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Editorial

Adjunct and Codicil-Eccrine Syringofibroadenoma

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Eccrine syringofibroadenoma occurs as an exceptionally discerned, benign neoplasm arising from eccrine duct cells. Lesions appear as erythematous, solitary or multiple papules and nodules frequently confined to acral sites or extremities. The neoplasm appears concurrent with Schöpf-Schulz-Passarge syndrome and Clouston syndrome.

Eccrine syringofibroadenoma is additionally designated as eccrine syringofibroadenoma of Mascaro, eccrine syringofibroadenomatous hyperplasia, acrosyringeal adenomatosis, eccrine poromatosis, linear eccrine poroma, acrosyringeal nevus or acrosyringeal nevus of Weedon and Lewis or nevus syringoadenomatosus papilliferum.

Tumefaction is composed of proliferating and anastomosing cellular cords and ductal articulations encompassed within a fibro-vascular stroma. Attenuated, anastomosing cellular cords, strands and ductal structures composed of monomorphic, basaloid cuboidal epithelial cells appear to expand from basal epidermal layer into subjacent dermis and are encompassed by fibro-vascular stroma.

The exceptionally discerned neoplasm preponderantly emerges within 7th decade to 8th decade although no age of disease emergence is exempt. Lesions concurrent with Schöpf-Schulz-Passarge syndrome are frequently encountered within adolescent subjects. Neoplasms associated with Clouston syndrome are observed in young adults. A specific racial or gender predilection is absent [1,2].

Eccrine syrinofibroadenoma predominantly occurs upon acral sites, especially extremities. However, implication of non acral sites as eyelids, perianal region or scalp is documented [1,2].

Eccrine syringofibroadenoma is posited to emerge from eccrine duct cells as postulated by morphological and ultrastructural examination along with cogent immunohistochemistry. The reactive subtype is hypothesized to be engendered from tissue repair or remodelling process of eccrine ducts [2,3].

Of obscure aetiology, lesion may arise as a reactive process, hamartoma or as a tumefaction with neoplastic genesis. Concurrence with ectodermal dysplasia appears indicative of tumorigenesis or hamartoma. Exceptionally, malignant metamorphosis may ensue, a feature which is indicative of neoplastic transformation. Lesions associated with inflammatory dermatoses and benign or malignant epithelial neoplasms suggest induction of a reactive process [3,4].

Eccrine syringofibroadenoma represents as a gradually progressive, solitary lesion or multiple nodules of tan or flesh coloured hue.

Neoplasm manifests with distinct clinical subtypes as

- Solitary subtype is a common variant delineating lesions frequently confined to lower extremities
- Multiple subtype demonstrating hidrotic ectodermal dysplasia or Schöpf-Schulz-Passarge syndrome and Clouston syndrome. Commonly, lesions are confined to palms and soles of young subjects between 15 years to 25 years

- Multiple subtype where lesions are devoid of associated cutaneous features and are emerge upon palms and soles
- Nevoid subtype configured of exceptionally discerned, non familial variant exemplifying unilateral, linear papules and plaques of eccrine syringofibroadenoma
- Reactive subtype which appears concordant with conditions such as
- Inflammatory dermatoses or disease processes as erosive lichen planus, uncontrolled psoriasis, bullous pemphigoid, epidermolysis bullosa, primary cutaneous amyloidosis, thermal scar, venous stasis, chronic diabetic foot ulcer, neuropathy or peristomal cutis
- Infectious diseases or aetiologies as leprosy or human papilloma virus (HPV) infection
- Neoplastic conditions as nevus sebaceous, clear cell acanthoma, acantholytic dyskeratotic acanthoma, squamous cell carcinoma, basal cell carcinoma or Merkel cell carcinoma
- Associated syndromes as
- Schöpf-Schulz-Passarge syndrome which emerges as an autosomal recessive disorder demonstrating homozygous genetic mutation within WNT10A gene confined to chromosome 2q35. Aforesaid syndrome represents with specific clinical features as palmoplantar keratoderma, hypodontia, hypotrichosis, hyperhidrosis, hidrocystomas confined to eyelids, nail dystrophy and eccrine syringofibroadenoma
- Clouston syndrome which emerges as an autosomal dominant disorder occurring due to heterozygous chromosomal mutations confined to gap junction β 6 (GJB6) gene located at chromosome 13q12.11 which encodes gap junction protein β 6 or connexin 30. Associated clinical features emerge as palmoplantar keratoderma, alopecia, nail dystrophy and eccrine syringofibroadenoma [3,4].

Grossly, eccrine syringofibroadenoma manifests with verrucous, erythematous papules, nodules and plaques. Ulceration of superimposed cutaneous surfaces may ensue. Multiple nodules appear as symmetrical lesions or configure unilateral, nevoid lesions [4,5].

Upon microscopy, neoplasm represents with proliferation of attenuated cords and strands of anastomosing, reticulated epithelial cells. Frequently, a lattice may be articulated which extends from superimposed epidermis into subjacent dermis [4,5].

Tumour cells appear as monomorphic, basaloid cuboidal epithelial cells with magnitude beneath < adjacent keratinocytes. Cords of epithelial cells are impregnated with structures simulating eccrine ducts. Intervening stroma appears fibrotic and vascularized [4,5].

Upon ultrastructural examination, tumour cells are pervaded with tonofilaments, desmosomes and innumerable glycogen granules. Besides, a distinct basal lamina, globular keratohyaline granules encompassing ducts and cystic structures may be discerned. Lamellar granules appear absent. Cellular envelopes appear inadequately developed and cornified [4,5].

Figure 1: Eccrine syringofibroadenoma demonstrating cords, tubular and reticular articulations comprised of monomorphic, basaloid epithelial cells impregnated with eccrine ducts. Surrounding stroma is fibrotic and vascularized [8].

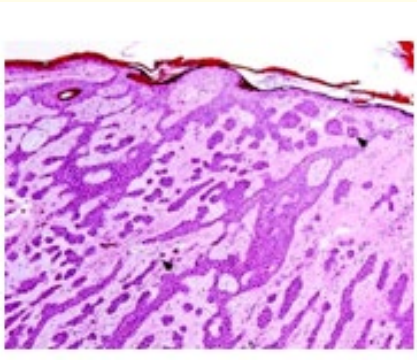

Figure 2: Eccrine syringofibroadenoma delineating cords, tubules and reticular structures comprised of monomorphic, basaloid epithelial cells pervaded with eccrine ducts. Circumscribing stroma is fibrotic and vascular [9].

Staging of cutaneous squamous cell carcinoma

Tumour staging is contingent to factors as magnitude of lesion, depth of neoplastic invasion, cellular differentiation and accompanying perineural invasion and is categorized as.

American Joint Committee on Cancer (AJCC) classification (seventh edition) applicable to cutaneous squamous cell carcinoma of the head and neck is comprised of

- T1: Tumour diameter < 2 centimetres and accompanying < 2 high risk factors
- T2: Tumour diameter ≥ 2 centimetres or lesion associated with ≥ 2 high risk factors
- T3: Tumour depicting invasion of orbit, maxillary, mandibular or temporal bone
- T4: Tumour delineating invasion of axial skeleton or appendicular skeleton or perineural invasion confined to base of skull.

Factors contributing to propensity of tumour emergence appear as tumour thickness > 2 millimetres, lesions with Clark level IV or V, poorly differentiated or undifferentiated tumefaction, lesions associated with perineural invasion or neoplasms confined to ear or lip.

American Joint Committee on Cancer (AJCC) classification (eighth edition) is comprised of

- T1: Tumour diameter ≤ 2 centimetres
- T2: Tumour diameter ≥ 2 centimetres and < 4 centimetres
- T3: Tumour diameter ≥ 4 centimetres or lesions displaying minimally a singular high risk feature
- T4a: Tumour demonstrating gross invasion of cortical bone or marrow of bones as orbit, maxillary, mandibular or temporal bone
- T4b: Tumour demonstrating invasion of base of skull or neural foramina confined to skull base.

Features contributing to predilection of tumour emergence appear as perineural invasion, especially within a nerve subjacent to the dermis or nerve ≥ 0.1 millimetre thickness or clinical or radiographic involvement of specific nerves in the absence of invasion or transgression of base of skull, deep seated tumour invasion > 6 millimetre thickness or beyond subcutaneous adipose tissue and minor bony erosion.

Brigham and Women's Hospital (BWH) classification is expounded as

- T1: Lesions with absence of high risk factors
- T2a: Lesions associated with singular high risk factor
- T2b: Lesions associated with two to three high risk factors
- T3: Lesions demonstrating ≥ 4 high risk factors or bone invasion.

Factors associated with enhanced propensity of tumour emergence appear as tumour diameter ≥ 2 centimetres, poorly differentiated neoplasms, tumours with perineural invasion within nerves ≥ 0.1 millimetre diameter or tumour invasion beyond subjacent adipose tissue [3,4].

Ductal or peri-ductal structures of eccrine syringofibroadenoma appear immune reactive to carcinoembryonic antigen (CEA), epithelial membrane antigen (EMA), involucrin, filaggrin, CK6, CK8, CK10, CK18 or CK19. Strands and sheets of tumour cells appear immune reactive to CK6, cytokeratin AE1, AE3 or CK34 β E12.

Strands and sheets of tumour cells appear immune non reactive to cytokeratin KL1 or CK10 [5,6].

Eccrine syringofibroadenoma requires segregation from neoplasms as fibroepithelioma of Pinkus, poroma and

hidroacanthoma simplex, porocarcinoma, tumours of follicular infundibulum or squamous cell carcinoma with focal ductal differentiation [5,6].

Eccrine syringofibroadenoma may be appropriately ascertained upon surgical tissue samples subjected to precise histopathological examination. Additionally, clinico-pathological concurrence appears mandatory for cogent tumour categorization [6,7].

Singular lesions may be appropriately alleviated by surgical extermination of the lesion. Alternatively, cryotherapy appears beneficial.

Contingent to location and magnitude, multiple lesions may be subjected to diverse treatment modalities as laser therapy achieved with dual pulse width flash lamp, carbon dioxide (CO2) laser therapy, radiotherapy or topical corticosteroids. Diffuse lesions may be managed with oral therapy with etretinate.

Benign or reactive lesions of eccrine syringofibroadenoma may undergo spontaneous involution. Lesions of extensive duration may exceptionally demonstrate malignant metamorphosis [6,7].

Bibliography

1. Chen Y and Tian X. "A Case of Reactive Eccrine Syringofibroadenoma Associated with Dyshidrotic Eczema". *Clinical, Cosmetic and Investigational Dermatology* 16 (2023): 3407-3411.

2. Khan HA., *et al*. "Late-onset eccrine syringofibroadenoma of the feet in a patient with hypohidrotic ectodermal dysplasia". *Australasian Journal of Dermatology* 62.3 (2021): 383-385.

3. Cunha Filho RRD., *et al*. "Reactive eccrine syringofibroadenoma triggered by thermal damage: case report". *Anais Brasileiros de Dermatologia* 96.2 (2021): 255-256.

4. Chukwuma O., *et al*. "Recalcitrant erosive plaques on the palms and soles: a rare manifestation of eccrine syringofibroadenoma". *JAAD Case Report* 6.7 (2020): 590-592.

5. Lee JS., *et al*. "Eccrine syringofibroadenoma associated with bowen's disease: a case report and review of the literature". *Annals of Dermatology* 32.1 (2020): 57-63.

6. Zhou P., *et al*. "Solitary eccrine syringofibroadenoma of the dorsum of hand". *Indian Journal of Dermatology* 65.6 (2020): 560-561.

7. Sugita Y., *et al*. "Reactive eccrine syringofibroadenoma on the heel, clinically mimicking squamous cell carcinoma". *Case Reports in Dermatological Medicine* 2019 (2019): 4735739.

8. Image 1 Courtesy: Basic medical key.

9. Image 2 Courtesy: Perri Dermatology.