

## Rare Presentation of Proliferating Trichilemmal Tumor of the Left Shoulder Treated by Rhomboid Flap

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### Abstract

**Background:** Proliferating trichilemmal tumor (PTT), also known as giant hair matrix tumor or invading pilomatrixoma, is an uncommon adnexal neoplasm that is usually benign but occasionally shows malignant potential, for which postoperative histopathology is required to confirm diagnosis.

**Case Presentation:** A 53-year-old woman with an unremarkable medical and surgical history presented with a slowly enlarging lump on the posterior aspect of the left shoulder, present for 20 years.

**Intervention:** Ultrasonography showed a large, heterogeneous mass; fine-needle aspiration cytology (FNAC) was inconclusive, and incisional biopsy was performed prior to definitive excision with Rhomboid flap reconstruction.

**Histology:** Histopathological examination revealed a circumscribed nodule with proliferating squamous epithelium, abrupt trichilemmal keratinization, and mild focal atypia, features consistent with PTT.

**Outcome:** The patient recovered well after excision and flap reconstruction, with postoperative imaging showing no clinical evidence of recurrence.

**Key Learning Point:** PTT should be considered in the differential diagnosis of longstanding soft-tissue masses even at atypical extra-scalp sites such as the shoulder, where histopathological confirmation and appropriate reconstructive planning are essential for optimal outcomes.

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## 1. Introduction

A proliferating trichilemmal tumor (PTT), also called giant hair matrix tumor or invading pilomatrixoma, is a rare, benign, exophytic tumor arising from the isthmus of the outer root sheath of the hair follicle. It occurs predominantly in women over 60 years old (female-to-male ratio 2.5:1); about 90% of lesions are solitary scalp tumors, with the remainder arising at sites such as the face, ear, neck, shoulder, trunk, anogenital area, and extremities. Lesion size typically ranges from under 1 cm to 10 cm (largest reported, 25 cm) and may continue to enlarge if untreated [1]. Although PTT generally follows a benign course, a small proportion behave aggressively, with local recurrence and malignant transformation reported; postoperative histopathology is therefore required for definitive diagnosis [2,3]. Histologically, PTT may resemble squamous cell carcinoma (SCC); nevertheless, they can be distinguished from one another because PTTs typically exhibit abrupt keratinisation, distinct glycogen storage cells, and a clear separation

from the surrounding tissue without infiltration or replacement of neighbours [4]. Complete surgical excision is the treatment of choice with excellent prognosis [4]. Because radiographic findings are often nonspecific and cytology may be equivocal due to abundant keratinous debris, histological examination remains the diagnostic gold standard [3]. The mainstay of treatment is complete surgical excision with histologically clear margins; tissue-sparing techniques such as Slow Mohs or Mohs micrographic surgery are effective alternatives that maximize tissue preservation [5,6].

## 2. Case Presentation

A 53 y/o female with insignificant past medical and surgical history presented to the Outpatient Department with a complaint of a lump on her left shoulder that had been growing progressively for the past 20 years and had reached a noticeable size. On general physical examination, the patient was conscious, alert, and

oriented to time, place, and person, with no apparent pallor, jaundice, edema, cyanosis, or clubbing. Her vitals were stable as well. Her systemic examination was unremarkable.

An Ultrasonographic examination was performed on the lesion which showed a solid, 7x6 cm, heterogeneous mass on the right lower quadrant of the left shoulder. These findings suggested a diagnosis of Lipoma. To confirm the findings, a Fine Needle Aspiration Biopsy (FNAC) was performed which could only demonstrate hemorrhagic non-diagnostic aspirate, and was therefore rendered inconclusive. So, the next course of action was to perform an incisional biopsy which led us to the diagnosis of PTT. Having received her provisional diagnosis, the patient underwent an excision of the tumor with Rhomboid flap reconstruction and the excised specimen was sent for postoperative histopathology. Microscopically, the specimen was observed to be a circumscribed nodule lined by proliferating squamous epithelium with abrupt areas of trichilemmal keratinization and mild focal atypia with very few mitoses and no necrosis. These findings were also consistent with a PTT. The patient was counseled and discharged.

After a month, a post-operative MRI was performed to look for any residual tumor which demonstrated an abnormal MR signal intensity area in posterior aspect of left shoulder subcutaneous planes adjacent to left trapezius muscle associated with adjacent significant fat strandings and post-surgical fibrotic changes at the surgical site. However, clinical evaluation ruled out any disease process.

### 3. Discussion

Proliferating trichilemmal tumors are rare lesions thought to originate from trichilemmal cysts (TC) [1]. They are composed of squamous epithelium with trichilemmal and no granular layer, and although typically benign, they can histologically mimic squamous cell carcinoma [2,4]. The scalp, neck, and face are the most common sites of involvement, with occasional occurrences on the trunk, back, buttocks, and vulva [5].

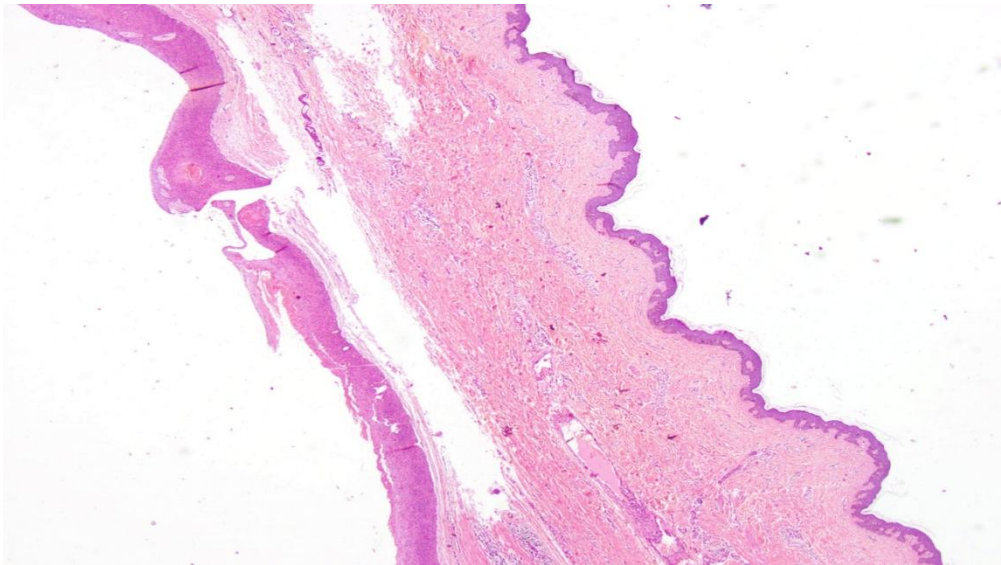
The presentation described here is a longstanding, slow-growing PTT on the shoulder rather than the scalp adds to a small body of literature documenting this tumor at atypical sites. This atypical location carries a diagnostic implication: because clinicians and radiologists are less likely to consider PTT outside the scalp, such lesions are more prone to being mistaken for other soft-tissue masses, delaying definitive diagnosis until histopathological confirmation is obtained [1].

Regardless of site, the management principles for PTT remain consistent: complete surgical excision is the treatment of choice [5]. Reconstructive options such as the Rhomboid flap offer a reliable means of closing the resulting defect with minimal tension and a favorable cosmetic outcome, which is particularly relevant in regions such as the shoulder where skin laxity may be more limited than on the scalp [3,6]. PTT can be treated with radiation, chemotherapy, lymph node dissection, or local surgical excision, but mainstay of treatment continues to be adequate surgical excision [7,8]. Mohs' micrographic surgery might be the best plan of action for treating trichilemmal cancers that are growing [9]. Wide surgical excision with margin analysis is necessary for MPTTs [10,11]. Treatment options have included chemotherapy for metastatic disease and radiation therapy for local recurrence [9].

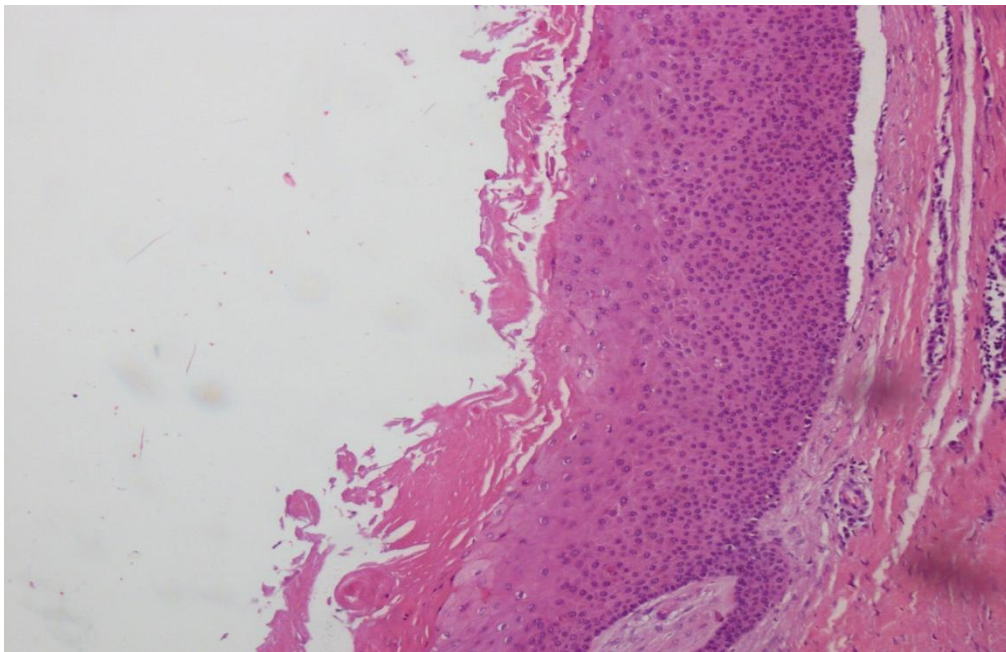
Malignant transformation with local invasion, recurrence, and rare metastasis has been described even though PTT typically follows a benign course, underscoring the importance of histological confirmation and long-term postoperative follow-up, particularly for lesions with atypical features [12,13]. A case of recurrent MPTT with regional lymph node metastasis in a young woman, illustrating that nodal spread, though uncommon can occur even outside the typical elderly demographic has also been reported [14]. Radiotherapy has also been proposed as an alternative when surgery is not feasible; Sutherland et al. achieved complete clinical response with radical radiotherapy alone in an elderly patient with significant comorbidities, sparing him extensive surgery [15]. Histopathological examination remains the gold standard for diagnosis and for distinguishing PTT from squamous cell carcinoma and other follicular tumors [16,17].

Postoperative imaging in PTT can be difficult to interpret, since post-surgical fibrotic change may radiologically mimic residual or recurrent disease. This underscores the value of correlating imaging findings with clinical examination, rather than relying on imaging alone, during postoperative surveillance.

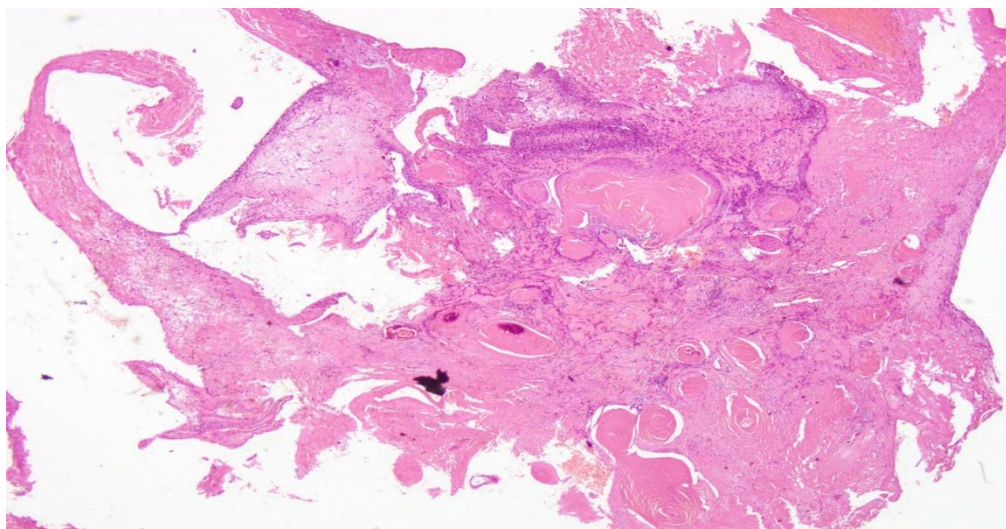
This report has several limitations. As a retrospective single-case description, detailed local examination findings, intraoperative details, and formal margin status were not consistently documented in the available clinical records, limiting the granularity of the operative and pathological description. Additionally, follow-up was relatively short, and long-term recurrence risk could not be assessed. Despite these limitations, the case adds to the small body of literature on PTT at atypical anatomical sites and highlights the diagnostic and reconstructive considerations relevant to such presentations.



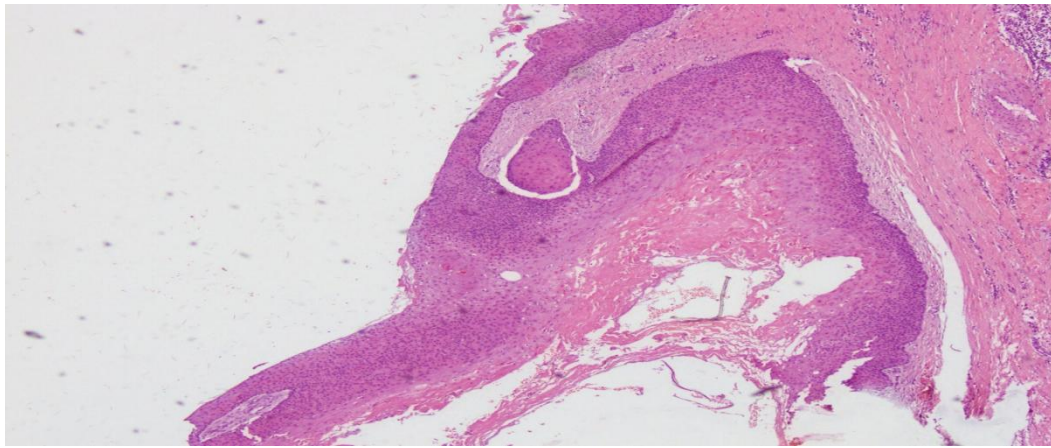
**Figure 1.** 10x image of skin exhibiting a cyst wall lined by squamous epithelium.



**Figure 2.** 20x image of proliferative squamous epithelium with uniform cells.



**Figure 3.** Proliferating squamous epithelium with abrupt trichilemmal type of keratinization.



**Figure 4.** proliferating squamous epithelium with abrupt trichilemmal type of keratinization.



**Figure 5.** Post-operative gross image of the surgical site post-procedure showing the defect repaired by a Rhomboid flap.

#### 4. Conclusion

This case underscores that proliferating trichilemmal tumor, though classically a scalp lesion of elderly women, can arise at atypical sites such as the shoulder after decades of indolent growth and should remain in the differential for any longstanding soft-tissue mass, regardless of location. Diagnosis was only achieved through histopathology after ultrasound and FNAC both proved inconclusive, reaffirming that imaging and cytology alone cannot exclude PTT. Complete excision with Rhomboid flap reconstruction achieved an excellent oncologic and cosmetic outcome, illustrating that even large, longstanding lesions in cosmetically sensitive regions can be managed successfully with careful surgical planning. Given the rare but documented potential for malignant transformation, histopathological confirmation and sustained postoperative follow-up remain essential, even when a lesion appears clinically benign.

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#### Ethics Statement

This case report was conducted in accordance with the ethical standards of the responsible institutional research committee and with the 1964 Helsinki Declaration and its later amendments. Formal ethical approval was waived by NISHTAR MEDICAL UNIVERSITY AND HOSPITAL as this is a single case report and does not constitute human subject research.

## Informed Consent Statement

The written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal. Patient confidentiality and anonymity have been maintained throughout the manuscript, and no identifying information is included.

## Data Availability Statement

All data generated or analyzed during this study are included within the published article.

## Author Contributions

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Manuscript drafting and revision: All authors.

All authors reviewed and approved the final manuscript.

## Conflict of Interest

The authors declare that they have no competing interests.

## Generative AI Statement

No generative artificial intelligence tools were used in the preparation of this manuscript.

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