Malignant Proliferating Trichilemmal Tumor: Clinical Presentations, Treatment, and Outcomes

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Abstract

Background: Malignant proliferating trichilemmal tumor (MPTT) is very rare malignant tumors of hair follicles derived from outer root sheath. This tumor is mostly located on the head and neck of elderly women. Regional or distant metastasis is possible. In this study, we present clinical features and treatment outcomes of cases diagnosed as MPTT. Furthermore, we aimed to emphasize a different clinical form of the tumor that can be misdiagnosed clinically. Patients and Methods: A retrospective evaluation of five cases operated between September 2009 and February 2017 Celal Bayar University Faculty of Medicine, Plastic Reconstructive and Aesthetic Surgery Department at were included in the study. Clinicopathological features of patients, type of surgery, and follow-up information were evaluated. Results: Four patients were female. Average age was 72.2. All of the lesions were located on the head and neck. All patients had a history of rapid growth of lesions. Patients were scanned with computed tomography. There was no metastasis at the time of diagnosis. None of the patients needed adjuvant therapy. Mean follow-up time was 11.8 months. None of the patients developed recurrence or metastasis. Conclusions: These tumors resemble basal cell or squamous cell carcinoma. Rapid progress of benign form of the tumor should address malignant transformation. There is no consensus about adjuvant therapy. Screening for metastasis and close follow-up are mandatory.

Keywords: Hair follicle, malignant outer root sheath tumor, malignant proliferating trichilemmal tumor

Introduction

Malignant proliferating trichilemmal tumor (MPTT) is one of the malignant tumors of hair follicles derived from outer root sheath. This tumor was first described using the term “proliferating epidermoid cyst” by Jones in 1966, and later, Saida described MPTT in 1983.\(^1\) This tumor commonly affects the scalp of elderly women found as a nodular mass. Invasion of the tumor into neighboring tissues and being accompanied with necrosis are accepted as findings of malignancy. Squamous cell carcinoma (SCC) should be considered in differential diagnosis because of similar morphology.\(^2,3\)

MPTT is extremely rare and can be aggressive. Regional and distant metastasis have been reported.\(^4,5\) We report clinical presentation and treatment outcomes of five cases diagnosed as MPTT.

Patients and Methods

All authors state to the effect that the principles of the 1975 Declaration of Helsinki were followed during this research. Patient informed consent form was taken from all patients for publication. Patients operated between September 2009 and February 2017 diagnosed as MPTT according to the incisional or excisional biopsy results were included in the study. Age and gender of the patients, localization and size of the tumor, duration of tumor before the first examination, and any previous surgery were recorded. All patients were asked about acceleration of tumor growth at any time. Tumors were resected with 1 cm margin, and defect reconstructions were performed. Histopathological examinations including immunohistochemical methods were performed. Patients were scanned with computed tomography (CT) (SOMATOM Force, Siemens, Erlangen, Germany) for regional or distant metastasis. None of the patients needed adjuvant therapy. Mean follow-up time was 11.8 months. None of the patients developed recurrence or metastasis.

Conclusions: These tumors resemble basal cell or squamous cell carcinoma. Rapid progress of benign form of the tumor should address malignant transformation. There is no consensus about adjuvant therapy. Screening for metastasis and close follow-up are mandatory.

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metastasis and consulted to the radiation oncology department for adjuvant therapy. Follow-up was performed monthly for the first 3 months, then at 6th month, and at the end of the 1st year. After the 1st year, yearly examinations were suggested. Patients were evaluated with physical examination and evaluated with ultrasonography (USG). Reconstruction method, scan data for metastasis, necessity of adjuvant therapy, follow-up time, and tumor recurrence data were recorded.

RESULTS

Five patients diagnosed as MPTT were included in the study. Four patients were female (M/F: 1/4). The average age was 72.2 (ranged between 55 and 94). Two patients had previous surgery for their lesions at different centers. Both of them were reported as PTT. Duration of lesions until surgery was between 1 and 12 years with a mean of 7.6 years. All patients had rapid growth of their lesions for the last couple of months. Four of the lesions were located on the scalp, and the other was located on the right temporal aspect of the face [Figures 1 and 2]. Dimensions of the tumors were between 3 cm × 2.5 cm and 15 cm × 16 cm. Ulceration was detected on the lesions of two patients [Table 1].

Tumors were resected with 1 cm margin. The defect reconstruction of three patients was performed with split-thickness skin graft (STSG), one of them was reconstructed with SCALP flap and STSG [Figures 3 and 4] and the other one with primary suture. The margins were tumor free in all patients.

Histopathologically, multilobulated tumors characterized by atypical squamous cells, cystic areas, widespread trichilemmal keratinization, and extensive necrosis were seen [Figure 5]. Peripheral palisading was usually present. Calcification was present in one lesion. When we evaluate these tumor groups immunohistopathologically, high molecular weight cytokeratin positivity and negative carcinoembryonic antigen were detected.

Follow-up time ranged from 6 to 18 months with the mean of 11.8 months. One patient did not accept CT scan and the other patients had no regional or distant metastasis at the time of diagnosis. One patient had bilateral occipital multiple lymphadenomegaly evaluated intraoperatively. Bilateral occipital lymph node resection was performed. Totally, 10 lymph nodes were reported as reactive by histopathologic evaluation [Figures 6 and 7]. None of the patients needed adjuvant therapy. One patient died as a result of cerebrovascular accident 15 months after surgery. There was no recurrence, regional, or distant metastasis during follow-up [Table 1].

DISCUSSION

Cutaneous adnexal carcinomas are extremely rare, reported to represent 0.005% of all skin tumors. These carcinomas have heterogeneous origin and are believed to originate from undifferentiated stem cells.[6] MPTT is categorized in the follicle-originated malign tumors according to their differentiations and originates from outer root sheath epithelium.[7]
Malignant proliferating trichilemmal tumor (MPTT) develops in the course of time from the foci of proliferating epithelial cells in the trichilemmal cysts due to factors such as trauma or chronic inflammation. More than 80% of the patients are elderly women. About 90% of cases occur on the scalp. Our results were consistent with the literature. One of the tumors was located on the face in our report. Unusually, it was a pigmented dark blue lesion that was resembling malignant melanoma.

Ye et al. classified PTT into three groups based on pathological characteristics: (a) Group 1 - benign (circumscribed tumor with “pushing” margins, modest nuclear atypia; no absence of pathologic mitoses, necrosis, and invasion of nerves or vessels), (b) Group 2 – low-grade malignant (similar to Group 1 but manifested irregular, locally invasive tumor with involvement of the deep dermis and subcutis), and (c) Group 3 – high-grade malignant lesions (invasive growth patterns, marked nuclear atypia, pathologic mitotic forms, and geographic necrosis, with or without involvement of nerves or vascular structures). Our cases had invasive borders and nuclear atypia but no necrosis, atypical mitosis, or vascular-perineural invasion. Then, they can be classified as low-grade MPTT according to this classification. The absence of regional or distant organ metastasis and recurrence supports this conclusion.

All patients gave a history of rapid growing over the last couple of months. Two of the patients had previous surgery and were diagnosed as PTT at the first time. This should point

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**Table 1: Clinical features of tumors and previous surgery information of the patients with pathologic diagnosis, treatment methods, and follow-up information**

<table>
<thead>
<tr>
<th>Patient</th>
<th>Gender</th>
<th>Age</th>
<th>Localization</th>
<th>Size</th>
<th>Duration</th>
<th>Previous surgery</th>
<th>Treatment</th>
<th>Metastasis</th>
<th>Follow-up (month)</th>
<th>Relapse</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Female</td>
<td>80</td>
<td>Scalp</td>
<td>9 cm × 10 cm × 4 cm</td>
<td>12 years, rapid growing for 6 months</td>
<td>2007, reported as PTT</td>
<td>E + scalp flap + STSG</td>
<td>None</td>
<td>18</td>
<td>None</td>
</tr>
<tr>
<td>2</td>
<td>Female</td>
<td>66</td>
<td>Scalp</td>
<td>4.5 cm × 4 cm × 3 cm with ulceration</td>
<td>10 years, rapid growing for a year</td>
<td>No</td>
<td>E + STSG</td>
<td>None</td>
<td>6</td>
<td>None</td>
</tr>
<tr>
<td>3</td>
<td>Female</td>
<td>55</td>
<td>Scalp</td>
<td>7 cm × 7 cm × 3.5 cm</td>
<td>7 years, rapid growing for 2 months</td>
<td>No</td>
<td>E + STSG</td>
<td>None</td>
<td>12</td>
<td>Exitus</td>
</tr>
<tr>
<td>4</td>
<td>Male</td>
<td>94</td>
<td>Temporal</td>
<td>3 cm × 3 cm × 0.4 cm</td>
<td>1 year, rapid growing for 2 months</td>
<td>No</td>
<td>E + primary suture</td>
<td>Unknown</td>
<td>16</td>
<td>None</td>
</tr>
<tr>
<td>5</td>
<td>Female</td>
<td>66</td>
<td>Scalp</td>
<td>15 cm × 16 cm × 5.5 cm with ulceration</td>
<td>8 years, rapid growing for one and a half year</td>
<td>2016, reported as PTT</td>
<td>E + STSG + LNB</td>
<td>None</td>
<td>7</td>
<td>None</td>
</tr>
</tbody>
</table>

E: Excision, STSG: Split-thickness skin graft, LNB: Lymph node biopsy, PTT: Proliferating trichilemmal tumor

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**Figure 4:** Postoperative view of the patient 18 months after surgery

**Figure 5:** Infiltrative squamous island with central trichilemmal keratinization (H and E, ×40)
out that malignant transformation of these tumors from benign precursors may occur.

Diagnosis of MPTT is based on histopathologic examination of hematoxylin-eosin stained slides. Trichilemmal keratinization and peripheral palisading are signs which represent follicular root sheath origin of tumors. Nuclear atypia and marked cellular pleomorphism with atypical mitoses are associated with malignant behavior.\(^\text{[12]}\)

Metastatic rate of MPTT is up to 25% for Grade 3 lesions.\(^\text{[11]}\) As a result, radiographic evaluation is imperative once the diagnosis has been established. Siddha et al. used adjuvant radiotherapy to treated area and neck of the patient with regional metastasis to prevent recurrence in MPTT.\(^\text{[13]}\) Considering the aggressive nature of the malignant variant and high rates of locoregional as well as distant failures, Dubhashi et al. advocated adjuvant radiotherapy with regional lymph node metastasis in their report.\(^\text{[4]}\) On the other hand, Siddha et al. suggested that wide local excision with a 1 cm margin of normal tissue is the treatment of choice for MPTT.\(^\text{[13]}\) Sharma et al. recommend a 2 cm margin for recurrent lesions.\(^\text{[10]}\)

Furthermore, Mohs surgery was reported without recurrence with 6-month follow-up.\(^\text{[14]}\) We treated all our patients with wide local excision with 1 cm margin. We scanned our patients with cranial, cervical, and thoracoabdominal CT except one because he refused further investigations. None of them had regional or distant metastasis, and adjuvant RT was not prescribed. Closed follow-up was recommended to the patients as monthly visits for the first 3 months, at the 6th month, at the 1st year and once a year after the 1st year. Patients were evaluated with cervical USG, and no recurrence or metastasis was detected.

This study evaluates the clinical behavior of MPTT by investigating the rapid progression of these tumors before diagnosis. All patients gave a history of rapid growing of their tumors for a couple of months. This may be a sign of malignant transformation of benign tumors, especially for MPTT. As our knowledge, this is the first study focusing on rapid progression before diagnosis. Furthermore, we represent different clinical forms of MPTT with pigmentation and peduncle.

**Conclusions**

MPTT is a rare malignant tumor of the hair follicle that can present with different clinical features. These tumors resemble SCC clinically. Although these tumors are rarely reported, actual incidence is unknown. Regional or distant metastasis can be seen. There is no consensus about adjuvant therapy but screening for metastasis and close follow-up are mandatory.

**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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Nil.

**Conflicts of interest**

There are no conflicts of interest.

**References**

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