ABSTRACT

Pilomatricoma, also known as calcifying epithelioma of Malherbe, is a rare skin tumor originating from the hair follicle matrix. We report a case of pilomatricoma in a 50-year-old woman, presenting as a rapidly growing pretibial mass. Malignant pilomatricoma is associated with potentially fatal metastases and are clinically and histologically indistinguishable from benign pilomatricoma. Thus, an 18F-fluorodeoxyglucose positron emission tomography-computed tomography (18F-FDG PET/CT) scan was requested for staging, revealing marked FDG uptake restricted to the primary lesion and no evidence of separate disease. The purpose of this report is to demonstrate the importance of PET/CT in the staging of this FDG-avid tumor; the malignancy of which is often first revealed by metastases. Our case also demonstrates that pilomatricoma should be included in the differential diagnosis of a rapidly growing peripheral soft-tissue mass; conventionally, the domain of sarcoma.

Keywords: Imaging, pilomatricoma, positron emission tomography-computed tomography, sarcoma

INTRODUCTION

Pilomatricoma is a superficial skin tumor originating from hair follicle cells. Its rarity combined with its varied histology and clinical presentation means that it often evades clinical diagnosis. This is significant because malignant pilomatricoma is associated with potentially fatal metastatic capabilities.1 While malignant pilomatricoma is less common than benign, it is currently impossible to differentiate between benign and malignant pilomatricoma histologically, and there is evidence that benign pilomatricoma may be capable of malignant transformation.2,3 Thus, metastases could be the first sign of serious disease in an otherwise benign-appearing lesion. This emphasis on the detection of metastasis makes positron emission tomography-computed tomography (PET/CT) an extremely powerful tool for the staging of pilomatricoma.

CASE REPORT

A 50-year-old woman presented with a rapidly enlarging, symptomatic superficial subcutaneous mass immediately lateral to the left tibial tuberosity (Figure 1a, axial T1 magnetic resonance imaging [MRI]). An ultrasound scan revealed a 20-fold increase in volume over a period of 10 months, as well as new peripheral and central neovascularization (Figure 1b, Doppler Ultrasound of Mass). Ultrasound-guided biopsy of the lesion was performed, revealing fibrous tissue containing necrotic epithelial cells and abundant keratin; histology initially suggestive of squamous cell carcinoma. However, further review at mechanical diagnosis and treatment...
updated the diagnosis to probable pilomatricoma. Due to the diagnostic doubt, and rapid enlargement, a staging whole-body $^{18}$F-fluorodeoxyglucose ($^{18}$FDG) PET/CT scan was performed (C, axial fused $^{18}$FDG PET/CT on SUV 0-6 scale) demonstrating the lesion as a markedly FDG avid 3 cm × 1 cm × 3 cm soft-tissue mass (SUVmax = 10.4), with no other abnormal scan findings. The lesion was excised, with final histology confirming it as a pilomatricoma with no evidence of mitoses.

DISCUSSION

Pilomatricoma is a rare skin appendage neoplasm arising from hair follicle cells. Diagnosis of pilomatricoma is a challenge, with literature review demonstrating an accuracy rate of preoperative diagnosis ranging from 0% to 30%. This is due to the low specificity of imaging techniques such as CT and MRI combined with the often misleading results of fine-needle biopsy, as was the case in our patient even with a core biopsy. For this reason, preoperative diagnosis of pilomatricoma is often based on the clinical presentation: generally as a slow-growing mass in the head-and-neck regions of children. Thus, the presentation of a pilomatricoma in a middle-aged woman as a rapidly growing pretibial mass is unusual.

Treatment of pilomatricomas is through wide local excision because of their potential for malignant transformation, and the significant metastatic potential of malignant pilomatricoma. Our case adds to the increasing body of evidence to suggest that pilomatricomas are significantly FDG avid, with a July 2020 PUBMED literature search using the keywords “Pilomatricoma” and “PET” retrieving four articles reporting on four adult patients with benign pilomatricoma. Unlike our case, all of these tumors presented more typically for pilomatricoma as a slowly growing mass of the upper body but crucially they all demonstrated FDG avidity (SUVmax ranging from 3.86 to 16.5). This makes PET/CT staging beneficial in the management of pilomatricomas; hence, the detection of these metastases by PET/CT may be the only indication that the tumor is malignant due to its ability to detect early and therefore treatable metastases. There are currently no molecular or immunohistochemical markers able to differentiate between benign and malignant pilomatricomas, In conclusion, our case demonstrates that pilomatricoma is an important differential diagnosis for a rapidly growing peripheral soft-tissue mass; conventionally, the domain of sarcomas. Furthermore, the case illustrates the importance of PET/CT staging in the management of pilomatricoma, due to its potential for metastatic disease.

DECLARATION OF PATIENT CONSENT

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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CONFLICTS OF INTEREST

There are no conflicts of interest.

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