

# Solid and Cystic Clear Cell Hidradenoma with Focal Intracystic Carcinoma in-Situ

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## Citation

B Wang, D Sarma, B Javadzadeh, J Shehan. *Solid and Cystic Clear Cell Hidradenoma with Focal Intracystic Carcinoma in-Situ*. The Internet Journal of Dermatology. 2007 Volume 6 Number 2.

## Abstract

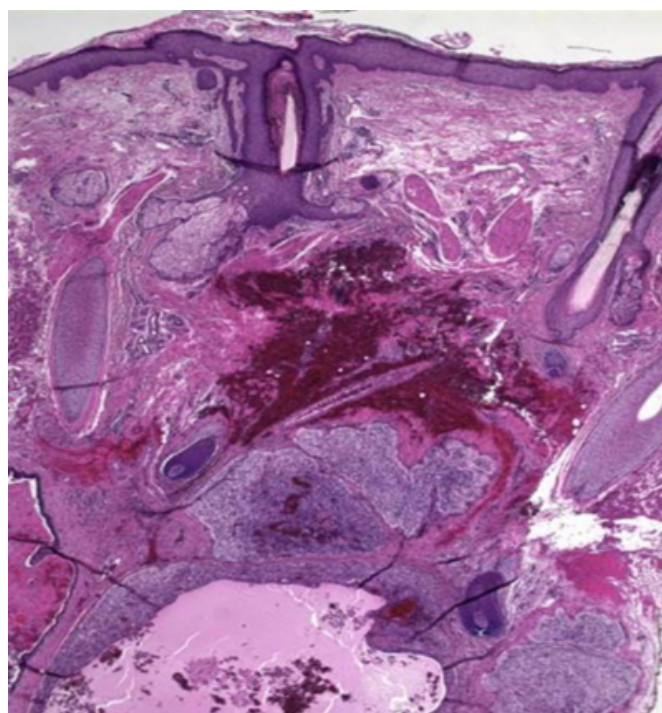
A rare case of a nodulocystic hidradenoma with intracystic carcinoma in-situ occurring in the scalp of a 79-year-old man is reported.

## CASE REPORT

A 79-year-old male presents an asymptomatic cystic nodule on his scalp. The excisional biopsy specimen shows a demarcated dermal neoplasm with both solid and cystic components. The solid area demonstrates sheets and nests of clear polygonal epithelial cells with small uniform nuclei. The nuclear chromatin is dark and condensed without discernable nucleoli. The similar clear cells are present at peripheral areas of cystic component. The large cystic areas show intracystic hemorrhage, proteinaceous material, and necrotic cellular debris. Focally, the cyst lining shows dysplastic squamoid cells projecting in the lumen in a papillary configuration. The dysplastic cells exhibit nuclear pleomorphism, large eosinophilic nucleoli, and mitotic figures. The coagulative necrosis is also noted in the intracystic papillary structures. There is no overt stromal invasion. The neoplasm has high proliferative index with 15% Ki67 positive cells as well as approximately 5% P53 positive cells. The neoplasm is immunoreactive to EMA, but non-reactive to P63, GCDPF-15, androgen receptor and HMB-45. We interpret the tumor as a cystic clear cell hidradenoma associated with focal intracystic carcinoma in-situ.

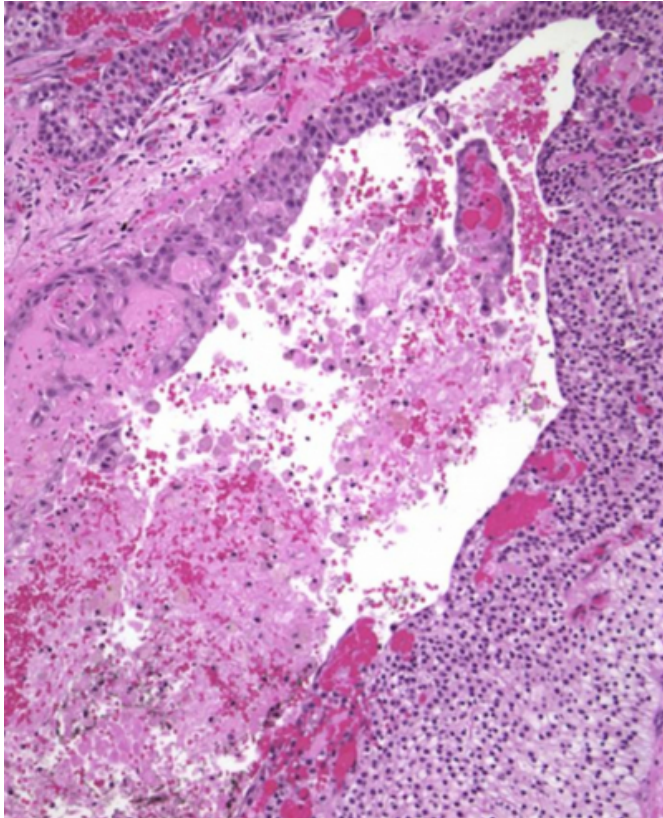
## Figure 1

Figure 1: Excisional biopsy of scalp nodule shows well demarcated solid and cystic tumor in the dermis.



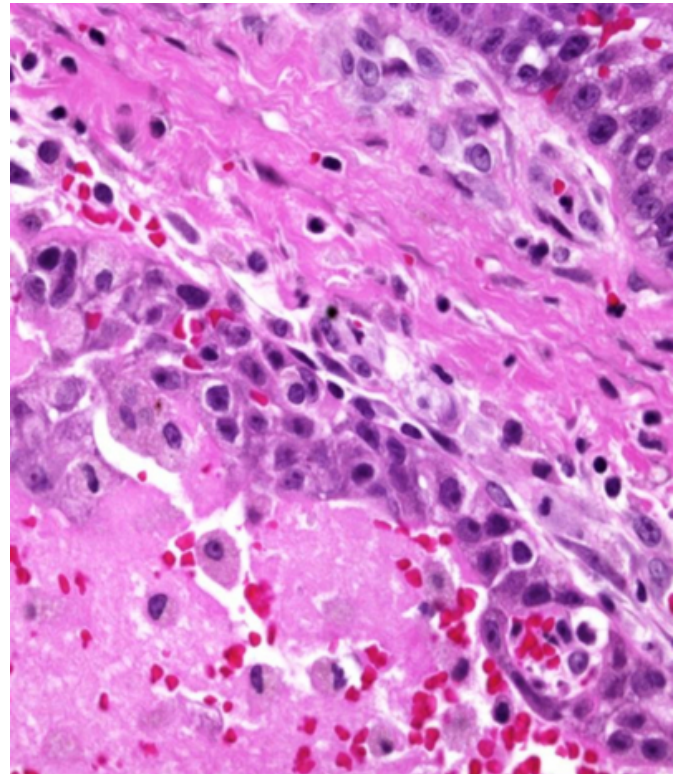
**Figure 2**

Figure 2: The cystic tumor is composed of small, uniform clear cells with dark, uniform nuclei (right) merging into dysplastic neoplastic cells over the upper and left part of the cyst. Cyst lumen contains necrotic cells (left).



**Figure 3**

Figure 3: High magnification: Intracystic carcinoma in-situ exhibits large squamoid cells with nuclear pleomorphism, prominent nucleoli and necrosis.



## DISCUSSION

Clear cell hidradenoma of the skin is a benign skin appendage neoplasm, which presumably arises from the folliculo-sebaceous-apocrine unit or eccrine glands. It may show apocrine, trichogenic, myoepithelial, or eccrine differentiation. Clear cell hidradenoma usually presents as a firm, asymptomatic skin nodule on a wide age range with slightly female preponderance. It can present in many locations, but is commonly seen in head and face (<sup>1</sup>). It has recently been demonstrated that chromosomal translocation t(11; 19) of clear cell hidradenoma of the skin is associated with TORC1-MAML2 gene fusion (<sup>2</sup>). Malignant clear cell hidradenomas or clear cell hidradenocarcinomas are rarely encountered in practice and have been reported as rare case reports (<sup>3,4,5,6</sup>). The clinical signs of a malignant adnexal tumor include rapid growth and ulceration of overlying skin with histopathologic evidence of local invasion as well as angiolymphatic or perineural invasion. The presence of nuclear pleomorphism, increased mitotic counts, or apoptosis is helpful, but not absolutely diagnostic for clear cell hidradenocarcinomas (<sup>4</sup>). Whether the malignant counterpart of clear cell hidradenoma occurs de novo or arises from malignant degeneration of existing hidradenoma

is not known. However, the new evidence of TORC1-MAML2 gene fusion in clear cell hidradenomas is interesting. Its fusion gene product, identical to that of mucoepidermoid carcinoma of the salivary gland, leads to the aberrant activation of downstream cAMP/CREB signaling genes, which results in deregulation of cAMP/CREB and Notch pathways and is associated with epithelial tumorigenesis (2). DNA aneuploidy has been reported in a recurrent clear cell hidradenoma (3). The molecular and cytogenetic evidence may suggest that clear cell hidradenocarcinoma may evolve from existing clear cell hidradenoma through multiple genetic mishaps.

Our case of hidradenoma exhibits overt dysplasia in the luminal epithelial cells, which represent, in our opinion, a nascent carcinoma in the clear cell hidradenoma. A complete excision of the clinical tumor is warranted to prevent recurrence and progression into invasive carcinoma.

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#### **References**

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