Case Report

Multiple Proliferating Trichilemmal Tumors: A Report of Two Cases and Literature Review

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Abstract
Proliferating trichilemmal tumors are rare tumors of the skin appendages derived from the outer root sheath of the hair follicle. It occurs predominately in the elderly women and often occurs on the scalp. Multiple proliferating trichilemmal tumors of the scalp are relatively uncommon. The author(s) report two cases of multiple trichilemmal tumors. A 40-year-old man who presented with two years history of multiple lesions on his scalp. The second, a 56-year-old man presenting with a familial multiple trichilemmal tumors located on the scalp for ten years, which has recurred after excision. All the lesions on the two patients scalp were totally excised. Then histopathological examination revealed the diagnosis of multiple proliferating trichilemmal tumors. They are alive and without evidence of recurrence after 13 and 16 months follow up respectively. Multiple proliferating trichilemmal tumors are mostly located on the scalp. Total excision is the main treatment and scalp reconstruction is necessary if it develops ulceration. Radiotherapy and Chemotherapy may be needed when malignant proliferating trichilemmal tumors are diagnosed. As a result of it malignant and local recurrence or distant metastasis potential, long-term closed clinical follow up after treatment is needed.

Key words: Malignant proliferating trichilemmal tumors; Multiple proliferating trichilemmal tumors; Proliferating trichilemmal tumor

Introduction
Proliferating trichilemmal tumor (PTT) also called Proliferative trichilemmal cyst or Pilar cyst, or Pilar tumor, a well-documented series was first reported in 1966 as “proliferating epidermoid...
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cysts” by Wilson-Jones (1). It is defined as a rare
eoplasm derived from the outer root sheath of
the hair follicle. Various diagnostic terms and
different names for this lesion have been used
throughout the medical literature (2). It is usually
a solitary lesion and most commonly occurs in the
elderly women (3). The lesion is in areas of dense
hair follicular concentrations, such as the scalp, in
90% of patients, with 10% occurring on other
anatomic locations (4,5).

Multiple proliferating trichilemmal tumors
are relatively uncommon. To our knowledge only
nine cases of multiple proliferating trichilemmal
tumors had been reported in the English Medical
Literature (6-12,19). The previous reported cases
provided limited information about multiple
proliferating trichilemmal tumors. The Clinical
features and Management of multiple proliferating
tumors remain unclear. We present two new cases
of multiple proliferating trichilemmal tumors of
the scalp and review the cases reported till date.

CASE DESCRIPTION

1. Case 1

A 40-year-old man presented with a 2- year
history of multiple (four) slowly enlarging lesions
on his scalp (Figure 1 A and B). There is no
previous family history of similar scalp lesions.
On physical examination, the patient’s scalp was
studded with four mobile masses, ranging from
1cm to 2.5cm in size. The masses extended from
the anterior hairline to the posterior hairline with
two in the parietal region, one in the occipital
region and one in the left temporal region. The
masses were fluctuant and soft, and all were
covered by hair. There was no fixation to the
underlying bone area and cervical lymph nodes
were non- palpable. With a preliminary clinical
diagnosis of multiple scalp masses, all lesions
were totally excised. The four gross specimens
had complete capsular, the lesions ranged from
0.8cm to 1.6cm in diameter and all were cystic

Microscopically, histopathological slides
revealed the characteristic structures of multiple
proliferating trichilemmal tumor; trichilemmal-type keratinization, ordinary
trichilemmal cyst, calcification, an eosinophilic
hyaline membrane surrounding the tumor lobules
and palisading of cells at the periphery of the
tumor lobules (Figure 1 D and E). A final
histopathological diagnosis of multiple
proliferating trichilemmal tumors was made. No
clinical recurrence or distant metastasis was seen
after 13 months of regular follow-ups.

2. Case 2

A 56-year-old man presented with 10 years
history of multiple (eighteen) slowly enlarging
lesions on his scalp after multiple trichilemmal
tumor excision ten years ago (Figure 2 A and B).
Some masses in the parietal and occiput regions
of the patient’s scalp were growing quickly in size
in the last one year (Figure 2 A). The patient has a
family (father, mother, brothers and sisters)
history of similar multiple scalp masses (dates
were not obtained). The patient was otherwise
healthy with no significant past medical history.

On physical examination, the patient’s scalp
was studded with multiple fluctuant masses,
ranging in size from 0.5cm to 4cm in diameter.
The masses extended from the anterior hairline to
the posterior hairline with most localized in the
occiput and parietal regions. The masses were
fluctuant and soft, and they were all covered by
hair. One mass in the occiput region has a
tendency of ulceration (Figure 2 A). There was no
fixation to the underlying bone area, and cervical
lymph nodes were non-palpable. An initial
clinical diagnosis of multiple scalp masses was
made, and every mass was totally excised. Eighteen gross specimens that have completed
capsular were obtained. The lesions ranged from
0.3cm to 3.5cm in diameter and all were soft and

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cystic (Figure 2 C). The resected tissue was placed in 10% formalin for histopathological examination.

Histopathological examination revealed the characteristic structures of multiple trichilemmal tumors; trichilemmal-type keratinization, calcification, an eosinophilic hyaline membrane surrounding the tumor lobules and palisading of cells at the periphery of the tumor lobules (Figures 2 D and E). Multiple proliferating trichilemmal tumors were diagnosed at last. At 16 months of follow up, there was no evidence of local recurrence or distant metastasis.

Figure 1. Gross pathology specimen demonstrated and histological assessment of multiple proliferating trichilemmal tumors of Case 1.
1. A and B: a 2-year history of multiple (four) slowly enlarging lesions on his scalp.
2. C: gross pathology specimen of four lesions ranged from 0.8 to 1.6 cm in diameter and all were cystic.
3. D and E: histologic characteristic of multiple proliferating trichilemmal tumors: trichilemmal-type keratinization, ordinary trichilemmal cyst, And calcification, an eosinophilic hyaline membrane surrounding the tumor lobules and palisading of cells at the periphery of the tumor lobules (hematoxylin and eosin [H&E], original magnification, 200 ×.

Discussion

Proliferating trichilemmal tumors (PTT), also called proliferative trichilemmal cyst or Pilar cyst, or Pilar tumor, a well-documented series was reported first in 1966, as “proliferating epidermoid cysts” by Wilson-Jones (1), is a rare neoplasm derived from the outer root sheath of the hair follicle. The same lesion has been reported under various names, including “subepidermal acanthoma”, “invasive hair matrix tumor”, “invasive pilomatrixoma”, “trichochlamydocarcinoma”, “hydatiform keratinous cysts”, “giant hair matrix tumor”, “trichochlamydoacanthoma” and Pilar cysts and tumors, which were review by Satyaprakash AK (13). It is usually a solitary lesion and most commonly occurs in elderly women (3).
Figure 2. Gross pathology specimen demonstrated and histological assessment of multiple proliferating trichilemmal tumors of Case 2.

1. A and B: a 10-year history of multiple (eighteen) slowly enlarging lesions on his scalp after multiple trichilemmal tumors excision ten years ago, the multiple fluctuant masses ranged in size from 0.5 to 4 cm in diameter, and one mass in the occipital region has a tendency of ulcerating.

2. C: Eighteen gross specimens from 0.3 to 3.5 cm in diameter and all were soft and cystic.

3. D and E: histological characteristic of multiple proliferating trichilemmal tumors: trichilemmal-type keratinization, calcification, an eosinophilic hyaline membrane surrounding the tumor lobules and palisading of cells at the periphery of the tumor lobules (hematoxylin and eosin [H&E], original magnification, 200×).

It is in areas of dense hair follicular concentrations, such as the scalp in 90% of patients, with 10% occurring on the other anatomical locations (4, 5). Although much less common, including the neck, trunk, groin, mons pubis, vulva and gluteal region; the upper and lower extremities, including the elbow, the dorsum of the hand and the index finger; the face including the forehead, nose, eyelid, lip, and intraoral; And even the base of the skull have been reported in the literature and were reviewed by Satya Prakash AK (13). The usual clinical presentation of PTT is a subcutaneous cystic nodule that has been present for many years and slowly progresses to a large nodular mass, often following a history of trauma or chronic inflammation, and it may present as masses with ulceration (3, 6, 14).

The diagnosis of PTT is dependent on the histopathological examination. The pathologic findings of PTT are trichilemmal-type keratinization, ordinary trichilemmal cyst, and calcification, an eosinophilic hyaline membrane surrounding the tumor lobules and palisading of cells at the periphery of the tumor lobules (1-4). According to the PTT histological characteristics, it is divided into benign PTT, low-grade malignant PTT and Malignant PTT (MPTT) (13). The clinical treatment of various kinds of PTT is different. The accepted treatment for benign PTT is simple local excision, low-grade malignant PTT require wide local excision with 1-cm margin of normal tissue to
Although PTT is a benign tumor, it is known to recur and trend to malignant transformation (13). PPTs may recur after local excision, and the reported recurrence rate of the disease is 3.7% (4, 17, 18). Although the true rate of local recurrence of PTT and metastatic MPTT is unknown, PTT local recurrence or distant metastasis has been reported in many of the literature (5, 9, 11, 13, 15).

We found two (2/11) cases of the multiple PTT were multiple MPTT and local recurrence or metastasis after excision (Table 1). One was reported by Yoleri et al. (10). A 64-year-old man presented with rapid growth and ulceration in one of his masses on the scalp. The right semi-scalp skin including galea was excised, and a split-thickness skin graft was applied over the periosteum. Eight months later, local metastasis was seen on the right pre-aurlicular area, and wide excision with a skin graft was performed. The patient was referred for radiotherapy. Another case was reported by Makiese et al. (11). A 51-year-old female patient presented with multiple lesions on the scalp. After the multiple lesions were excised, MPTT was confirmed diagnosis, and chemotherapy (CAV protocol, cisplatin, Adriamycin, and vindesine) was given. The second excision after local recurrence. There was no evidence of local recurrence or distant metastasis for 54 months following up.

Also, Gallant et al (19) reported a case of a 58-year-old previously healthy white female presented to her primary care provider with the desire to remove a right posterior scalp cyst for cosmesis. The lesion was excised and recurred locally eight months post resection along with palpable right posterior cervical lymph node. Following the recurrence, modified radical posterior neck and lymph node dissection, and adjuvant chemotherapy (carboplatin plus paclitaxel) with concurrent radiation were given. There was evidence of disease recurrence and chemotherapy (consisting of docetaxel and crisplatin) were given even. The patient then opted for no aggressive measures and was symptomatically treated.
Table 1 Published reports of multiple proliferating trichilemmal tumors.

<table>
<thead>
<tr>
<th>Study</th>
<th>Cases</th>
<th>Age/ Sex</th>
<th>Time (years)</th>
<th>History</th>
<th>Size (cm) / No. of masses</th>
<th>Rapid Growth</th>
<th>Ulcer Time (mo)</th>
<th>Growth Pattern</th>
<th>Location</th>
<th>Treatment given</th>
<th>Histology</th>
<th>Recurrence or Metastasis</th>
<th>Follow up (mo)</th>
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<tbody>
<tr>
<td>Our study</td>
<td>2</td>
<td>40/M</td>
<td>2</td>
<td>No</td>
<td>1-2.5 4</td>
<td>No</td>
<td>No</td>
<td>Circumscribed</td>
<td>Scalp</td>
<td>Excision</td>
<td>PTT</td>
<td>Local recurrence</td>
<td>NED, 13</td>
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<tr>
<td>Gallant JN [19]</td>
<td>1</td>
<td>58/F</td>
<td>10</td>
<td>NS</td>
<td>1-2 Solitary</td>
<td>Yes</td>
<td>NS</td>
<td>Circumscribed +Infiltrative</td>
<td>Scalp</td>
<td>Excision</td>
<td>CAV protocol</td>
<td>Local Metastasis</td>
<td>NED, 26</td>
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<td>Wang x [12]</td>
<td>1</td>
<td>47/M</td>
<td>20</td>
<td>NS</td>
<td>1.5-6 Numerous</td>
<td>Yes</td>
<td>No</td>
<td>Circumscribed +Infiltrative</td>
<td>Multiple</td>
<td>Excision</td>
<td>PTT</td>
<td>No</td>
<td>NS</td>
</tr>
<tr>
<td>Makiesee O [11]</td>
<td>2</td>
<td>51/F</td>
<td>NS</td>
<td>Trauma</td>
<td>12 Numerous</td>
<td>No</td>
<td>No</td>
<td>Circumscribed</td>
<td>Scalp</td>
<td>Excision</td>
<td>CAV</td>
<td>No</td>
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<tr>
<td>Yoleri L [10]</td>
<td>1</td>
<td>64/M</td>
<td>30</td>
<td>No</td>
<td>1-7 Numerous</td>
<td>Yes</td>
<td>Yes</td>
<td>Circumscribed + infiltrative</td>
<td>Scalp</td>
<td>Excision</td>
<td>TR SR</td>
<td>Local metastasis</td>
<td>NED, 54</td>
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<td>Hendricks DL [9]</td>
<td>1</td>
<td>47/F</td>
<td>8</td>
<td>NS</td>
<td>1.5-6 Numerous</td>
<td>Yes</td>
<td>Yes</td>
<td>Circumscribed</td>
<td>Scalp+ Back</td>
<td>Excision</td>
<td>SR PTC + MPTT</td>
<td>NED, 8</td>
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<td>59/F</td>
<td>20</td>
<td>NS</td>
<td>1-5 15</td>
<td>Yes</td>
<td>Yes</td>
<td>Circumscribed</td>
<td>Scalp</td>
<td>Excision</td>
<td>SR PTT + MTC</td>
<td>No</td>
<td>NS</td>
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<td>Chang SJ [7]</td>
<td>1</td>
<td>69/F</td>
<td>25</td>
<td>Inheritance</td>
<td>3-15 Numerous</td>
<td>Yes</td>
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<td>Circumscribed</td>
<td>Scalp</td>
<td>Excision</td>
<td>SR PTC</td>
<td>No</td>
<td>NS</td>
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<tr>
<td>Erdem H [6]</td>
<td>1</td>
<td>70/F</td>
<td>23</td>
<td>Trauma</td>
<td>1.2-15 8</td>
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<td>Yes</td>
<td>Circumscribed</td>
<td>Scalp</td>
<td>Excision</td>
<td>SR PTC</td>
<td>No</td>
<td>NS</td>
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</tbody>
</table>

Note: PTT, proliferating trichilemmal tumor; PTC, proliferating trichilemmal cyst; MTC, multiple trichilemmal cyst; MPTT, malignant proliferating trichilemmal tumor; NED, no evidence of disease; CAV, cisplatin, Adriamycin, and vindesine; TR, radiotherapy; SR, scalp reconstruction; NS, not state.
following development of community acquired pneumonia until her death.

Conclusion

Multiple Proliferating Trichilemmal Tumors are lesions localized in dermis or subcutaneous tissue, which may become circumscribed or infiltrative, sometimes exhibit ulceration, and are solid or partially cystic. Multiple PTTs are also mostly located in the scalp. Totally excised is the main treatment, scalp reconstruction is necessary if it with ulceration, and wide surgical excision and radiotherapy or chemotherapy may be also needed when it diagnosed MPTT. Because of its malignant and local recurrence or distant metastasis potential, careful histologic examination and long-term closed clinical follow up after it treated is needed.

Declarations

1) Consent to publication
We declare that all authors agreed to publish the manuscript at this journal based on the signed Copyright Transfer Agreement and followed publication ethics.

2) Ethical approval and consent to participants
Written consent was taken from patients.

3) Disclosure of conflict of interests
We declare that no conflict of interest exists.

4) Funding
None

5) Authors’ Contributions
Authors contributed to this paper with the design (XH, YY, and IK), literature search (XH, YY, IK , AM, and ZG ), drafting (XH, YY, and IK), revision (AM and ZG), editing (IK, XH and YY) and final approval (XH, YY, IK, AM and ZG).

6) Acknowledgement
None

7) Authors’ biography
None

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