Boffin Access
FOSTERING KNOWLEDGE

Open Access Journal | Volume: 3.2

Biomedical Research and Reviews

ISSN 2631-3944

The Adnexal Malformation-folliculosebaceous Cystic Hamartoma

Anubha Bajaj[1*]

[1]Histopathologist, AB Diagnostics, New Delhi, India

Abstract

Folliculosebaceous Cystic Hamartoma (FSCH) was initially scripted by Kimura et al in 1991 and is cogitated as an exceptional, non neoplastic, cutaneous adnexal malformation comprised of distended follicular and sebaceous glandular unitsoccurring in association with mesenchymal elements [1]. Nomenclature of folliculosebaceous cystic hamartoma emerges from cogent histological features comprised of follicular, cystic articulations appearing with adjacent, multiple sebaceous lobules accompanied with modifications of mesenchymal stroma. Folliculosebaceous cystic hamartoma requires a segregation from asymptomatic, flesh coloured nodules or papules, particularly when originating within the face and scalp [1].

Disease Characteristics

Folliculosebaceous cystic hamartoma is an infrequent, benign, cutaneous, nodular neoplasm essentially arising from the hair follicle. On account the multitudinous, mature, ectodermal articulations and an admixture of mesodermal elements, the neoplasm is cogitated as a hamartoma or can be contemplated asan extension of or akin to sebaceous trichofolliculoma. Folliculosebaceous cystic hamartoma demonstrates a female preponderance, emerges in adulthood and progresses gradually. Folliculosebaceous cystic hamartoma is commonly exhibited betwixt 4 years to 84 years and exemplifies a mean age of occurrence at 51 years while the neoplasm is infrequent in infants beneath < one year [2,3]. Folliculosebaceous cystic hamartoma can concur with neurofibromatosis and the tumefaction can be misinterpreted as neurofibroma. Giant folliculosebaceous cystic hamartoma can arise as a congenital lesion or appears in infants, enlarges within the pubertal phase and is frequently discerned upon the face or adjunctive areas [2,3].

Clinical Elucidation

Folliculosebaceous cystic hamartoma enunciates nodules which are commonly situated upon the head, scalp,earlobe, superior dorsal region, posterior thigh, genital region, nose whereas centric region of the faceis a frequent location. An estimated 30% lesions arise within the nasal and paranasal region. Uncommonly, lesions on the scrotum and nipple can be discerned [2,3]. Majority of lesions are configured as solitary, rubbery, firm or soft, flesh coloured, sessile or pedunculated, polyps, papules or exophytic nodules with a smooth exterior and an adherence to the superimposed cutaneous surface. The magnitude usually varies from 0.5 centimetre to 1.5 centimetre whereas the dimensions generally beneath < 3 centimetres. The superficial cutaneous surface demonstrates multiple nodules admixed with scattered, distended pores exuding a viscous, oily discharge. Clinical segregation from various, benign cutaneous tumours such as a sessile papilloma, intradermal nevi, sebaceous hyperplasia, lipoma, dermoid cyst and neurofibroma is mandated [2,3].

Histological Elucidation

On gross examination, a flesh coloured or pinkish white, pedunculated or sessile nodule of variable diameter is exemplified which elevates the superimposed epidermis. Cut surface demonstrates multiple cysts impacted with whitish yellow substance. The neoplasm can be appropriately categorized as a hamartoma upon a cogent histological examination. Diagnostic morphological features of folliculosebaceous cystic hamartoma are delineated as sebaceous lobules adherent to infundibular cystic articulations, circumscribed with bundles of collagen, an admixture of mature adipocytes and an augmentation of miniature venules with a superimposed hyperkeratotic, acanthotic, stratified squamous epithelial lining [4,5]. The dermal region depicts multiple, distended pilo-sebaceous units delineating an infundibular variety of keratinization along with enunciation of several, radiating sebaceous lobules. Aforesaid cystic follicular arrangements are enveloped by mesenchymal tissue. Enhancement of muscular element abutting the sebaceous lobules is exemplified. Secondary hair follicles are absent. Folliculo-cystic configurations are surrounded by dense aggregates and bundles of laminated collagen admixed with abundant adipose tissue and an accompaniment of vascular proliferation [4,5]. The neoplasm is predominantly lobular and

Article Information

DOI:	10.31021/brr.20203121
Article Type:	Review Article
Journal Type:	Open Access
Volume:	3 Issue: 2
Manuscript ID:	BRR-3-121
Publisher:	Boffin Access Limited
Received Date:	03 February 2020
Accepted Date:	19 February 2020
Published Date:	22 February 2020

***Corresponding author:**

Anubha Bajaj
Histopathologist
AB Diagnostics, New Delhi, India
E-mail: anubha.bajaj@gmail.com

Citation: Anubha B. The Adnexal Malformation-folliculosebaceous Cystic Hamartoma. Biomed Res Rev. 2020 Feb;3(2):121

Copyright: © 2020 Anubha B. This is an open-access article distributed under the terms of the Creative Commons Attribution 4.0 international License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

articulated, enlarged cystic structures are preponderantly layered by mature, stratified squamous epithelium, identical to the infundibular epithelium. Multiple, mature sebaceous lobules appear circumscribed. Rudimentary hair follicles can be discerned within the mid-centric dermis. Folliculo-sebaceous cystic structures encompassed within the fibro-vascular stroma demonstrate aggregates of adipose tissue cells and elements of neural tissue. Collagen bundles configured within the reticular dermis are preponderantly enhanced and appear amidst sebaceous follicles. Foci which recapitulate angio-fibromas can be observed. A moderately disseminated perifollicular lymphocytic infiltrate is delineated [4,5]. The well circumscribed, dermal lesion is comprised of numerous follicular articulations and sebaceous lobules accompanied by distended, cystic spaces which directly extend to the superficial epidermis. The epithelial articulations are circumscribed by a fibro-hyaline stroma in concurrence with a fibrous component demonstrating an admixture of adipocytes [4,5] (Figures 1-8).

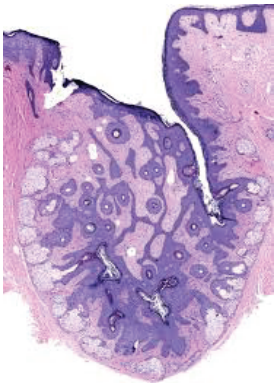

Figure 1: Folliculosebaceous cystic hamartoma demonstrating a lobular architecture with squamous epithelium lined cystic spaces and surrounding bundles of collagen.

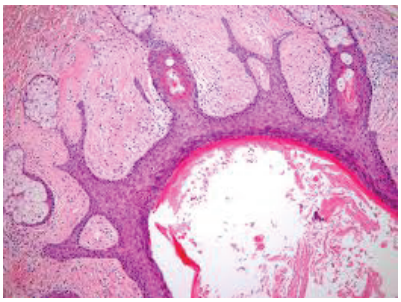

Figure 2: Folliculosebaceous cystic hamartoma with radiating cords and cystic spaces lined with stratified squamous epithelium and an enveloping stroma of fibro-vascular tissue and sebaceous lobules.

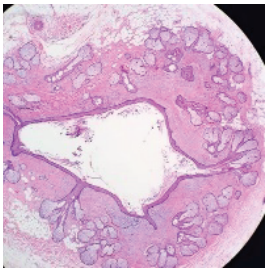

Figure 3: Folliculosebaceous cystic hamartoma depicting variously sized, epithelium lined cystic spaces, a circumscribing fibrous tissue stroma, a mild perivascular lymphocytic infiltrate and an absence of secondary hair follicles.

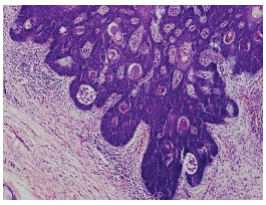

Figure 4: Folliculosebaceous cystic hamartoma delineating lobules of epithelium lined cystic spaces, encompassing fibrous tissue stroma, occasional sebaceous lobule and a mild, perivascular lymphocytic infiltrate.

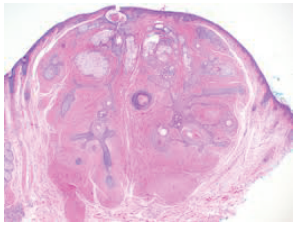

Figure 5: Folliculosebaceous cystic hamartoma with a lobular array of epithelium lined cystic architecture surrounded by bundles of collagen, scant inflammatory exudate and rudimentary hair follicles.

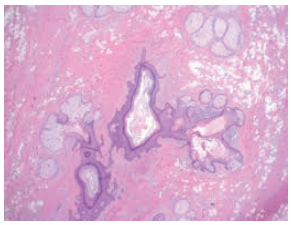

Figure 6: Folliculosebaceous cystic hamartoma with epithelium layered cystic spaces of varying magnitude, abutting sebaceous lobules and an admixture of collagenous tissue.

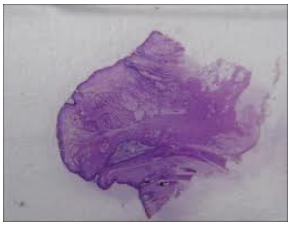

Figure 7: Folliculosebaceous cystic hamartoma demonstrating an admixture of epithelial and mesenchymal elements and a preponderantly lobular architecture.

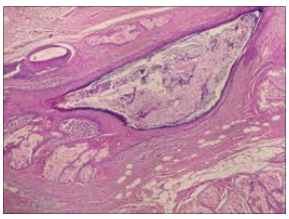

Figure 8: Folliculosebaceous cystic hamartoma exhibiting enlarged, cystic arrangements with a squamous epithelial lining, adjacent collagen bundles and sebaceous lobules with mild perivascular infiltrate and absent hair follicles.

Differential Diagnosis

Folliculosebaceous cystic hamartoma requires a demarcation from lesions such as sebaceous gland hyperplasia, steatocystoma, nevus lipomatosus superficialisor a cutaneous dermoid cyst, whereas the cystic hamartoma delineates an admixture of adipocytes and fibrous stroma. Various papulo-nodular or cyst like cutaneous lesions require segregation from folliculosebaceous cystic hamartoma [6,7]. Nevus sebaceous of Jadassohn demonstrates miniature, aberrant, budding sebaceous glands appearing adjacent to hyperplastic, stratified squamous epithelium with papillomatous articulations. Sebaceous trichofolliculoma resembles folliculosebaceous cystic hamartoma on morphological grounds although a distinction is possible. Multiple, refractile, vellus hair shafts are cogitated in sebaceous trichofolliculoma along with numerous sebaceous lobules in the absence of a centric, cyst-like cavity. Characteristically, mesenchymal stroma is absent in sebaceous trichofolliculoma although proliferation of nerve fibres along with adjunctive mesenchymal components can be enunciated [6,7]. Differentiation betwixt folliculosebaceous cystic hamartoma and sebaceous trichofolliculoma is challenging as the lesions primarily exemplify an infundibular cyst with encompassing sebaceous lobules. However, sebaceous trichofolliculoma depicts distinctive features such as excavated lesions on clinical examination and emergence of hair shafts within follicular structures. On the contrary, folliculosebaceous cystic hamartoma is represented as papules or nodules and is devoid of hair shafts within follicular structures [6,7]. Histological manifestations of folliculosebaceous cystic hamartoma emerge on account of secondary follicular regression encountered within sebaceous trichofolliculoma and aforesaid lesion is considered asa delayed phase of trichofolliculoma. Nevertheless, occurrence of congenital folliculosebaceous cystic hamartoma remains unexplained. Therefore, the lesions are contemplated to be composed of diverse histo-genesis. Additionally, exclusion of congenital cutaneous lesions is necessitated from folliculosebaceous cystic hamartoma [8,9].

Therapeutic Options

A comprehensive, surgical extermination of the neoplasm with a wide perimeter of uninvolved tissue is mandated. On further monitoring, individuals are asymptomatic and devoid of reoccurrence of the neoplasm [8,9].

References

1. Kimura T, Miyazawa H, Aoyagi T, Ackerman AB. Folliculosebaceous cystic hamartoma- a distinctive malformation of the skin. Am J Dermatopathol. 1991 Jun:13(3);213-220.
2. Noh S, Kwon JE, Lee KG, Roh MR. Folliculosebaceous cystic hamartoma in a patient with neurofibromatosis type I. Ann Dermatol. 2011 Oct;23(Suppl):S185-187.
3. Bobde VM, Helwatkar SB, Sathawane PR, Raut WK. Congenital giant folliculosebaceous cystic hamartoma of thigh: A rare case. Indian J Dermatol. 2019 Nov-Dec;64(6);490-492.
4. Bouhaja L, Jones M, Rammeh S. Folliculosebaceous cystic hamartoma. Our Dermatol Online. 2016:7(4):477-478.
5. Haw S, Lee MH. A case of giant folliculosebaceous cystic hamartoma. Ann Dermatol. 2009 Feb;21(1):63-65.
6. Merklen-Djafri C, Batard ML, Guillaume JC, Kleinclauss I, Cribier B. Folliculosebaceous cystic hamartoma: Anatomo-clinical study. Ann Dermatol Venereol. 2012 Jan;139(1):23-30.
7. Lee SY, Lee DR, You CE, Park MY, Son SJ. Folliculosebaceous cystic hamartoma on the nipple. Am J Dermatopathol. 2006 Jun;28(3):205-207.
8. Hamada M, Kiryu H, Satoh E, Moroi Y, Urabe K, et al. A case of genital folliculosebaceous cystic hamartoma with an unique aggregated manifestation. J Dermatol. 2006 Mar;33(3):191-195.
9. Brücher JJ, Franke I, Ulrich J, Gollnick H, Leverkus M. Giant genital variant of folliculosebaceous cystic hamartoma- successful management by a CO2 laser and acitretin therapy. Br J Dermatol. 2007 Oct;157(4);833-835.