A Case Study and Review of Literature of Eruptive Syringoma in a 6-Year-Old

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BACKGROUND

Definition
- Syringoma is a benign, adnexal tumor of the eccrine sweat gland ducts.

History
- The word Syringoma is derived from Greek language where Syrinx refers to a tube. [1]
- Eruptive syringoma, a rare variant first described by Jacquet and Darier in 1887 [2]

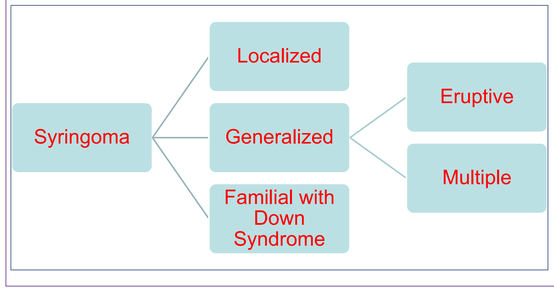
Age range
6 years to 70 years

Origin
- Intraepidermal portion of the eccrine sweat ducts

Common sites
- Periorbital area and neck

Associations
- Down Syndrome

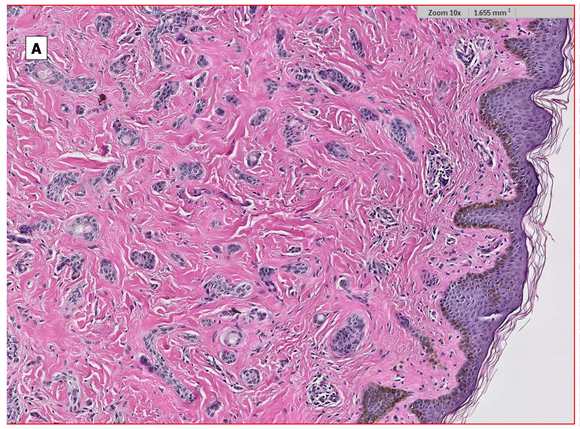
Proposed Variants of Eruptive Syringoma by Friedman & Butler [3]



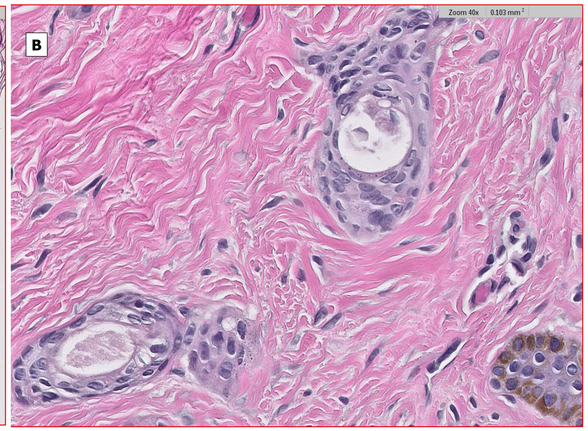
CASE DISCUSSION

Case Report

- A six year old African-American child presented with several lesions that begin under the chin and upper neck.
- In few months she developed extensive 2-3 mm hyperpigmented small linear papules over the neck, upper chest and axilla bilaterally.
- The lesions were non-painful and did not itch or bleed.
- There was also an incidental finding of Café au lait macule seen in the mid back of the patient.



Low power (10x) shows multiple small ducts which are showing tad-pole shaped or paisley-tie pattern with dense red sclerotic or fibrotic stroma.



High power (40x) shows small ducts with 2 layers of epithelium which have nests and strands of cells with basaloid appearance. The dilated glands with eosinophilic material.

EPIDEMIOLOGY

Salient features of Eruptive Syringoma

Location of tumor	Number of cases	Ethnicity	Number of cases	Sex	Number of cases
Face	9	Caucasian	7	Male	10
Neck	12	Asian	2	Female	15
Trunk	18	South East Asian Indian	4		
Abdomen	8	African /African-American	3		
Back	5	Middle Eastern	1		
Upper extremities	13	Not provided	5		
Lower extremities	8				
Groin	1				

TREATMENT

- Dermabrasion
- Various methods of excision
- Cryosurgery
- Electrodesiccation
- Chemical peeling
- Oral and topical retinoids
- Carbon dioxide laser
- Topical atropine

PATHOGENESIS

- The histogenesis of syringomas is most likely related to eccrine elements or pluripotential stem cells. [5]
- However, distinguishing between eccrine and apocrine ducts is sometimes difficult, and many tumors that were traditionally thought to be eccrine have been shown to have apocrine differentiation.
- The immunohistochemical pattern of cytokeratin expression indicates differentiation toward both the uppermost part of the dermal duct and the lower intraepidermal duct (ie, sweat duct ridge).
- Few authors believe, eruptive syringoma may represent a hyperplastic response of the eccrine duct to an inflammatory reaction rather than a true adnexal neoplasm.
- Some cases of syringoma are associated with diabetes mellitus.

LITERATURE REVIEW

Papules Characterization	Percentage
Hyperpigmented	96%
Erythematous	12%
Positive Darier Sign	4%
Down Syndrome-related	4%

Treatment Modalities	Percentage
Cryotherapy	8%
Oral Isotretinoin	16%
Topical Tretinoin	8%
Pulsed Dye Laser	4%

Clinical outcome	Percentage
Improvement	2
No Improvement	5

FOLLOW UP AND PROGNOSIS

- The patient was reassured that it was a benign lesion
- Treated conservatively due to the age of the patient.
- Follow up revealed that the lesions subsided without medical or surgical intervention over the years.

DIFFERENTIAL DIAGNOSIS

Conditions	Histological Findings
Acne vulgaris	Follicular dilatation with surrounding perifollicular infiltrate with neutrophils, lymphocytes and histiocytes.
Sebaceous hyperplasia	Increased clusters of normal sebaceous glands
Eruptive xanthoma	Epithelial/epidermal hyperplasia with keratinocyte necrosis, vacuolated or foamy macrophages
Darier's disease	Suprabasal clefts and acantholytic, dyskeratotic cells in spinous and granular layers
Fox-fordyce disease	Keratotic plugging of infundibulum and follicular spongiosis with perifollicular and periductal inflammation
Verruca plana	Exophytic papillomatous proliferation with hypergranulosis, intracorneal hemorrhage, keratohyaline inclusions
Lichen planus	Band-like inflammatory infiltrate obscures the dermoepidermal junction
Mastocytosis	Multifocal compact or diffuse compact mast cell infiltrates and diffuse interstitial infiltration pattern
Granuloma annulare	Interstitial histiocytes perivascular lymphocytes with central mucin with giant cells
Lichen nitidus	Acanthotic rete ridges forming claw-like epidermal grasp with dense subepidermal infiltrate
Pseudoxanthoma elasticum	Elastic fibers are fragmented, swollen, and clumped in middle and deep reticular dermis
Trichoepithelioma	Multiple lobules and nests of basaloid cells involving superficial dermis with keratinizing cysts
Sarcoidosis	Naked noncaseating granulomas with Schaumann bodies and Asteroid bodies

CONCLUSION

- Eruptive Syringomas are benign adnexal neoplasms that can mimic many inflammatory and malignant tumors.
- Biopsy is required to histologically differentiate with other skin lesions.
- Various options of treatment is available
- Associations with Down's syndrome and diabetes mellitus is important.

MATERIALS AND METHODS

- A Pubmed search performed using the search terms "eruptive syringoma" and "eruptive syringomas"
- A total number of about 73 studies found on Pubmed
- 24 cases out of the 73 studies were included in this study.

Inclusion criteria :
- All studies that describe a case or cases regarding Eruptive syringomas
- Studies that are published in English only

Exclusion criteria:
- Studies that describe aspects of clinical features, histology and treatment for their respective cases.
- All articles that were free and described all the above-mentioned aspects were excluded.

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