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Editorial

Trough and Conduit - Syringoma Nipple

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Bajai.

Syringoma nipple emerges as a benign neoplasm of cutaneous adnexa. Engendered from sweat glands and of eccrine origin, tumefaction is posited to arise from sweat duct ridge. Neoplasm is commonly discerned upon lower eyelids of implicated female subjects. The lesion may expound a malignant counterpart which is designated as syringomatous carcinoma or sweat gland carcinoma.

Frequently multiple, syringoma nipple configures firm papules of magnitude varying from one millimetre to 4 millimetres. Preponderantly confined to dermis, the well circumscribed lesion articulates ductules demonstrating a 'comma' pattern, layering by basaloid cells and encompassed within a stroma with significant sclerosis.

Syringoma expounds a female preponderance. However, the predominantly asymptomatic vulvar syringoma may remain undocumented.

Characteristically, neoplasm is frequently discerned within reproductive years or with commencement of puberty, possibly due to contributory hormonal factors.

Although preponderant in Asians, enhanced disease incidence is observed within Japanese women. Additionally, conditions such as Down's syndrome, Ehlers-Danlos syndrome, Marfan's syndrome, Nicoloau-Balus syndrome or Brook-Spiegler syndrome exhibit elevated disease incidence. An estimated 20% neoplasms depict a familial concordance [1,2].

Site of occurrence of syringoma is contingent to the clinical variant. Localized variant demonstrates lesions confined to periorbital region, vulva, penis, scalp, axillary region, face and forearm or hand in decreasing order of frequency.

Eruptive variant delineates lesions within anterior trunk, neck and axillary region in decreasing order of frequency.

Commonly, neoplasm is encountered within neck and anterior trunk of female subjects <15 years [1,2].

Of obscure aetiology, hormonal factors may influence emergence of syringoma nipple. Lesion may represent as an adenoma of acrosyringium which is an intra-epidermal segment of eccrine sweat duct. Nevertheless, pathogenesis is currently incompletely elucidated. Clear cell variant may expound concurrence with adult onset diabetes mellitus [2,3].

Multiple syringomas are associated with an autosomal dominant mode of disease inheritance. Neoplastic origin appears concordant to chromosome 16q22 [2,3].

Generally asymptomatic, syringoma nipple frequently represents with pruritus wherein pruritus may be exacerbated during menstruation and summer season. Ingestion of oral contraceptives or gestation may exacerbate cogent clinical symptoms [2,3].

Characteristically, lesions are symmetric and bilateral. Implicated subjects depict multiple, miniature, firm, flesh coloured nodules of magnitude varying from one millimetre to 4 millimetres. Concomitant extra-genital lesions are frequently encountered [2,3].

Syringoma nipple depicts distinct clinical variants as

- Localized
- Generalized
- Multifocal
- Eruptive
- Familial
- Lesions concurrent with Down's syndrome [3,4].

Cytological smears expound aggregates of basaloid cells. Alternatively, clear cell variant appears to be composed of clear cells. Tumour cells are devoid of cytological atypia. Mitotic figures are extremely exceptional [3,4].

Grossly, tumefaction appears as a firm, flesh coloured papule. Cutaneous eruption may or may not concur. Tumour nodules vary from one millimetre to 4 millimetres in diameter [3,4].

Upon microscopy, the well circumscribed neoplasm is generally confined to superficial reticular dermis wherein the tumefaction may exceptionally expand into subjacent dermis [3,4].

Neoplasm is comprised of proliferating dual cellular component denominated as

 Epithelial cells configuring ductules, cords and cellular nests along with articulation of cysts. Tumour cells are basaloid, cuboidal epithelial and delineate dual layering within ductules. Lesion depicts an eosinophilic layer of cuticle. Neoplastic ducts may configure a 'comma' or 'tadpole' or 'paisley' pattern.

Intra-cystic and luminal content is constituted of quantifiably variable proteinaceous debris or keratin.

 Sclerotic or fibrous tissue rich stroma which encompasses the lesion. Cytological atypia is absent. Mitotic figures are extremely exceptional [3,4].

The variant of clear cell syringoma is constituted of clear epithelial cells impregnated with glycogen. Aforesaid variant is commonly encountered in diabetics. Few miniature cysts may be coated with squamous epithelium.

Ultrastructural examination confirms eccrine genesis of the neoplasm [4,5].

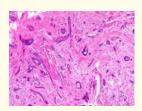


Figure 1: Syringoma demonstrating cords, nests and ductules layered by cuboidal epithelial cells. Surrounding stroma is sclerotic with significant fibrosis. Cytological atypia or mitotic figures appear absent [8].

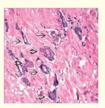


Figure 2: Syringoma delineating cords, nests and ductules layered by cuboidal epithelium. Encompassing stroma is sclerotic with significant fibrosis. Cytological atypia or mitotic figures appear absent [9].

Nottingham Bloom-Richardson grading system is contingent to

- Configuration of tubules by the tumour
- Quantifiable mitotic figures per 10 high power fields as exemplified within actively proliferating, cellular areas
- Occurrence of nuclear pleomorphism.

Configuration of tumour tubules contributes to pertinent scoring and is classified as

- 1 point: Tubules representing > 75% of tumefaction
- $\bullet \hspace{0.4cm} 2$ points: Tubules articulating 10% to 75% of tumefaction
- 3 points: Tubules manifesting < 10% of tumefaction.

Mitotic figures are appropriately evaluated upon tumour periphery and are aptly quantified within mitotically active areas. Estimation of mitotic figures confined within 10 high power fields (hpf) constituting a singular, non contiguous neoplastic area is optimal.

Nuclear pleomorphism is classified as

- 1 point: Neoplasms depicting minimal variation of nuclear magnitude and outline with configuration of miniature, regular, uniform neoplastic cells
- 2 points: Neoplasms delineating moderate variation in nuclear magnitude and outline
- 3 points: Neoplasms demonstrating significant variation in nuclear magnitude and outline.

Carcinoma breast is graded and scored as

- 3 5 points: Accumulated by well differentiated, grade I carcinoma breast
- 6 7 points: Accumulated by moderately differentiated, grade II carcinoma breast
- 8 9 points: Accumulated by poorly differentiated, grade III carcinoma breast [3,4].

Syringoma breast appears immune reactive to carcinoembryonic antigen (CEA), epithelial membrane antigen (EMA) or CK5.

Tumour cells appear immune non reactive to oestrogen receptors (ER), progesterone receptors (PR) or SOX10 [5,6].

Syringoma nipple requires segregation from neoplasms as microcystic adnexal carcinoma, desmoplastic trichoepithelioma or morpheaform basal cell carcinoma [5,6].

Syringomatous lesions may arise upon the face and appear clinically concordant with lesions confined to the nipple. Neoplasm may be appropriately discerned upon histological evaluation of surgical tissue samples. Therapeutic intervention is necessitated for cogent cosmetic outcomes [6,7].

Surgical extermination of the lesion is commonly adopted. Alternatively, cryotherapy or laser therapy may be beneficially employed. However, employment of carbon dioxide laser ablation may induce scarring or hyperpigmentation [6,7].

Superior cosmetic outcomes may be obtained with therapeutic manoeuvers as micro-insulated needle radiofrequency.

Topical atropine is beneficially employed in order to alleviate associated clinical symptoms as pruritus and decimate lesion magnitude.

Generally, lesion appears unresponsive to topical steroids [6,7].

Syringoma nipple emerges as a benign, non-progressive lesion. However, multiple lesions may be disfiguring and delineate inferior cosmetic outcomes.

Following precise therapy, complications as lesion reoccurrence, scarring and dis-pigmentation may ensue [6,7].

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- 8. Image 1 Courtesy: Biomed central.
- 9. Image 2 Courtesy: Basic medical key.