Cannula, Conduit, Subway: Syringoma

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Preface

The term “Syringoma” is a derivative of the Greek word “Syrinx” which connotes a pipe or a tube. Syringoma is a benign adnexal tumour commonly affecting adolescents’ females, delineated in an estimated 1% population and configuring well differentiated ducts and cellular strands of basaloid epithelium. The tumour recapitulates the glandular epithelial structures. The neoplasm usually articulates as an intra-dermal or subcutaneous nodule. As syringoma frequently appears at puberty it can manifest a significant cosmetic impact [1,2].

Clinical Elucidation

Individuals remains asymptomatic or rarely demonstrate pruritus while perspiring. Syringoma usually exhibits miniscule, multiple, skin coloured, light brown or mildly pigmented papules. The peri-orbital or supra-clavicular region, neck, anterior or posterior trunk are incriminated sites, particularly in the eruptive form. Adjunctive sites such as axillae, abdomen, lower eyelids, cheeks and extremities can be implicated.

Four major clinical variants are categorized by Friedman and Butler as i) localized, ii) familial, iii) a generalized subtype which incorporates eruptive and multiple lesions and iv) associated with Down’s syndrome [2,3]. Syringoma is also prominently associated with Down’s syndrome (22.2%), diabetes mellitus (2.1%), Ehlers Danlos syndrome, Marfan’s syndrome and hyperthyroidism. Syringoma appearing in conjunction with milia, cysts and atrophoderma vermiculata are cogitated as Nicolau- Balus syndrome [2,3].

Histological Elucidation

Syringoma as a benign adnexal tumour comprises of minia- ture, dilated and cystic spaces lined by a dual layer of cuboidal epithelium interspersed with solid epithelial strands and columns of identical cuboidal cells and ductal articulations embedded in a dense, fibrotic stroma. A predominant fibro-connective tissue stroma envelops numerous miniature ducts lined with a dual layer of epithelial cells. The cellular epithelium appears flattened in a majority of the cases. The internal row of epithelium occasionally displays vacuoles. The duct Lumina frequently contain amorphous cellular debris. A few ducts and cystic spaces exhibit miniscule tail like, comma shaped elongations of epithelial cell configurations resembling tadpoles. A cluster of such articulations can produce a distinctive paisley like pattern. In addition to the ducts, solid strands of basophilic epithelial cells can be elucidated [1,3].

Cystically dilated ductal Lumina are impacted with keratin and often depict keratohyaline granules. The duct Lumina display a lining epithelium and generally about the superficial epidermis. The keratin cysts may rupture and incur a prominent foreign body giant cell reaction with subsequent though infrequent calcification. Exceptionally, several tumour cells appear as clear cells on account of glycogen accumulation. A few ductular structures and epithelial cords with preponderant cellular islands of irregular outline and magnitude can co-exist. Precluding the peripheral cell layer, the lining epithelium is preponderantly comprised of clear cells. The lesions are conventionally situated in upper to mid-dermis [3,4].

Enzyme Histochemistry

Discerns the syringoma as a tumour which differentiates towards intra epidermal eccrine sweat ducts and glands. Eccrine enzymes such as succinic dehydrogenase, phosphorylase and leucine aminopeptidase abundantly constitute the syringoma. The tumour cells display a weak reaction to lysosomal enzymes. Apocrine structures react to enzymes such as acid phosphatase and beta glucuronidase. Electron microscopic studies similarly cogitate the genesis of the tumour as being of eccrine sweat glands and ducts [4,5] (Figures 1-14).
Figure 1: Syringoma: Nests and Tubules of Cuboidal Cells, Keratinous Cysts and a Fibrous Tissue Stroma.

Figure 2: Vulval Syringoma with a Lining of Stratified Squamous Epithelium, Cords and Tubular Structures with Cuboidal Epithelium.

Figure 3: Eruptive Syringoma with Tadpole like Configurations and Glandular Lumina.

Figure 4: Syringoma with Glandular Lumen Lined with Cuboidal to Polygonal Epithelium, Tail like Structures and a Dense, Fibrous Stroma.

Figure 5: Syringoma: Solid Clusters of Cuboidal to Polygonal Epithelium with a Fibrotic Stroma.

Figure 6: Eruptive Syringoma with Dense Stroma, Solid Epithelium and Comma like Projections.

Figure 7: Syringoma with Epithelium Lined Cystic Spaces, Tail like Structures and Fibro-Adipoid Stroma.

Figure 8: Cutaneous Syringoma with a Stratified Squamous Epithelial Covering and Epithelium Coated Tadpole like Structures.

Figure 9: Cutaneous Syringoma with Glandular Spaces Lined by Cuboidal and Polygonal Epithelium and a Dense, Fibro-Collagenous Stroma.

Figure 10: Chondroid Syringoma with Epithelial Solid and Cystic Spaces and a Myxo-Chondroid Stroma.
Distinguishing Diagnosis

A clinical distinction of syringoma or a chondroid variant is mandated from lesions such as epidermal inclusion cyst, amelanotic nevus, sebaceous cyst, dermoid cyst, schwannoma, neurofibroma, pilomatrixoma and basal cell carcinoma [5,6]. An eruptive syringoma necessitates a demarcation from the clinically identical papular childhood dermatosis such as plane warts, acne vulgaris, lichen planus, granuloma annulare, papular sarcoidosis, milia, sebaceous hyperplasia, eruptive xanthoma, urticaria pigmentosa, Darier’s disease, pseudoaxanthoma elasticum and hidroicystoma.

Solid strands and columns of basophilic epithelial cells appear entrenched in a fibrous tissue stroma, a morphologically identical manifestation of a syringoma and a fibrosing basal cell epithelioma. However, fibrosing basal cell epithelioma is devoid of ductular configurations encompassing amorphous, eosinophilic material. Keratinous horn cysts abutting the epidermis delineated in syringoma often recapitulate the horn cysts of trichoepithelioma and can be misdiagnosed as the aforementioned tumours in a singular lesion. Although trichoepithelioma depicts solid columns and strands of basophilic epithelial cells and keratinous horn cysts, it frequently lacks ductal articulations [4,6]. Eruptive syringoma requires a histological distinction from conditions such as sclerosing (morphoea-like) basal cell carcinoma and desmoplastic trichoepithelioma. Syringoma also necessitates a distinction from tumours of identical histology such as a microcystic adnexal carcinoma. The tumour tends to infiltrate the deep dermis and subcutaneous tissue [7,8].

Therapeutic Options

Surgical excision, dermabrasion, cryotherapy, chemical peels particularly trichloro-acetic acid, topical atropine, carbon dioxide laser or pulsed dye laser can be beneficially adopted to treat syringomas. Post therapeutic adverse reactions such as scarring and dys-pigmentation can appear with employment of aforementioned options. Oral administration of isotretinoin in cumulative doses is advantageous and ensures a decline in the quantity and magnitude of syringomas.

Occasionally, administration of isotretinoin can be ineffective [8,9]. Several therapeutic strategies are available for managing eruptive syringoma or variants. However, the efficacy is restricted on account of the tumours being situated in the dermis with possible recurrences. Lesions can be eradicated by mild electrodesication or shaving. Carbon dioxide laser therapy with the pinhole technique and fractional thermolysis appears to be efficacious. Enlarged lesions necessitate a surgical extermination. Adjunctive modalities include cryosurgery, chemical peeling, dermabrasion and oral or topical retinoids. Appropriate for a syringoma or the chondroid variant is a comprehensive surgical excision with a prevention of future recurrences. Surgical excision of the tumefaction appears effortless on account of the circumscribing capsule. A recalcitrant, enucleated or an incompletely excised tumour lobule enhances the risk of tumour reappearance and malignant transformation. Thus, an annual post-operative monitoring is a pre-requisite. Infrequently, tumours reappear following a comprehensive eradication within one to two years [9-11].

Variants of Syringoma

Chondroid syringoma (CS) is an infrequent dermal tumour arising from the sweat glands, initially enunciated by Billroth in 1859. The neoplasm is a dual epithelial – mesenchymal lesion and
is cogitated as the “mixed tumour”. Chondroid differentiation is
cogent to the mesenchymal component of the tumour. The tumour
morphologically recapitulates mixed tumours of the salivary and
lacrimal glands. Hirsch and Helwig rephrased “myxoid” skin tu-
mour with the terminology of “chondroid syringoma” in 1961 on
account of the sweat gland elements articulating in a cartilaginous
stroma [1,2]. The nomenclature demarcates betwixt the mixed
tumours of salivary or lacrimal glands and the exceptional sweat
gland tumours. Majority of the mixed sweat gland tumours are be-
nign. As a mixed tumour, the neoplasm can be cogitated as a pleo-
morphic adenoma of skin. Reoccurrences, benign and malignant
conformities are described.

Head and neck region are frequently implicated, especially the
cheek, eyelid, forehead, nose, upper lip and infrequently, the orbit.
Chondroid syringoma also arises in the scalp, hand, foot, axilla, ab-
domen, penis, vulva and scrotum [3,4]. Chondroid syringoma has an
estimated prevalence of 0.01% to 0.1% of the primary cutane-
ous tumours and is frequently elucidated at an average age of 50
years with a minimal male preponderance. The majority (80%) of
the neoplasm are distributed in the head and neck preferentially
on the nose, upper lip and eyebrows. Chondroid syringoma ap-
ppears clinically as a gradually evolving, solitary, painless, well de-
defined, firm, papule or tumour aggregate varies from 0.5 centimetre
to 3.0 centimetre magnitude. The neoplasm is located in the der-
mis or sub-dermis. Superficial surface can be non-ulcerated, ery-
thematous, purple or skin coloured and the nodule is unattached
to the subjacent tissues [4,5]. On histology, a characteristic admix-
ture of epithelial and mesenchymal constituents is confirmatory of
the “mixed tumour” chondroid syringoma. Chondroid syringo-
ma demonstrates dual histological variants - apocrine and eccrine.

The apocrine subtype is frequent with randomly dispersed aggre-
gates of squamous epithelial cells, tubular or branching glandular
lumina and a double layered epithelial lining. The eccrine subtype
typically depicts homogenous epithelial elements with miniscule,
tubular and glandular lumina and a singular epithelial lining.

Focal immune reactivity to keratin, vimentin, desmin and S-100 protein is elucidated. Distinct, diagnostic histological crite-
ron for chondroid syringoma incorporate 1) clusters of cuboidal or
polygonal epithelial cells 2) inter-digitating tubule-aleolar
configurations with a dual or multi-layered cuboidal epithelial lin-
ing 3) ductal articulations lined by one or two columns of cuboidal
cells 4) occasional keratinous cysts 5) a matrix composed of mes-
enchymal constituents in variable quantities. Tubules, alveoli and
glandular articulations lined by two or more layers of cuboidal ep-
ithelium are exemplified. Islands of cuboidal or polygonal epithel-
ium embedded in a fibro-adipoid, chondroid, hyaline or mucinous
hypo-cellular stroma can be delineated [4,5].

A typical lesion of chondroid syringoma may display the entire
spectrum of histological modifications or manifest only a few fea-
tures. Electron microscopy, immune histochemistry and enzymat-
ics analysis determine an apocrine or eccrine genesis of the tumour.
Although benign, chondroid syringoma cogitates an exceptional
malignant subtype. Atypical or malignant tumours elucidate a de
novo emergence or may follow an inadequate resection. Malign-
ant transformation in a chondroid syringoma is exceptional and
is characterized by nuclear atypia, enhanced mitosis, infiltrative
tumour margins, satellite tumour nodules, necrosis and rapid tu-
mour progression. A neoplasm exceeding 3 centimeters in magni-
tude may possibly be malignant. Radiological assay of the tumour
can analyse attributes such as anatomical expanse of the tumefac-
tion, tumour invasion or adherence to adjacent anatomical struc-
tures. A chondroid syringoma necessitates a demarcation from
adjunctive benign tumours of epidermal or mesenchymal append-
age such as pleomorphic adenoma, dermoid cyst, neurofibroma,
lymphangioma, cavernous haemangioma, lipoma, sebaceous cyst,
lymphoma and histiocytic tumours [6,7].

Therapeutic strategies for managing chondroid syringoma in-
clude electrodessication and tumour vapourisation with car-
bon dioxide or Argon laser. However, a total surgical extermina-
tion remains the preferential therapy. Consistent monitoring for
excluding tumour recurrence and malignant transformation is a
pre-requisite. Chondroid syringoma may exemplify a salivary
gland tumour and comprises of varying quantities of mucoid and
cartilaginous stroma. Eruptive syringoma was first scripted by
Jacquet and Darier in 1887. Eruptive syringoma as an exceptional
tumefaction emerges as multitudinous, sequential lesions arising
at puberty or childhood. Eruptive or disseminated syringomas typ-
ically develops hundreds of miniscule, poor to well demarcated,
smooth surfaced, skin colored, pink, yellow or brownish papules
ranging from one centimeter to five centimeters in magnitude. The
lesions occur on the anterior chest, neck, eyelids, upper cheeks, up-
per abdomen, axilla, trunk, genitalia, extremities and peri-umbil-
ical area. Eruptive syringoma arises in concordance with Down’s
syndrome and Ehlers Danlos syndrome. Individuals with eruptive
syringomas infrequently demonstrate a family history of identical
lesions. Familial eruptive syringoma is an exceptional, autosomal
dominant condition [5,6]. Eruptive syringoma is a disorder of ob-
scure pathogenesis although a hyperplastic reaction of the eccrine
duct to an inflammatory stimulus such as contact dermatitis, shav-
ing, laser hair removal, alopecia areata or radiation dermatitis can
be implicated. Alternatively, eruptive syringomas can emerge due
to autoimmune damage to the acrosyringium and are cogitated
as an acrosyringitis with ductal cysts. Eruptive syringoma can be
cosmetically debilitating for individuals. A characteristic histolo-
gy is diagnostic of syringoma. The lesions depict a follicular and
non-follicular morphology [5,6].

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