CASE REPORT

Digital papillary adenocarcinoma of the ankle

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ABSTRACT

Digital Papillary Adenocarcinoma (DPA) is a very rare neoplasm of sweat glands, with approximately 100 cases in the literature. We present a rare case of DPA of the ankle in a 66-year-old African American man. DPA has a high potential for metastases to the lung and lymph nodes, and initial treatment often entails wide excision or complete digital amputation. DPA typically presents as a painless lesion or mass of the distal digits of the hands or feet. With the limited available literature regarding this malignancy, there is no standard approach to treatment. Wide excisional margins with sentinel lymph node mapping and biopsy are based on extrapolation for other tumors. In the following report, we discuss a representative case and the current literature on the presentation, treatment and pathology of this rare neoplasm.

Key Words: Digital papillary adenocarcinoma, Skin adnexal cancer

1. INTRODUCTION

Digital Papillary Adenocarcinoma (DPA) is a rare malignant adnexal tumor arising from the sweat gland. It was first described in 1979 by Helwig as an eccrine acrospiroma, but later understood to be a papillary adenocarcinoma. These tumors may also present as digital papillary adenomas which are extremely rare. These tumors are most common in Caucasian men in their 5th-7th decades of life, and most often present on the volar surface of fingers and toes, the palmar surface of the hand, and the soles of the feet. They are three times more common on the digits of the hand compared to the foot.

DPA is asymptomatic, typically presenting as a solitary mass, cyst or ulcer. The majority appear as a flat or slightly elevated lesion, while some are more swollen and thickened. DPA is often undiagnosed for months to years due to its rarity, typically slow progression and lack of symptoms. Left undiagnosed, DPA will metastasize in 14% of cases, 70% of which will be pulmonary metastases. Standard treatment of DPA includes surgery, with either excision or amputation of the digit involved. While there is no consensus regarding excision margins, a retrospective study has suggested superior results with amputation versus wide local excision.

Because DPA is rare, it can be misdiagnosed as metastatic papillary adenocarcinoma of the colon, thyroid, or breast. It is essential to rule out other forms of malignancy before diagnosing DPA and expert pathologic consultation is imperative. Continual follow up with thorough exam and possible chest X-rays to monitor for lung metastasis is important due to the high rate of recurrence and metastasis.

2. CASE REPORT

A 66-year-old African American male presented with a 10-year history of a slowly growing, asymptomatic lesion on his left lateral heel. He had a significant past medical history of hypertension, obesity, sleep apnea, localized prostate cancer previously treated by radical prostatectomy, gyneco-
mastia, hypogonadism, arthritis, and tobacco abuse for over 30 years. The lesion recently caused him occasional shooting pain, leading to excision of the lesion by the patient’s podiatrist. The pathology report revealed digital papillary adenocarcinoma. The neoplasm was composed of many dilated tubular structures lined by multiple layers of cuboidal epithelium with eosinophilic cytoplasm, oval vesicular nuclei and prominent nucleoli. In deeper sections, tubules showed an infiltrative growth pattern and have irregular and hyperchromatic nuclei with a central comedo type necrosis. The neoplasm extended to the base of the biopsy and re-excision was recommended. He was then referred to surgical oncology at Wake Forest University for further evaluation (see Figures 1-3).

Figure 1. Dilated tubular structures lined by multiple layers of cuboidal epithelium. In deeper part, tubules show an infiltrative growth pattern (H&E, ×10).

Figure 2. Tubules with irregular and infiltrative growth pattern and central necrosis (H&E, ×40).

Figure 3. Tubules lined by 2-3 layers of cuboidal epithelium with eosinophilic cytoplasm, round to oval vesicular nuclei, prominent nucleoli, and central comedo type necrosis (H&E, ×200)

Prior to re-excision, the left lateral heel showed a well-healed scar at the excision site with no palpable inguinal, popliteal, cervical or axillary lymphadenopathy on exam. On May 2nd 2017, the patient underwent a wide local excision of his scar, with a 2 cm margin, closed with split thickness skin graft and sentinel lymph node mapping with biopsies of the popliteal and inguinal basins without complication. The pathology report revealed negative margins of the 6 cm × 4.2 cm × 0.4 cm excision of the left heel with left popliteal and inguinal lymph nodes negative for malignant neoplasm. The patient’s post-operative course was uncomplicated with excellent graft uptake and donor site healing.

3. DISCUSSION

DPA is a rare malignancy with approximately 100 reported cases. DPA of the ankle is a particularly unique presentation of the disease. In 1987 Kao et al. published a retrospective study of 57 cases of DPA. They examined the basic clinical presentation of DPA, the disease course, treatment, and differential diagnosis. Of the cases studied, 75.4% were of the hand, 23% of the foot, and 1.6% of the calf. Additionally, all 31 cases presented by Suchak et al. occurred on either a finger or toe. None of the cases presented on the ankle. One case report conducted by Carter et al., presented a similar case of a 54-year-old African American male with DPA of the left ankle. The patient similarly underwent wide excision of the lesion with negative margins. Due to the paucity of data, the importance of negative margins with excision remains unclear, but can be extrapolated from other tumor types.

The evaluation of DPA suggests it must be distinguished
from other more common sweat gland neoplasms, such as eccrine acrospiroma and chordoid syringoma. Kao et al. suggests primarily distinguishing DPA based on histological evidence. While all three types of neoplasms share a glandular appearance with focal papillary epithelial projections, DPA has a complex tubuloalveolar pattern distinguishing it from its more common counterparts. Unlike DPA, both eccrine acrospiroma and chordoid syringoma typically present on the head, neck or proximal limbs and have low rates of recurrence or metastases. These factors help further clinically separate them from DPA, and underscore the importance of expert dermatopathologic consultation. In addition, DPA must be differentiated from distant adenocarcinoma metastatic to the skin, with primary sources such as breast, thyroid, and lung being in consideration. When the diagnosis is in doubt, immunohistochemical staining for p63 may be of utility as many sweat gland tumors will demonstrate greater than 25% staining for p63, whereas metastatic adenocarcinoma should not stain for p63. The importance of clinical history and location is also crucial in the diagnosis, as sweat gland neoplasms can stain positively for CK7 and negative for CK20, thereby mimicking a metastatic breast cancer.

The primary therapy for DPA is wide excision or digital amputation for lesions without evidence of metastases. There is no well-accepted recommendation for, or against, sentinel lymph node biopsy based on the current literature. Kao et al. determined an overall recurrence rate of 57.1% with local excision, and concluded that without evidence of metastasis, amputation is superior to local excision. When this study was re-examined by Duke et al., recurrence rates dropped to only 5% if adequately resected. In a retrospective study conducted by Suchak et al., 31 cases of aggressive digital papillary adenocarcinoma were examined. Of the 31, nine had complete excisions and eight had digit amputation. Of those with amputations, four patients had sentinel lymph node biopsies which returned negative, and they did not develop metastatic disease. Of the total, six patients developed metastases, one to a lymph node, four to the lung, and one to both a lymph node and lung. Three of these patients developed metastases after 5, 14 and 20 years. From these results, the authors were unable to draw conclusions regarding the benefit of sentinel lymph node biopsy but emphasized the importance of long-term follow-up. However, with the modest risk of sentinel node biopsy and the potential benefit, it seems prudent to offer the procedure with a wide excision in otherwise fit patients.

While the majority of cases have shown a high rate of aggressive neoplastic behavior such as recurrence and metastasis, some cases have existed for years without metastasis. A case of a DPA of the third digit of the hand reports that the patient had the lesion for 20 years while it slowly increased in size. After complete excision there was no recurrence or metastasis for two years. An additional case presented a patient with a 15-year history of a painless mass found to be DPA; it was excised and the patient remained without recurrence or metastasis for two and a half years. An additional case presented a patient with a 15-year history of a painless mass found to be DPA; it was excised and the patient remained without recurrence or metastasis for two years. These cases, in addition to our case, suggest a low-grade type of DPA seems to differ in clinical course to the more common aggressively malignant DPA.

DPA is a rare malignancy typically with slow rates of progression but the potential for highly aggressive behavior. Expert dermatopathologic opinions in these cases is important to secure the correct diagnosis. Wide excision with negative margins is recommended with consideration of sentinel node mapping.

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