Pilar and Eccrine-Microcystic adnexal carcinoma

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Disease Characteristics

Microcystic adnexal carcinoma is a neoplasm displaying dual differentiation constituted of eccrine and pilar glandular variants although within contemporary protocols of disease discernment the tumefaction is contemplated as an apocrine tumor. Microcystic adnexal carcinoma is engendered from pleuri-potent keratinocytes demonstrating an ability for adnexal differentiation. Factors enunciating a predilection for emergence of microcystic adnexal carcinoma are exposure to ultraviolet radiation, radiotherapy and immune suppression [1,2].

Clinical Elucidation

Microcystic adnexal carcinoma commonly appears as a gradually evolving, asymptomatic tumefaction upon the head and neck. Individuals demonstrate a whitish, non-pigmented, flattened, well circumscribed, scar-like lesion or a plaque, a few millimeters in magnitude which preponderantly appears upon the face. Generally, the neoplasm exemplifies as a flesh colored, indurated plaque or nodule, predominantly situated upon the upper lip or facial regions apart from the axilla, extremities, cutaneous surfaces of genitalia, trunk and scalp [2,3].

Histological Elucidation

The neoplasm is predominantly infiltrative and confined to the dermis. Extraneous surface of the tumefaction demonstrates numerous keratin impacted cysts accompanied by foci of calcification. Underlying reticular dermis depicts locally invasive nests and clusters of basaloid cells configuring slender strands and syringomatomoid cellular aggregates which often extend to the subcutaneous tissue. Suspicious or definitive foci of perineural invasion can be enunciated within the tumefaction [2,3]. Tumor infiltration can extend to the dermis, subcutaneous tissue or the underlying skeletal muscle tissue. Numerous keratinous cysts are configured upon superficial tumor segment. Solid islands, strands, nests and cords of basaloid and squamous epithelial cells are intermingled along with cystic configurations and an encompassing desmoplastic stroma. Tumor cell aggregates can delineate ductal differentiation along with foci of micro-calcification, clear cell metamorphoses, prominent glandular lumina and arborizing tubular structures [3,4]. Cords and strands of basaloid epithelial cells and ductular articulations amplify within the mid-dermis although keratinous cysts are quantifiably diminished. Focal sebaceous and follicular differentiation is enunciated. Deep-seated tumor component demonstrates a scirrhous configuration along with articulation of miniature nests, cords and strands of basaloid cells disseminated within a dense, hyalinized stroma. Epithelial elements are preponderantly reduced and display miniature cellular clusters comprised of two to three cells [4,5]. On cytological assessment, tumor cells are uniform and mitotic figures are exceptional. Glandular component can be prominent and engendered tumefaction is therefore denominated as sclerosing sweat duct carcinoma or malignant syringoma [4,5]. Tumor cells demonstrate an immune reactivity to epithelial membrane antigen [EMA], cytokeratin 7 [CK7] and cytokeratin 15 [CK15]. Luminal cells are immune reactive to carcino-embryonic antigen [CEA]. Few tumor cells are immune reactive to S100 protein and the circumscribing stroma is immune non-reactive to CD34 [4,5] (Figure 1-12).
Figure 1: Microcystic adnexal carcinoma demonstrating solid cords and strands of basaloid epithelial cell intermingled in a desmoplastic stroma [6].

Figure 2: Microcystic adnexal carcinoma with superimposed keratinous cysts with underlying solid strands and cords of basaloid epithelial cells surrounded by a desmoplastic stroma along with intermingled miniature epithelial cell clusters [6].

Figure 3: Microcystic adnexal carcinoma demonstrating keratinous cysts, cords, nests and aggregates of basaloid epithelial cells with an encompassing fibrous tissue stroma [7].

Figure 4: Microcystic adnexal carcinoma delineating aggregates and clusters of basaloid epithelial cells with intermingled, abundant fibrous tissue stroma [8].

Figure 5: Microcystic adnexal carcinoma displaying nests, cords, strands and miniature clusters of basaloid epithelial cells with a circumscribing desmoplastic stroma [9].

Figure 6: Microcystic adnexal carcinoma exhibiting superficial keratinous cysts, attenuated epithelium, cords, nests and tiny aggregates of basaloid epithelial cells with enveloping fibrous tissue stroma [10].

Figure 7: Microcystic adnexal carcinoma with elongated cords, aggregates and minimized cell clusters of basaloid epithelial cells loosely disseminated amidst abundant fibrous tissue stroma [11].

Figure 8: Microcystic adnexal carcinoma with slender cords, strands and miniature clusters of basaloid epithelial cells surrounded by a desmoplastic stroma [12].

Figure 9: Microcystic adnexal carcinoma demonstrating foci of perineural invasion along with cords and strands of basaloid epithelial cells [12].
Differential Diagnosis

Microcystic adnexal carcinoma requires a segregation from conditions such as morphea-like or fibrosing basal cell carcinoma and desmoplastic trichoepithelioma. Additionally, eccrine syringoid carcinoma as an exceptional cutaneous malignant adnexal neoplasm requires a demarcation [5,15]. Microcystic adnexal carcinoma necessitates a distinction from lesions such cutaneous metastasis of carcinoma breast, syringoma and papillary eccrine adenoma [5,15]. Differentiation is mandated betwixt microcystic adnexal carcinoma and fibrosing basal cell carcinoma with follicular differentiation. Morphea-like basal cell carcinoma lacks the configuration of glandular lumen or specific tumor zones. Immune reactivity to Ber Ep4, carcino embryonic antigen [CEA] within tumor lumina and intense staining with cytokeratin 15(CK15) is enunciated. Although lacking complete specificity, aforesaid immune reactive pattern is indicative of microcystic adnexal carcinoma, in contrast to a fibrosing basal cell carcinoma. Exemplification of Ber Ep4 is cogitated in an estimated (38%) of microcystic adnexal carcinomas and in entirety [100%] with basal cell carcinoma. Enunciation of cytokeratin 15(CK15) is around 92% with microcystic adnexal carcinoma whereas basal cell carcinoma is immune non-reactive [5,6]. Segregation of microcystic adnexal carcinoma is mandated from desmoplastic trichoepithelioma wherein the stroma is immune reactive to CD34. Discernible values of Ki-67 are reduced, thus indicating a minimal proliferative index. Desmoplastic trichoepithelioma and syringoma are devoid of an aggressive pattern of tumor evolution or perineural tumor dissemination. In contrast, microcystic adnexal carcinoma morphologically displays foci of calcification, epitheloid cell granulomas, adherence to follicles and adnexal structures, enunciation of keratin, immune reactivity to pleckstrin homology –like domain family A member 1 (PHLDA1) and immune non reactivity to formyl K77, thereby indicating a follicular histogenesis [15,16].

Investigative Assay

Dermatoscopic evaluation of microcystic adnexal carcinoma exhibits a dense, white, amorphous, centrally dispersed structureless zone accompanied by finely disseminated, linear, branching, centriodal vascular elements. Upon superior and peripheral tumor segments distinctive white clods of variable magnitude can be delineated. White blobs with aforementioned pattern are also observed within frequent adnexal cutaneous neoplasms such as trichoepithelioma and are probably indicative of keratin retention cysts. Cogent dermatoscopic evaluation is a contemporary investigative methodology for discerning microcystic adnexal carcinoma. Assessment of pertinent tumor tissue followed by an extensive histopathological examination is mandated to achieve an appropriate diagnosis. Tissue sampling as obtained with shave or superficial punch biopsies are inadequate for tumor evaluation [15,16].

Therapeutic Options

Comprehensive surgical extermination is an optimal mode of treatment for managing a microcystic adnexal carcinoma. Currently, adoption of Moh’s micrographic surgery is a preferential therapeutic modality. Localized tumor reoccurrence is enunciated in an estimated 50% instances. Tumor reappearance is minimized in instances where perimeter of surgical extermination is devoid of tumor cell aggregates, particularly as discerned with initially obtained tissue samples [16,17].

References

6. Image 1 and 2 Courtesy: Pathology Outlines
7. Image 3 Courtesy: MD Edge
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