form which comprises multiple and eruptive syringoma.² Although its most widely used treatment modality is ultrapulsed CO₂ laser, no studies have confirmed its efficacy. Also, recurrences are very frequent even after treatment.³ Hence, it remains a serious cosmetic problem even till date.

Case study

Here, we are going to present three cases of generalized syringoma.

Case 1:

A 25-year old female presented with asymptomatic crops of hyperpigmented flat topped papules with

1-3 mm diameter, started from forehead five years back and gradually progressed to involve whole face (Figure 1 & 2). Six months back, she also noticed few similar lesions over chest and bilateral arms.



Fig1: Multiple skin to tan colored flat topped papules on face



Figure 2: Multiple skin to tan colored flat topped papules on temporal region

Case 2: A 26-year-old male presented with mildly pruritic skin colored to hyperpigmented papules of 2-3 mm size, gradually increasing in number for last 12 years duration. They were distributed bilaterally symmetrically over face, neck, chest, upper back and abdomen (Figure 3 & 4).



Fig 3: Multiple skin to tan colored flat topped papules on face



Fig 4: Multiple skin to tan colored flat topped papules on chest & upper abdomen

Case 3:

A 25-year-old male presented with ten years history of skin to tan-colored, flat-topped papules of 1-3 mm in diameter, on the face, neck, axilla, chest and upper abdomen. They were bilateral, symmetrical; and had both follicular as well as non-follicular distribution



Figure 5: Multiple skin to tan colored flat topped papules on neck & chest



Figure 6: Multiple skin to tan colored flat topped papules on axilla & lateral trunk

The remaining physical examinations were unremarkable in all cases. In all the cases, there was no history of diabetes mellitus, hypertension, prolonged exposure to radiotherapy, intake of significant systemic medications, and use of over-the-counter topical preparations. There was no spontaneous resolution in the period of observation. No similar illness in the family members.

All three patients were subjected for punch biopsy from the representative lesion, which were consistent with syringoma. Biopsy specimen demonstrated solid epithelial islands with ductal differentiation. The ducts were lined by 2-3 layers of cuboidal epithelium and few of them even showed comma-shaped extensions. The stroma showed thick bundles of collagen. Based on the clinic-pathological co-relations, we made final diagnoses of generalized syringoma. After explaining the possible treatment outcome, we went for a trial of radiocautery and electrocautery of few exposed lesions; but all of them denied intervention of the remaining ones because of the scarring and recurrence.

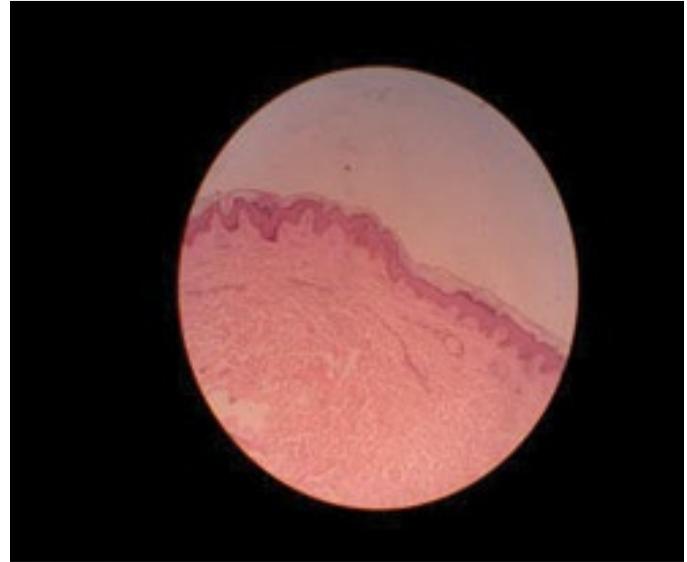


Figure 7: (10x zoom) Showing epithelial neoplasm involving upper reticular dermis



Figure 8: (40x zoom) Showing ductal structure lined by 2-3 layers of cuboidal cells

DISCUSSION

Syringoma is a benign adnexal tumor arising from eccrine ductal epithelium. Friedman & Butler, 1987 has classified it into four principal clinical variants: localized form, familial form, associated with Down's syndrome, and generalized form which comprises multiple and eruptive syringoma². Its classical presentation is characterized by multiple skin-colored to brown, flat topped papules, ranging from 1-3 mm in size, commonly present over eyelids

and upper cheeks¹. But in generalized eruptive syringoma, a rare clinical variant first described by Jacquet and Darier in 1987, the lesions comprised of small (usually <3 mm in diameter), skin-colored papules, present in the axillae, chest, abdomen, along with other usual sites. They usually appear in symmetrical clusters as seen in our cases.⁴

Although the pathophysiology of eruptive syringoma is not yet completely understood; some authors have proposed that it is a reactive process in hyperplastic eccrine duct rather than a true neoplasm, resulting from a previous cutaneous inflammation.⁵ Past reports had shown association of the eruptive syringoma with diabetes, Down's syndrome, during radiotherapy, milia, sarcoidosis, elevated serum carcinoembryonic antigen (CEA), and psychiatric disorders etc.^{1,6}

Histopathological study of the lesion shows epithelial neoplasm involving the upper reticular dermis comprising of cystic tubules. Those tubules are lined by 2 to 3 layers of basophilic granular cells and central eosinophilic cuticle with ductal differentiation. Its lumen contains amorphous eosinophilic debris. The most characteristic feature is the strand of epithelial cells projecting from a side of duct into the stroma; which resembles a "Tadpole's tail" or "Comma".^{1,7}

Treatment of syringoma is mainly for cosmetic purpose. Though, many treatment options like: surgical excision, electrocautery, cryotherapy, ablative carbon dioxide laser, dermabrasion, topical retinoic acid etc are available; none of these treatment modalities are found to have satisfactory outcome.⁴ Moreover, as they are located in the dermis, none of these modalities are found to be devoid of scar formation. Also, since recurrence is another common problem with syringoma, its treatment is quite frustrating.³ Hence, syringoma still remains a real cosmetic problem.

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