



ORIGINAL RESEARCH PAPER

Dermatology

MULTIPLE FAMILIAL TRICHOEPITHELIOMA WITH BASAL CELL CARCINOMA- AN UNDER-REPORTED ENTITY

KEY WORDS: Multiple familial trichoepithelioma, nodules, Basal cell carcinoma

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ABSTRACT

Trichoepithelioma is a benign adnexal neoplasm, originating from hair follicles. It can present as a solitary type, or rarely as a multiple familial form. An autosomal dominant condition called multiple familial trichoepithelioma is characterized with large number of nodules and papules, mostly on the face and sometimes on the scalp. These can be associated with multiple cylindromas as well as trichoepitheliomas. Trichoepitheliomas can undergo conversion to malignant skin tumors such as basal cell carcinoma. [1]

SUMMARY

Trichoepithelioma is a benign neoplasm arising from hair follicles, differentiating towards both follicular germinative and stromal cells.^[2] Rarely, lesions of multiple trichoepitheliomas can undergo malignant conversion to basal cell carcinoma, which can manifest as ulcerations over the nodular lesions of trichoepitheliomas.^[3] It is important to evaluate the lesion to rule out the possibility of malignancy and manage the case accordingly.

Case Report

A 65-year-old man arrived with multiple raised lesions over his nose, that had been progressively growing for 40 years. He noticed the lesions at the age of 25 years, which over time grew larger and more numerous. Despite that, over the years he did not seek treatment regarding his asymptomatic lesions. The patient's children also had similar lesions. Patient gave history of raw lesions over the raised lesions on the left ala of nose for the last one year. History of bleeding on touch was present. On examination, multiple skin colored papulo-nodules, round, discrete to confluent lesions were seen over the nasal bridge, dorsum and ala of nose and over the nasolabial folds. Lesions were soft to firm in consistency. Over the left alar aspect of nose, a solitary ill- defined ulcer of 2 cm x 1.5 cm over the nodular lesions was seen, with pale granulation and peripheral hyperpigmented, rolled-out/beaded edge. Crusting over the ulcer floor was present. (Figure 1) Histopathology evaluation showed nodules of basaloid cells with a peripheral palisade arrangement, cells containing elongated nucleus, large cystic spaces were seen; stroma was arranged in parallel bundles around tumor. This was suggestive of basal cell carcinoma. Patient was referred to plastic surgery for further management of the case.

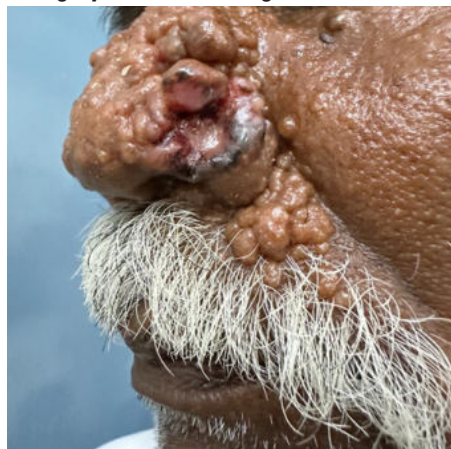


Figure 1- Clinical image showing Multiple Trichoepitheliomas, with nodulo-ulcerative lesion over the

ala of nose suggestive of basal cell carcinoma

DISCUSSION

Trichoepitheliomas are rare forms of genodermatosis that arise from germinative cells of the follicular-apocrine-sebaceous apocrine unit that are not differentiated. Typically, two clinical variants are described: giant solitary trichoepithelioma, which typically manifests in childhood and lacks a family history, and multiple trichoepitheliomas, which primarily affects the face and begins to show throughout puberty. Multiple familial trichoepitheliomas have been linked to genetic mutations, such as those in the CYLD gene. It is currently believed that the unusual occurrence of trichoepithelioma transforming into basal cell carcinoma typically occurs when there are several trichoepitheliomas. Theories about our case include the possibility that a single lesion of trichoepithelioma may evolve into a basal cell carcinoma or that two separate entities-basal cell carcinoma and trichoepithelioma—could overlap.^[2]

CONCLUSION

In this clinical image, we show a case of Multiple familial trichoepithelioma undergoing transformation to basal cell carcinoma, which is a rare entity. It is imperative that the lesions are evaluated for this malignant conversion to control the local spread of the basal cell carcinoma.

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