



Pilomatricoma of the scalp mimicking poorly differentiated cutaneous carcinoma on positron emission tomography/computed tomography (PET/CT) scan and fine-needle aspiration (FNA) cytology

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INTRODUCTION

Pilomatricoma is a common benign cutaneous tumor arising most commonly in the first 2 decades of life. Despite its frequency, pilomatricoma remains a diagnostic challenge to nondermatologists and is misdiagnosed in 45% to 75% of cases.^{1,2} A concerning clinical and cytologic appearance after fine-needle aspiration (FNA) has contributed to the misdiagnosis of malignancy in several cases.³⁻⁵ Fludeoxyglucose (FDG) avidity of these lesions at the time of positron emission tomography/computed tomography (PET/CT) scan can also raise concern for a malignant process.^{3,6} Clinical suspicion must remain high, and appropriate biopsy techniques should be used to avoid the misdiagnosis of pilomatricoma.

REPORT OF A CASE

The patient is a 21-year-old woman with medical history significant for myotonic dystrophy. She presented originally to the otolaryngology department with a new 1.2-cm draining cyst on the right anterior scalp. She also had a 1-cm nodule on the left posterior neck, along the hairline. She reported that the lesions were painful when she brushed her hair. They had not bled and were otherwise asymptomatic. She is a nonsmoker and had no history of skin cancer. She reported no fever, chills, nausea,

Abbreviations used:

| | |
|---------|--|
| FDG: | fludeoxyglucose |
| FNA: | fine-needle aspiration |
| FNAC: | fine-needle aspiration cytology |
| PET/CT: | positron emission tomography/ computed tomography |

vomiting, fatigue, decreased appetite, night sweats, or weight loss at presentation.

FNA of the anterior scalp and posterior neck lesions was performed. FNA cytology (FNAC) of both specimens found hypercellular aspirate composed of numerous clusters and singly dispersed immature epithelial cells with scant cytoplasm, high nuclear-to-cytoplasmic ratio, fine granular chromatin, inconspicuous nucleoli, occasional mitoses, and frequent apoptotic bodies. This cytomorphologic appearance suggested a diagnosis of poorly differentiated carcinoma. Cell block staining found some cells positive for AE1/AE3 and p40, consistent with a diagnosis of squamous cell carcinoma (Fig 1).

Whole-body PET/CT scan was subsequently performed for staging purposes and initial treatment strategy. The study found focal intense abnormal FDG uptake within the right-sided frontal scalp subcutaneous soft tissue density (maximum standard uptake value, 9.1). The symptomatic lesion on the left posterior neck also showed FDG avidity. Both

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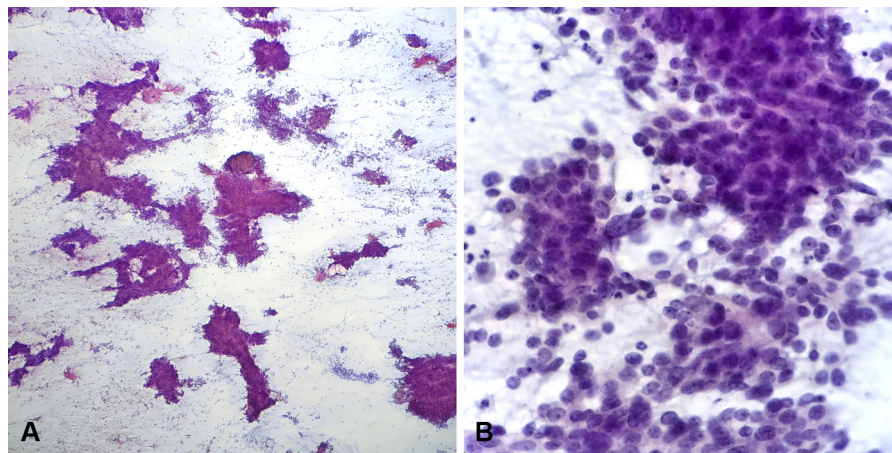


Fig 1. A and B, Fine-needle aspirate shows hypercellular sample composed of epithelial cells with scant cytoplasm, overlapping nuclei, high nuclear-to-cytoplasmic ratio, irregular nuclear membrane, inconspicuous nucleoli, and frequent apoptosis. (Pap stain.)

lesions were found to be partially calcified. There were no other areas of FDG avidity noted on the examination.

The patient underwent wide excision of both lesions with frozen section evaluation. Histologic examination of the frozen and paraffin fixed tissue found ghost cells, inflammatory cells, and hair follicles containing rare clusters of primitive basaloid cells (Fig 2). These features supported a final diagnosis of 2 lesions of pilomatricoma. FNA samples were re-reviewed, and showed predominantly primitive epithelial cells. No ghost cells were identified in the specimen (Fig 1). The findings, although initially suggestive of poorly differentiated carcinoma, were determined to represent the primitive follicular epithelium of pilomatricoma when reassessed in concert with the resected lesions.

DISCUSSION

Pilomatricoma, also known as *calcifying epithelioma of Malherbe*, is a benign cutaneous tumor arising in the dermal layer of hair-bearing skin. Clinically, pilomatricoma presents as a solitary slow-growing, firm and mobile mass usually ranging in size from 0.5 to 5 cm. Overlying skin may take on a blue or red hue and will classically exhibit the tent sign. It is most commonly found in the head and neck. It is described more frequently in females and is thought to occur most commonly in the first 2 decades of life.^{2,7}

Most cases of pilomatricoma appear to be sporadic, although rare familial forms of the disease exist. Pilomatricoma has been found to occur in association with other diseases and syndromes such as Gardner syndrome, Rubinstein-Taybi syndrome,

Turner syndrome, and myotonic muscular dystrophy such as in our case.^{1,2}

Simple enucleation is the treatment of choice for these lesions. FNA of these lesions can result in the inappropriate diagnosis of cutaneous malignancy.^{1,3,4} Findings of ghost cells and a monomorphic population of basaloid cells are pathognomonic for pilomatricoma. However, the limited sample from FNA can result in a predominance of basaloid cells, which can easily be mistaken for basal cell carcinoma or basaloid squamous cell carcinoma. Also, ghost cells are not always evident on cytologic specimens.⁶ Given the high rate of misdiagnosis of pilomatricoma with FNA, most investigators agree that the diagnostic effectiveness of FNAC is variable and depends heavily on the experience of the pathologist and overall clinical suspicion.⁵

The immunostaining results in this case further contributed to misdiagnosis. Given the cytomorphologic evaluation of the FNA specimens, malignancy was suspected. With a working diagnosis of malignancy and positive epithelial markers (CK AE1/AE3 and p40), a carcinoma of squamous cell etiology was favored. Although immunohistochemical staining can be useful in differentiating the cell type of a clonal population of cells, it is critical to consider alternative diagnoses, which can also have similar staining patterns. For epithelial markers like these, the differential diagnosis is expansive and includes many benign and malignant entities—one of which is pilomatricoma.⁸

PET/CT is a common imaging modality used in the workup of malignancy. It is well established that false-positive results frequently occur secondary to nonmalignant processes that increase glucose uptake such as infection, inflammation, lymphoid

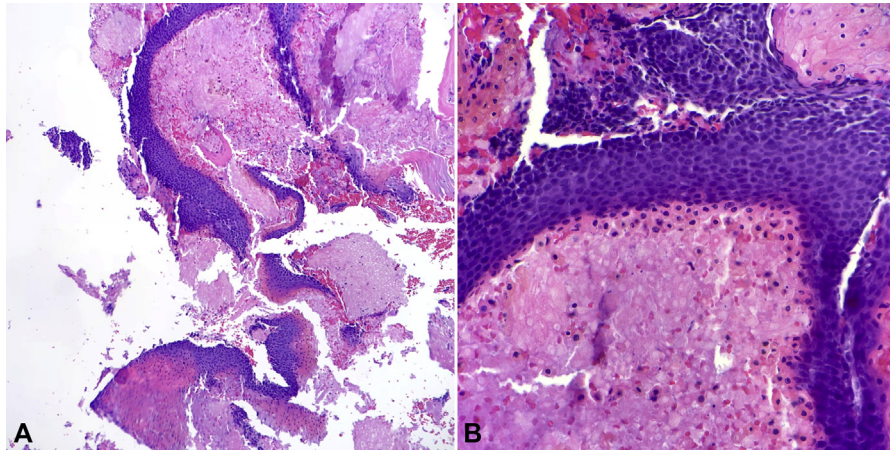


Fig 2. **A** and **B**, Histology findings of the resected lesion show abundant keratinous debris, ghost cells, and basaloid cells with high nuclear-to-cytoplasmic ratio and inconspicuous nucleoli. (Hematoxylin-eosin stain.)

follicular hyperplasia, and granulomatous diseases.⁹ FDG avidity in pilomatricoma is hypothesized to occur secondary to intralesional inflammatory cell recruitment and increased mitotic rate.^{7,10} Rare instances of local FDG-avid lymph nodes in cases of pilomatricoma have occurred, and a similar mechanism is proposed.^{7,10}

The diagnosis of pilomatricoma can be challenging. Noninvasive modalities such as FNAC and PET/CT scan can lead to the misdiagnosis of malignancy and unnecessary procedures. Simple biopsy techniques, which allow for histologic evaluation of the entire lesion, can both confirm the diagnosis and serve as an effective treatment for this benign entity.

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