Well-Differentiated Squamous Cell Carcinoma Arising within a Proliferating Pilar Tumor

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INTRODUCTION

Proliferating pilar tumors (PPTs) are rare cutaneous neoplasms that arise from the outer root sheath of hair follicles. While most cases are benign and have a favorable prognosis, a small subset of PPTs are termed malignant based on their histologic features and potential for metastasis. Malignant PPTs may arise de novo or develop within an existing trichilemmal cyst.¹ These are rare entities with less than 100 documented cases in the literature; thus, malignant PPTs represent less than 0.1% of all skin cancers.¹,² In addition to being rare, malignant PPTs can be particularly challenging to diagnose and are often confused histologically with metastatic or invasive squamous cell carcinoma (SCC).³,⁴ We present here a case of well-differentiated, invasive SCC arising within a PPT that was successfully treated with surgical excision.

CASE REPORT

A 69-year-old male with a history of hypertension, hyperlipidemia, type 2 diabetes mellitus, asthma, gout, and obstructive sleep apnea presented for evaluation of a mass on the right buttock, which had been present for 4 years. The patient initially thought the lesion was a pimple, but it subsequently grew to the size of a quarter before starting to drain clear, light yellow fluid. The lesion continued to grow and became both pruritic and painful, bleeding intermittently during the 6 months prior to presentation. The patient also endorsed decreased appetite and associated 45-pound unintentional weight loss over an 8 month period. He did not have any additional skin lesions, and denied any personal or family history of skin cancer.

Physical examination revealed a 10 x 8 cm firm, pink, ulcerated, pedunculated mass on the right buttock (Figure 1A-C). The clinical differential diagnosis included squamous cell carcinoma, verrucous carcinoma, and dermatofibrosarcoma protubersans. A shave biopsy was performed, which showed dermal nodules of stratified squamous epithelium connecting to the epidermis. The nodules showed palisading of the outer layer, extensive epithelial central proliferation, and trichilemmal keratinization (Figure 2). For the most part, the nodules were well-circumscribed, with pushing margins, but multiple areas showed disruption of the lesion’s silhouette with invasive features.
Cellular atypia and inconspicuous mitosis were also seen. No lymphovascular or perineural invasion was identified. Overall, the biopsy was felt to be most consistent with an invasive carcinoma arising within a PPT.

Figure 1. Physical examination revealed a 10 x 8 cm firm, pink, ulcerated, pedunculated mass on the right buttock.

Figure 2. Shave biopsy at 4X magnification (A) revealed dermal nodules of stratified epithelium with palisading of the outer layer, extensive epithelial central proliferation, and trichilemmal keratinization. The biopsy at 20X magnification (B) showed areas with invasive clusters and cords of atypical cells.

The patient subsequently underwent staging work-up with a CT abdomen and pelvis with contrast, which showed pathological right external iliac and right inguinal lymphadenopathy. Core needle biopsies of both areas of lymphadenopathy revealed reactive lymphoid hyperplasia, and it was determined that the patient had no clear evidence of distant disease. Radical resection was performed with 1 cm margins.
Figure 3. The excision from the right posterior thigh on 4X magnification (A) and 10X magnification (B) showed a large multi-lobulated neoplasm with nodules of stratified squamous epithelium, proliferating nests, and trichilemmal keratinization. Several tongue-like projections extended into the surrounding stroma which revealed prominent desmoplasia.

Histopathology revealed nodules of stratified squamous epithelium, proliferating nests, and trichilemmal keratinization (Figure 3). Occasional lobules showed dense homogenous keratinous contents. Some areas of the tumor demonstrated reverse maturation, focal stromal invasion, and associated desmoplasia. Such areas were best characterized as well-differentiated squamous cell carcinoma. Perineural or lymphovascular space invasion were not seen.

DISCUSSION

PPTs were first described by E. Wilson Jones in 1966 as “proliferating epidermoid cysts”. Since then, the same entity has been referred to by a number of different names, including pilomatrixoma, trichochlamydocarcinoma, giant hair matrix tumor, proliferating trichilemmal cyst, and trichilemmal pilar tumor. Clinically, these benign tumors may resemble keratinous or sebaceous cysts and are most commonly found on the scalp of older women. They often grow slowly over months to years, resulting in large, exophytic masses with variable degrees of ulceration. The prognosis of PPTs is generally favorable as surgical resection is curative; however, malignant transformation within these tumors has been reported.

Benign PPTs are diagnosed histologically based on the presence of circumscribed lobules of epithelial cells, often in continuity with the epidermis, with peripheral palisading. Trichilemmal keratinization is also an important feature that can help differentiate PPTs from SCCs. In 2004, Ye et al. examined 76 cases of PPTs and categorized them into 3 groups based on degree of stromal invasion and cytologic atypia: benign, low-grade malignant, and high-grade malignant. The lesion in our case revealed expansive nodules of squamous epithelium with trichilemmal keratinization. There were several invasive foci of clusters and cords of tumor invading into the surrounding stroma that demonstrated mild to moderate cytologic atypia and inconspicuous mitosis. The latter were surrounded by desmoplastic stroma and variable lymphoplasmacytic infiltrate.
overall features argued against a benign neoplasm, favoring a diagnosis of low-grade malignant PPT based on the proposed classification scheme by Ye et al.

To the author’s knowledge, this case represents only the second report of a SCC arising within a potential PPT. The first case, reported by Di Pace et al. in 2017, involved an 82-year-old woman who developed a SCC with areas of trichilemmal differentiation. The lesion had a thickness of 25 mm, infiltrating the hypodermis, and was noted to have evolved quickly, unlike the slow-growing tumor in this case. Similarly, staging work-up was negative for metastasis, and surgical resection was curative without the need for additional treatment. Di Pace et al. described the lesion as a “SCC of a proliferating trichilemmal tumour nature”.

In conclusion, PPTs may be malignant and have the potential to metastasize. As such, accurate diagnosis is important to facilitate treatment in a timely manner. We present this case to describe a rare clinical presentation of a well-differentiated SCC arising within a PPT and underscore the importance of clinicopathological correlation for diagnosing this rare disease entity.

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