Pilomatrixical Carcinoma in a 70-year old Hispanic Female

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CASE PRESENTATION

Our patient is a 70-year-old Hispanic Female former-smoker with an exophytic tumor on the right nasal ala. No other significant dermatologic history was reported. The patient reports a 3-year history of the lesion with rapid growth in the past 7 months. She denies pain and pruritus of the area. She admits to manipulating the lesions with her fingers, as she reports “trying to pop” the lesion. On physical exam, a 1.1 cm pedunculated friable tumor with crust is noted on the right nasal ala (Figure 1). On initial visit, a shave biopsy was obtained and histology demonstrated zones of basaloid cell that predominate over areas of necrosis (Figure 2). Nuclear pleomorphism is noted on higher power (Figure 3). Immunostaining was positive for CEA, CK7, CK-AE1/AE3, chromogranin, CK20 and synaptophysin. After discussion of treatment options, the patient elected to have the lesion treated with wide excision.

BACKGROUND

Pilomatrixical carcinoma is a rare cutaneous malignancy of follicular matrix origin that was first described in the English literature by Lopransri and Mihm in 1980.[1] Since that first description over 130 cases have been reported.[2] Pilomatrixical carcinoma is associated with mutations in the CTNNB1 gene responsible for encoding β-catenin, a protein implicated in cell differentiation and proliferation.[3] It demonstrates locally aggressive behavior with high recurrence rates.[2] Histological features include predominant hyperchromatic basaloid cells with high mitotic rate and nuclear pleomorphism. Anucleate matrical corneocytes or “ghost cells”, central necrosis and occasional dystrophic calcification can also be present. Lesions are most prevalent on the head and neck and predominate in a geriatric population and among male patients.[4] Treatment of the lesion is wide excision with negative margins. Radiation treatment can also be considered in the case of recurrence.[5]

DISCUSSION

The most described treatment for Pilomatrixical Carcinoma is wide excision. Wide excision of the malignancy has demonstrated lower rates of recurrence. In the Herrmann et al. review, tumors removed with simple excision recurred at a rate of 83% while recurrence in wide excision was only 23.8%. The data is less clear on whether wide excision is effective in preventing metastasis. A review by Melancon et al. showed reduced rates of metastasis in pilomatrixical carcinomas treated with wide excision (10.3%) verses simple excision (20.5%), however, these differences (with p=11) did not meet the criteria for statistical significance.[6] Wide excision of the tumor is effective in preventing recurrence, however rates of metastasis may not be improved.

Other treatment modalities for pilomatrixical carcinoma mentioned in the literature are Mohs surgery, radiation therapy, both alone and as an adjuvant, and chemotherapy. Radiation has shown mixed results, while chemotherapy has not been proven effective.[6] Mohs surgery shows promise as a treatment modality due to the ease of identifying pilomatrixical carcinoma with hematoxylin-eosin stain.[7] However, due to the limited reports in literature of treatment with Mohs surgery, further research is needed.

REFERENCES