

CASE REPORT

Malignant proliferating trichilemmal tumor of the forearm: a case report of an unusual location of a rare cutaneous adnexal tumor

Abelardo Quiñones-Martínez¹, Christian Nieves-Ríos^{2,*},
and Guillermo Bolaños-Ávila¹

¹Department of Surgery, St. Luke's Episcopal Hospital, Ponce 00733, Puerto Rico and ²Department of Medicine, Ponce Health Sciences University, Ponce 00716, Puerto Rico

*Correspondence address. Department of Medicine, Ponce Health Sciences University, Ponce 00716, Puerto Rico. Tel: +1-787-392-4408; Fax: +1-787-848-1306; Email cnieves17@stu.psm.edu

Abstract

Proliferating trichilemmal tumor (PTT) is a rare cutaneous adnexal neoplasm of the hair follicle that undergoes outer root sheath differentiation in the isthmus. Histological hallmarks include trichilemmal keratinization, lack of granular layer and lobular proliferation of squamous epithelium with glycogenated clear cells. It affects predominantly elderly women, especially over the scalp. In some cases, malignant transformation can occur. However, only a few cases are reported in the literature. We hereby describe the case of a patient diagnosed with malignant PTT (MPTT) in the dorsal forearm, 2 years after undergoing surgical excision of a squamous cell carcinoma in the same topography. Thus, providing a thorough description of the clinical presentation of MPTT will assist surgeons in diagnosing and treating this rare tumor.

INTRODUCTION

Proliferating trichilemmal tumor (PTT) is a rare skin neoplasm derived from the outer root sheath of a hair follicle that undergoes trichilemmal keratinization without granular layer interposition [1, 2]. This lesion affects primarily the scalp (90%) of women (84%) in the fourth to eighth decades of life [3, 4]. However, it has been found in other locations including the forehead, wrist and chest [3].

Histological features of PTT include dermal lobular proliferation of squamous epithelium, in some areas constituted by clear cells containing glycogen, circumscribed by a glassy and fairly cellular stroma [2]. These lobes exhibit trichilemmal keratinization with occasional calcification, usually in the central area [2]. Other reports have shown partial transformation to

spindle cell carcinoma with transition zones between squamous epithelium and spindle cells [5].

Clinically, PTT usually arises as a pilar cyst with a rapid increase in the size of a previously small, asymptomatic and solitary lesion [4]. Occasionally, it appears as an exophytic lesion or a large plaque with surface nodularities [6]. Grossly, PTT is well circumscribed, lobulated and sharply demarcated from the surrounding tissues [3]. The range in size is from 0.4 to 10.0 cm in diameter, and the rates of local recurrence and regional lymph node metastasis are 3.7 and 1.2%, respectively [3, 7].

Malignant transformation of PPT is a very rare and unusual finding that can be indicated by the presence of tissue invasion and cellular atypia [1, 8]. MPTT can be differentiated from its

Received: August 7, 2018. Accepted: October 9, 2018

Published by Oxford University Press and JSCR Publishing Ltd. All rights reserved. © The Author(s) 2018.

This is an Open Access article distributed under the terms of the Creative Commons Attribution Non-Commercial License (<http://creativecommons.org/licenses/by-nc/4.0/>), which permits non-commercial re-use, distribution, and reproduction in any medium, provided the original work is properly cited. For commercial re-use, please contact journals.permissions@oup.com

benign form by the presence of abnormal mitoses, high mitotic counts, cellular pleomorphism, cytological and architectural atypia, necrosis, infiltrating margins and aneuploidy [4]. However, a morphometric analysis of PTT found no major differences between benign and malignant PTT [9]. Furthermore, lesions in which benign and malignant areas coexisted have been described [6]. These might suggest that the biological behavior of PTT is not related to its histologic appearance, and that malignant transformation may have occurred within the pre-existing proliferating trichilemmal cyst [4, 6]. Here, we describe the clinical presentation of a MPTT for the purpose of aiding surgeons in the diagnosis and eventual treatment of this rare tumor.

CASE REPORT

A 81-year-old woman with a past medical history of arterial hypertension and diabetes mellitus type 2, presented to our hospital with urosepsis and *de novo* uncontrolled diabetes mellitus type 2. After admission and physical exploration, she was found with a right forearm ulcerated mass. The patient refers that since a year and a half ago, she presented with an initially slowly growing, painless protruding ulcerated mass with spontaneous bleeding. However, a sudden increase in size has been noted within the past several months. Medical history was positive for squamous cell carcinoma (SCC), which was excised 2 years ago. On gross examination an ulcerated, protuberant, and hard mass with irregular tissue, elevated margins and active bleeding was identified at the right dorsum forearm (Fig. 1). Prior trauma to the region was denied. Patient underwent a wide excision with 1.0 cm free margins of normal tissue with subsequent placement of skin graft from her right thigh over her right forearm (Fig. 2). A tissue sample was sent for pathological analysis.

Tissue examination report returned as a skin tumor, consisting of an elliptical segment of tan skin tissue measuring $9.5 \times 7.5 \times 4.5 \text{ cm}^3$. The lateral margins were red while the medial margins were blue inked. At the epidermal surface centrally located there was a rubbery to firm grayish-tan polyp flower-like tumor mass that measured $8.5 \times 5.5 \times 4.0 \text{ cm}^3$. On section, it was composed of a homogenous, solid white-tan tumor tissue and grossly appears to be located at 0.6 cm from the nearest inked lateral margin and at 0.4 cm from the deep margin. The resected margins were free of tumor. The diagnosis of ulcerated and invasive malignant PTT

(MPTT) was made. We could not find the recent status of the patient as she was lost to follow-up.

DISCUSSION

PTT is an uncommon cutaneous neoplasm derived from the outer root sheath of hair follicles, specifically in the isthmus. The histologic hallmark of PTT is the presence of trichilemmal keratinization without the formation of a granular layer [10]. It predominantly affects the scalp of elderly women. However, here we present a rare case of a patient with a past medical history of SCC on the forearm, reporting 2 years later at our hospital with a MPTT in the same topography.

Malignant transformation of PTT is a very unusual event, classifying it as one of the rarest trichilemmal tumors [8]. Histological features that distinguish MPTT from its benign form include cytological and architectural atypia, necrosis and infiltration [2]. Nonetheless, some of these characteristics have also been described in SCC. Moreover, there are only a few cases of MPTT reported in the literature. These factors could hinder the diagnosis of MPTT over other cutaneous tumors.

Considering that our patient had history of SCC and the fact that histologically, MPTT may mimic SCC [4], a clinical approach to differentiate both tumors might be appropriate. MPTT differs from a SCC by its sharp demarcation from the surrounding stroma [1]. Moreover, it has been described as a well circumscribed, lobulated, exophytic lesion that usually arises as a pilar cyst with a rapid increase in size. This description clearly matches the clinical presentation of our patient. Another feature that distinguish MPTT is the lack of origin from an overlying precursor epidermal lesion such as actinic keratosis [3]. This was corroborated in our report, favoring the diagnosis of MPTT. On the other hand, malignant transformation of PTT should be considered in long-standing lesions that show rapid growth or exophytic enlargement [6], a finding that clinched the diagnosis. The mainstay therapeutic approach of MPTT is wide local excision with 1 cm margin of normal tissue



Figure 1: Forearm before surgical excision showing a protuberant mass with irregular tissue and active bleeding.



Figure 2: Forearm after surgical excision and placement of skin graft from the right thigh.

[10]. On the other hand, the treatment of first choice for SCC is surgical excision including Mohs' micrographic surgery due to the low incidence of local recurrence and metastasis after this approach [11].

As previously mentioned, different cutaneous tumors commonly have overlapping features, complicating the diagnosis of these neoplasms. Therefore, the purpose of this report is to provide integral information about the clinical presentation of MPTT in an atypical location and to emphasize that special attention must be taken during the evaluation process, to ensure accurate diagnosis and treatment of this tumor.

ACKNOWLEDGEMENTS

None.

CONFLICT OF INTEREST STATEMENT

None declared.

REFERENCES

1. Mathis ED, Honningford JB, Rodriguez HE, Wind KP, Connolly MM, Podbielski FJ. Malignant proliferating trichilemmal tumor. *Am J Clin Oncol* 2001;**24**:351–3.
2. Noto G, Pravata G, Aricò M. Malignant proliferating trichilemmal tumor. *Am J Dermatopathol* 1997;**19**:202–4.
3. Brownstein MH, Arluk DJ. Proliferating trichilemmal cyst: a simulant of squamous cell carcinoma. *Cancer* 1981;**48**:1207–14.
4. Sutherland D, Roth K, Yu E. Malignant proliferating trichilemmal tumor treated with radical radiotherapy: a case report and literature review. *Cureus* 2017;**9**:e999.
5. Val-Bernal JF, Garijo MF, Fernández F. Malignant proliferating trichilemmal tumor. *Am J Dermatopathol* 1998;**20**:433.
6. Mehregan AH, Lee KC. Malignant proliferating trichilemmal tumors—report of three cases. *J Dermatol Surg Oncol* 1987;**13**:1339–42.
7. Ye J, Nappi O, Swanson PE, Patterson JW, Wick MR. A clinicopathologic study of 76 cases with a proposal for definition of benign and malignant variants. *Am J Clin Pathol* 2004;**122**:566–74.
8. Goyal S, Jain BB, Jana S, Bhattacharya SK. Malignant proliferating trichilemmal tumor. *Indian J Dermatol* 2012;**57**:50–2.
9. Aricò M, Noto G, Pravata G. Proliferating trichilemmal tumor: morphometric evaluations in four cases. In: Panconesi E, ed. *Dermatology in Europe*. Oxford: Blackwell, 1991, 810–3.
10. Satyaprakash AK, Sheehan DJ, Sangüeza OP. Proliferating trichilemmal tumors: a review of the literature. *Dermatol Surg* 2007;**33**:1102–8.
11. Motley R, Kersey P, Lawrence C. Multiprofessional guidelines for the management of the patient with primary cutaneous squamous cell carcinoma. *Br J Dermatol* 2002;**146**:18–25.