The Squamoid Cavitation: Trichilemmal Cyst

Anubha Bajaj*

Department of Histopathology, Panjab University, A.B. Diagnostics, India *Corresponding Author: Anubha Bajaj, Department of Histopathology, Panjab University, A.B. Diagnostics, India. Received: December 06, 2018; Published: January 11, 2019

Introduction

The infrequent pathology of a trichilemmal cyst may configure as a conventional dermatosis. Trichilemmal cyst may additionally adopt a nomenclature of "Pilar" cyst. Initially scripted by Wilson Jones in 1966, the condition arises from the hair follicles [1]. A Trichilemmal cyst may articulate as a neoplasm or a cystic lesion. Frequently, the Trichilemmal cyst may be discerned in the scalp besides consistently appearing within hair bearing zones such as the neck, trunk and gluteal region [2]. Trichilemmal cyst may commonly demonstrate multitudinous lesions, particularly in the middle aged females [2,3]. Trichilemmal cyst may be a sporadic condition or the disorder may be transmitted as a familial, autosomal dominant trait [2,4]. The quantification and magnitude of Trichilemmal cysts may be variable.

Disease characteristics

Researchers McGavran and Binnington elucidated the detailed morphology of "Sebaceous" cysts on electron microscopy [2,3]. The categorical site of keratinisation within the cysts re-capitulates the keratinisation of hair cortex and nails with an absence of configuring kerato-hyaline granules. The keratinisation was pre-meditated from the pilar apparatus, particularly the external root sheath of hair. The cyst may thus be denominated as "Pilar" cyst. The hair follicle with the articulated follicular isthmus of the external root sheath may compose the exact origin of the Pilar cyst [3,4]. The lesions were subsequently denominated as Trichilemmal cyst. A majority (90%) of the cysts arise in the scalp, as a frequent, painless tumefaction. Trichilemmal cyst may be considered as the most frequent cutaneous cyst of the scalp and the second most frequent cystic lesion situated in the head and neck [4,5].

An epithelium lined, restricted cavity with constituent viscous fluid or adjunctive debris may define a Trichilemmal cyst. Cysts in relation to skin are classified on the basis of evolution and pathogenesis. Cystic cavities admixed with dermal appendages may be frequently encountered, in contrast to the developmental cysts which ensue on account of the persisting vestigial remnants of the hair follicle. Trichilemmal cyst may thus configure within the subcategory of an appendageal cyst [5,7].

An estimated 5% to 10% of the general population may delineate the emergence of a Trichilemmal cyst, commonly within the zone of intensely concentrated hair follicles. The predominantly benign cyst may infrequently depict a malignant transformation with distant metastasis.

Trichilemmal cyst, as an uncommon clinical detection, may emerge at any age. The condition is rare during adolescence and frequent in young women. Trichilemmal cyst, though frequent in the scalp, seldom display a collusion of several, mammoth pilar cysts within the scalp. Lesions on the scalp may be elucidated in individuals with outdoor occupation, abundance of sunlight and/or a poor hygiene. Trichilemmal cyst may be indolent, depicting a gradual evolution and dimensional amplification. Occasionally, the Trichilemmal cyst may be painful. The cyst may be commonly enunciated within the face, neck, trunk, scalp, scrotum, groin, ear flap, gluteal and/or mammary region [3,4]. The nodular lesion progresses at an estimated proportion of 0.5 centimetre a year and may evolve over a period of decades. The lesions may be intra-dermal with a superficial covering epidermis and a focal punctum.

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Additionally, the nodules may be asymptomatic with a smooth dermal envelope, an absence of ulceration and may contain viscous sebum The dimensions usually range from a few millimetres to a few centimetres. Nevertheless, lesions exceeding a five centimetre magnitude may be labelled as a Trichilemmal cyst [5,7]. Exceptionally, the Trichilemmal cyst may extend up to twenty five centimetres in magnitude. The solitary swelling may resemble a Pott's puffy tumour or an osteomyelitis of the skull bones.

Epidermal ulceration when co-existent, may extend from 1 centimetre to 4 centimetre and may be associated with a ruptured pilosebaceous follicle or an epidermal blister. Concordant mesenchymal lesions such as subcutaneous lipomas and/or dermal fibromas or osteomas may be associated with pre-malignant colonic polyps and innate pachyonychia and may manifest as constituents of the Gardner's syndrome [7,8]. Numerically amplified Trichilemmal or "Sebaceous" cysts may demonstrate the appearance of a concordant squamous cell carcinoma, basal cell carcinoma, mycosis fungoides or a malignant melanoma in the geriatric individuals and the lesions may display an extended duration [2,3]. Trichilemmal cyst usually requires a demarcation from focal osteomyelitis, mycosis fungoides and Pott's puffy tumour.

Diagnostic characterization

In approximately 2% instances singular or multitudinous foci of proliferating cells may induce the formation of proliferating tumours which may be designated as the "Proliferating Trichilemmal Cyst" or tumour. The proliferating trichilemmal cyst may also appear de novo and may progress quickly. The biologically benign lesion may enlarge, ulcerate or be locally aggressive [8,9]. Appropriate characteristics to distinguish the benign proliferating trichilemmal cyst from a malignant trichilemmal cyst may be clinically lacking. A comprehensive histological elucidation of the tumour may be a pre-requisite to suitably categorize the nodule.

The categorical determination of cystic lesions of the scalp may be achieved with the histological evaluation of an excised surgical specimen. A careful surgical excision of the Trichilemmal cyst of the scalp or adjunctive location may be mandated followed by a meticulous pathological assessment in order to exclude a malignant conversion [7,8].

Trichilemmal cyst as a solitary, intra-dermal or a subcutaneous lesion. may clinically simulate an epidermal cyst, though are usually less frequent than the epidermal cyst. The cyst may be comfortably enucleated and may often lack a visible punctum, in contrast to the epidermoid cyst.

Gross examination

The smooth walled, well circumscribed Trichilemmal cyst depicts an off white external surface. Further sectioning displays an oozing of cream to whitish, semisolid or viscous, cheese-like keratinous substance [2,3].



Figure 1: TC: stratified squamous epithelium with hyperkeratosis, lining a crater [11].



Figure 2: TC: corrugated squamous epithelium overlying homogenous keratin [12].



Figure 3: TC: squamous epithelium with absent granular layer and abundant homogenous keratin with focal calcification [13].



Figure 4: TC: cystic cavity filled with keratin and a squamous epithelial lining [14].



Figure 5: TC: epithelial crater with impacted, homogenous keratin [14].



Figure 6: TC: peripheral palisade of basal layer and a keratin impacted cavity [15].



Figure 7: TC: a squamous palisade with a cystic cavity [16].



Figure 8: TC: thin epithelium lining the cystic, flaky keratin [11].



Figure 9: TC: plump epithelium, absence of granular layer, peripheral palisade [14].



Figure 10: TC: acanthotic squamous lining with peripheral palisade [17].



Figure 11: TC: peripheral parallelism with keratin imposition [18].



Figure 12: TC: florid squamous epithelial hyperplasia with palisade and fibro-connective stroma [19].

Microscopic examination

The Trichilemmal cyst may be lined with a stratified squamous epithelium. The intracellular bridges betwixt the various squamous epithelial cells may be indistinct. The basal epithelial cells may depict a characteristic peripheral palisading. The individual squamous epithelial cells may be dimensionally magnified and distend towards the cystic lumen. Squamous cells abutting the cystic cavity may be swollen and packed with pale staining cytoplasm. The keratinisation of squamous cells may be abrupt and expeditious. The interposed granular cell layer may be absent. The cystic cavity may demonstrate abundant, homogenous, eosinophilic substance which may differ from the lamellar keratin flakes elucidated in an epidermal cyst. Cholesterol clefts may be frequent within the keratinous material. The cystic constituents may be calcified in an estimated one fourths (25%) instances. The cyst may rupture and evoke a prominent foreign body giant cell reaction [8-10].

Keratinisation of the cyst may be considered analogous to the keratinisation of outer root sheath of the hair follicle or the trichilemmal.

Adjunctive investigations

Immune-histochemistry: An immune-reactive Keratin K10 and K17 may be elucidated. A polarized microscopic examination may exhibit perpendicular fragments of tonofibrils within the lining squamous epithelial cells. The anucleate squamous epithelial cells may suddenly become devoid of cytoplasmic organelles [2,3].

Tumour evolution: The appendageal tumour may configure a neoplasm which differentiates towards adnexal structures of the skin. The

peri-anal region may not depict the Trichilemmal cyst.

Proliferating trichilemmal tumour may be articulated as a solid to cystic neoplasm which may recapitulate the differentiation of a hair follicle isthmus [5,7]. The Trichilemmal cyst and the Proliferating Trichilemmal cyst/tumour may delineate a trichilemmal kind of cellular keratinisation. The lesion may depict a cyst with an associated tumour like proliferation and may co-exist. A well circumscribed, solid to cystic neoplasm emerging within the dermis or subcutaneous tissue may be delineated. A proliferative Trichilemmal cyst may be an exceptional and benign neoplasm. As per the nomenclature from Wilson Jones in 1966, it may be engendered from the cutaneous annexes of the hair follicles [1]. A proliferating Trichilemmal cyst may appear within or adjacent to the scalp. A proliferative Trichilemmal cyst as a benign tumour may originate from the external epithelial sheaths of the hair follicle. On histology it may manifest a trichilemmal pattern of keratinisation. A concordant and abrupt transition from the nucleated squamous epithelial cells to the anucleate keratinized cells may be exhibited with an absence of a granular cell layer. The majority (90%) of instances may arise from the scalp, especially upon the exposed skin [9,10]. The aetiology and pathogenesis of proliferative Trichilemmal cyst may be obscure though the majority progress from the wall of a preceding follicular cyst with a secondary trauma or inflammation. A debatable co-infection with the human papilloma virus (HPV) may be suggested. A proliferating Trichilemmal cyst necessitates a demarcation from a squamous cell carcinoma.

Immune-histochemical assay may be beneficial for the extrapolation of malignant conversion. The mitotic count may be evaluated with the Ki67 index as an enhanced cellular proliferation rate may accompany the malignant transformation and alteration of accessed cellular glycoprotein. A CD34 mutation within the p53 suppressor gene locus may additionally concur with emergence and progression of the cystic lesions to overt squamous cell carcinoma [3-5]. A squamous cell proliferation with abundant eosinophilic cytoplasm, abrupt cellular keratinisation and dense, homogenous, keratin occupying cystic spaces may be delineated. Focal epidermoid keratinisation may configure keratin pearls. The surrounding fibro-connective tissue stroma may be devoid of infiltration with the tumour cells. The lesion may be indolent or appear aggressive with the emergence of malignant proliferating trichilemmal tumours. A differentiation from a squamous cell carcinoma may be a pre-requisite for the follicular neoplasm.

The histology may vary from a Trichilemmal cyst with minimal epithelial proliferation and a smooth epithelial cell lining to cysts delineating marked epithelial hyperplasia, hypertrophy and cellular multiplication with few cystic regions, simulating foci of squamous cell carcinoma. The lobular proliferation of squamous epithelial cells may depict predominant epithelial invaginations within the cystic lumen. Peripheral palisading of the small basaloid squamous epithelial cells may be delineated abutting the dense, fibrotic, focal cyst wall. The squamous cells may differentiate along the lines of enlarged keratinocytes with abundant eosinophilic cytoplasm. Abrupt keratinization with trichilemmal configuration may ensue [2,3]. The concentrates of eosinophilic keratin may elucidate zones of dystrophic calcification and formulations of cholesterol clefts. The squamous lining/tumour cells may be monomorphic with insignificant cytological atypia and exceptional mitosis. The squamous cells aggregates may expand into the adjacent connective tissue. However, cellular proliferation may primarily be directed within the cystic lining and cystic lumen. Remnants of a Trichilemmal cyst may be exemplified at one pole. The cyst may predominantly be lined with smooth, squamous epithelium and patchy trabeculation.

The proliferating Trichilemmal cyst may mandate a distinction from a Trichilemmal cyst, a proliferating epidermoid and/or an infundibular cyst and a trichilemmal carcinoma. A Trichilemmal cyst may be enunciated in conjunction with a proliferating trichilemmal tumour, nevertheless, a Trichilemmal cyst cannot be ascertained as a neoplasm. The multi-lobular architecture of a neoplasm may be lacking. The cysts of the skin require an appropriate and comprehensive investigation and classification in order to avoid a misdiagnosis and misrepresentation [3,4].

Therapeutic protocol

The definitive therapeutic modality for managing a Trichilemmal cyst of the scalp may be surgical excision with a perimeter of normal, uninvolved tissue. The markedly vascular scalp may haemorrhage extensively during excision, thus an adequate pre-operative assessment may be mandated. An appropriate eradication may reduce the possibility of a cyst recurrence. A Trichilemmal cyst may necessitate a meticulous elimination with an absence of a rent in the cyst wall and an intact capsule[4,5].

A proliferating Trichilemmal cyst may be managed with a surgical resection and a margin of healthy, uninvolved tissue. The aggressive tumours may depict aspects such as localized infiltration, tumour reoccurrence following surgical excision and distant metastasis. Adjunctive therapeutic options such as a Moh's micrographic surgery, neo-adjuvant chemotherapy and radiation therapy may be employed. Radio-therapy may be applicable for lesions with malignant degeneration [7,8].

Characteristics	Epidermal Cyst	Trichilemmal Cyst
Microscopic background	Clean	Blotchy keratin or dirty, greasy discharge
Cellularity and cellular ap- pearance	Enhanced. Singly dispersed nucleate and anucleate squames	Minimal. Syncytial clusters of cells, occasional keratin globules.
Oily or viscous debris	Scant to moderate	Appears in older cysts
Keratinous debris	Cellular influx exceeds the keratin. Non-refringent crystals, stains blue on Giemsa stain	Keratinous debris exceeds the squamous cell component. Keratin globules stains magenta on Giemsa stain
Cystic calcification	Absent	Amorphous deposits or calcospherites

Table 1: Differences between Epidermal and Trichilemmal Cyst [6].

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