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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

cysts" by Wilson-Jones (1). It is defined as a rare neoplasm 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 (Figure 1C). The resected tissue was placed in 10% formalin for histopathological examination.

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

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.

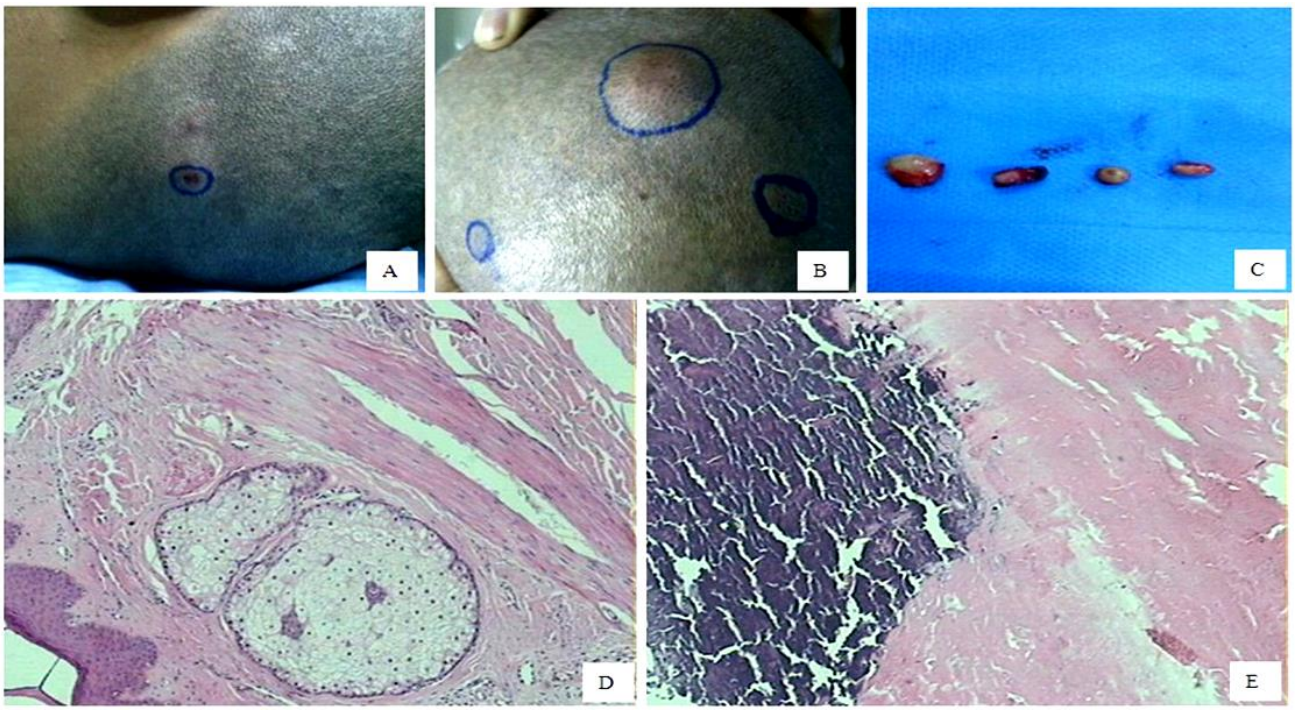

Figure 1. Gross pathology specimen demonstrated and histological assessment of multiple proliferating trichilemmal tumors of Case 1.

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: histological 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).

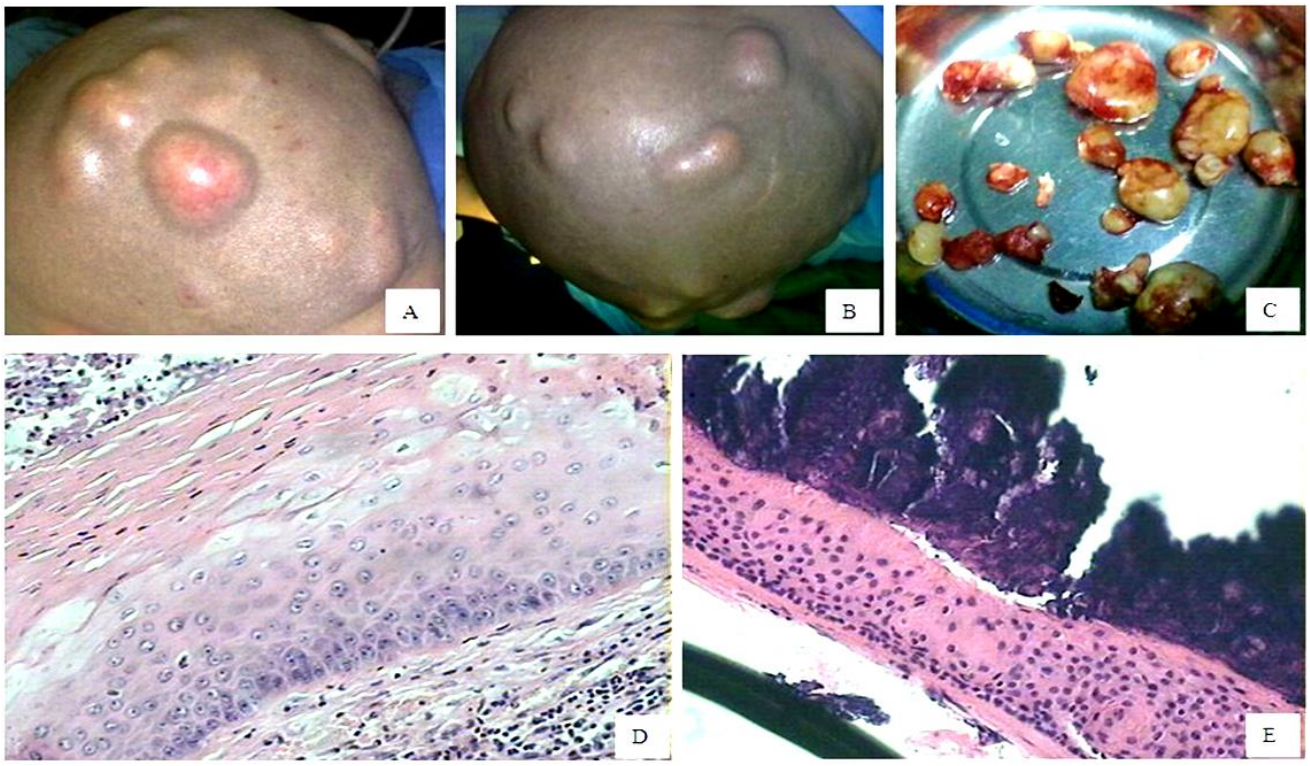

Figure 2. Gross pathology specimen demonstrated and histological assessment of multiple proliferating trichilemmal tumors of Case 2.

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

prevent recurrence and MPTT needs more aggressive therapeutic measures such as nodal dissection, radiotherapy, or chemotherapy should be considered in addition to wide local excision (13). As a result of its malignant and local recurrence or distant metastasis potential, long-term closed clinical follow up after treatment is needed for low-grade malignant PTT and MPTT (13).

PTT is usually a solitary lesion reported in the literatures, but it can exhibit multiple lesions at times (6-12, 19) and is relatively rare. To our knowledge only nine well-documented cases of multiple PTTs have been published to date in the English Language Literature (6-12,19). This paper presents two new multiple PTTs cases. Table 1 summarizes their clinicopathological features. There were 6 female and 5 male patients, ranging in age from 40 to 70 years (Table 1). But solitary lesion of PTT case was seen more frequently in women older than 50 years (3, 7). Like PTT, the most common site involved of multiple PTTs was also on the scalp (4, 5) (9/11, multiple PTTs of 9 cases occurred on the scalp, one case in several anatomic areas include bank, face, neck, axillae, gluteal region, and perineum, another case in her scalp and back). Multiple PTTs range in size from 0.3 to 16cm (Table 1). Duration of multiple PTTs ranges between 1 and 30 years (Table 1. Duration of cases reported). But duration of PTT ranges between 4 and 50 years in the literature (15). The number of lesions of the multiple PTTs ranges from 4 to 18 (Table 1. No. of masses). Some lesions of multiple PTTs sudden increase in size (7/11), and to become ulcerated (5/11) over the past 2 to 5 months (Table 1 Ulceration time) Three cases had history of trauma (Table 1).

Surgical excision was done in all the patients. Eight (8/11) cases were multiple benign PTT. Local excision was given for the multiple benign PTT (8/11), and reconstruction of the scalp defect after resection of a large lesion with ulceration (5/11) was also done. Close clinical follow-up was carried out and there was no evidence of local recurrence or distant metastasis for multiple benign PTT. 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-auricular 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.

Study	Cases	Age/ Sex	Time (years)	History	Size (cm) / No. of masses	Rapid Growth	Ulcer Time (mo)	Growth Pattern	Location	Treatment given	Histology	Recurrence or Metastasis	Follow up (mo)
		56/M	10	Inheritance	0.3-3.5 18	Yes	No	Circumscribed	Scalp	Excision	PTT	Local recurrence	NED,16
Our study	2	40/M	2	No	1-2.5 4	No	No	Circumscribed	Scalp	Excision	PTT	No	NED,13
Gallant JN [19]	1	58/F	10	NS	1-2 Solitary	Yes	NS	Circumscribed +Infiltrative	Scalp	Excision CAV protocol	MPTT	Local Metastasis	NED, 26
Wang x [12]	1	47/M	20	NS	1.5-6 Numerous	Yes	No	Circumscribed +Infiltrative	Multiple	Excision	PTT	No	NS
		65/M	NS	Trauma	12 Numerous	No	No	Circumscribed	Scalp	Excision	PTT	No	NED, 105
Makiese O [11]	2	51/F	NS	Trauma	16 Numerous	No	No	Circumscribed +infiltrative	Scalp	Excision CAV	MPTT	Local recurrence	NED,54
Yoleri L [10]	1	64/M	30	No	1-7 Numerous	Yes	Yes 2	Circumscribed + infiltrative	Scalp	Excision TR SR	PTC + MPTT	Local metastasis 8 mo	NED,8
Hendricks DL [9]	1	47/F	8	NS	1.5-6 Numerous	Yes	Yes 2	Circumscribed	Scalp+ Back	Excision SR	PTT + MTC	No	NED, 6
Bengoeche -Beeby MP [8]	1	59/F	20	NS	1-5 15	Yes	Yes NS	Circumscribed	Scalp	Excision SR	PTT + MTC	No	NS
Chang SJ [7]	1	69/F	25	Inheritance	3-15 Numerous	Yes	Yes NS	Circumscribed	Scalp	Excision SR	PTC	No	NS
Erdem H [6]	1	70/F	23	Trauma	1.2-15 8	Yes	Yes 5	Circumscribed	Scalp	Excision SR	PTC	No	NS

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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