



## Peri-Genital Proliferating Trichilemmal Cyst: A Clue to Underlying Malignancy

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### Abstract

*Proliferating trichilemmal cyst (PTC) is an uncommon tumour arising from outer root sheath of hair follicle which frequently present over the scalp as an isolated swelling 1 to 10 cm in size. PTC is assumed to arise from trichilemmal cyst with histological hallmark of trichilemmal keratinisation. Hardly ever, PTC can endure malignant transformation at which point, it is termed as malignant proliferating trichilemmal cyst (MPTC) which has a high chance of metastasis. We report a case of PTC at exceptional site in an elderly female at peri-genital area with later evolution to cutaneous squamous cell carcinoma (SCC) and metastasized to regional lymph nodes. On further exploration an underlying carcinoma cervix was also detected. So PTC served as a clue for cutaneous as well as cervical malignancy.*

**Keywords:** Proliferating Trichilemmal Cyst, Squamous Cell Carcinoma, Metastasis

### Introduction

Proliferating Trichilemmal Cyst (PTC) as an entity was first depicted by Wilson-Jones in 1966.<sup>[1]</sup> PTC is an rare tumour originating from the outer root sheath of hair follicle. It is thought to arise from trichilemmal cyst with histological hallmark of trichilemmal keratinisation.<sup>[2]</sup> Histologically this tumour have a capacity to imitate squamous cell carcinoma<sup>[3]</sup> and clinically PTC has the potential for malignant transformation as well as recurrences after surgical confiscation.<sup>[2]</sup> Scalp is frequently involved (>90% cases) site in individuals of more than 60 years of age, especially elderly females.<sup>[3]</sup>

PTC has various synonyms like pilar tumor of the scalp, giant hair matrix tumor, hydatidiform keratinous cyst, trichochlamydocarcinoma, and invasive hair matrix tumor.<sup>[2]</sup> We present a case of 70 year old elderly female who had a PTC at uncommon site in the perigenital area which later progressed to cutaneous squamous cell carcinoma of that region with metastasis to regional lymph nodes. On further investigations an underlying carcinoma cervix was also diagnosed.

### Case Report

A 70 year old female presented in dermatology OPD with a chief complaint of mildly painful

tumourous growth in the perigenital region for last 3 month. It started as a small pea-sized lesion which did remain non-progressive from last 2 years followed by rapid increase in size from past 3 months. There was a history of simultaneous appearance of two similar small lesions in the vicinity of old lesion. These lesions were asymptomatic initially but on presentation they were associated with mild pain. (Figure 1) There was also a history of scanty, watery discharge per-vaginum for 2 month duration. Patient was otherwise healthy with no other significant complaints with no history of post menopausal bleeding. There was no history suggestive of any underlying chronic disease like tuberculosis, hypertension and diabetes mellitus.

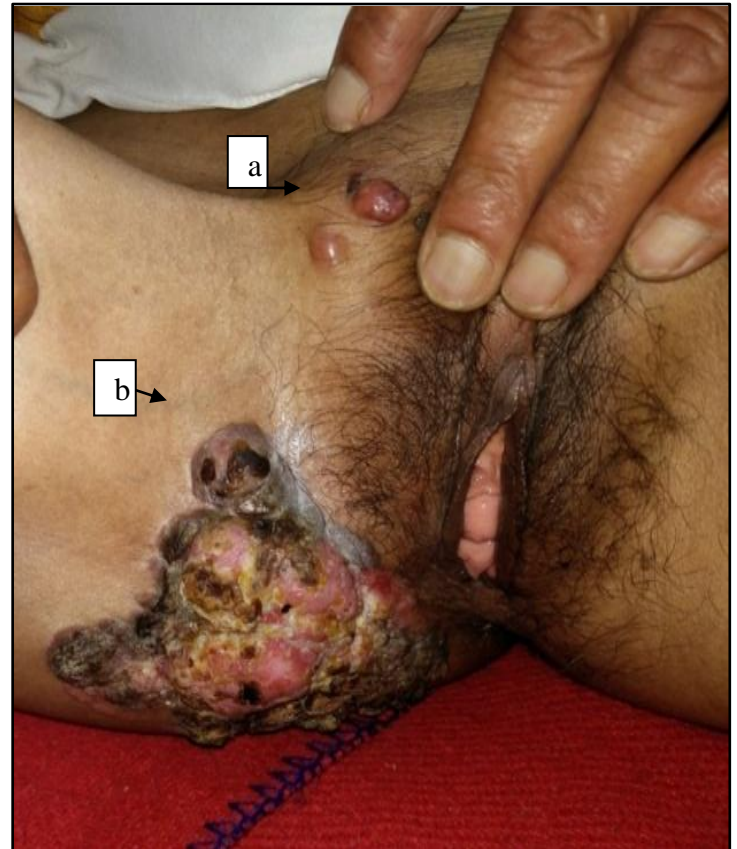
On general physical examination she was thin built elderly female with right inguinal (2x2 cm), axillary (1.5x1.5 cm) and submandibular (1x1 cm) lymphadenopathy. They were non-tender, mobile soft to firm in consistency. Otherwise there was no evidence of anemia, jaundice and raised JVP.

On mucocutaneous examination in the right upper perigenital area, there were two well-defined erythematous, firm nodules of size 1x2 cm approximately while in lower perigenital area and extending to medial thigh, there was a single well-to-ill defined erythematous, tumourous growth of size approximately 12 x 5 cm with overlying brownish crust and oozing of yellowish purulent discharge. On per-vaginal examination, hard indurated growth over cervix was observed.

Patient was investigated with clinical possibilities of squamous cell carcinoma, tuberculosis verrucosa cutis and chromoblastomycosis.

Complete blood count, biochemistry, urinalysis stool examination were within normal limits. HIV serology was non reactive. Chest X-ray and mountux test were non contributory. Fungal and mycobacterial culture [MGIT] were non-contributory. CECT chest and abdomen was done to rule out any metastasis and it was found that there was pre-aortic, para-aortic, pelvic and right inguinal lymphadenopathy, with heterogeneous and bulky cervix. (Figure.7)

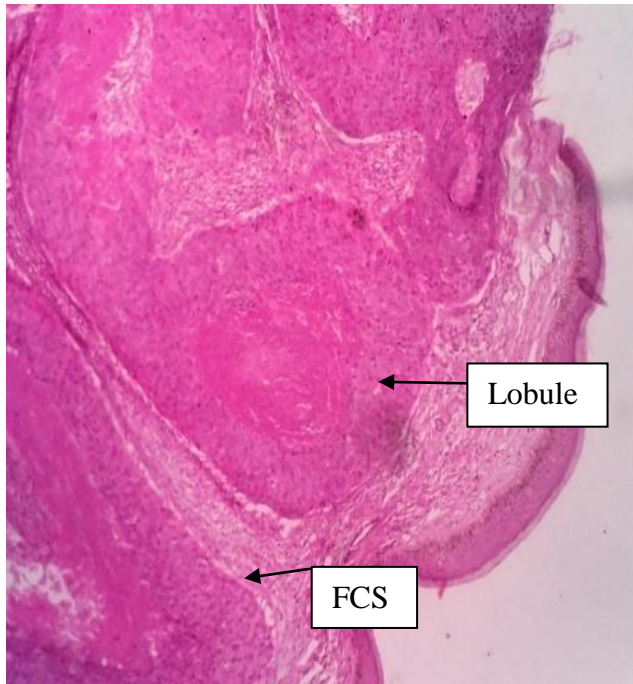
FNAC of the tumourous growth was done which demonstrate polymorphous lymphoid cell infiltrate with numerous foreign body type of multinucleated giant cells which suggested a granulomatous inflammation with no evidence of malignancy.



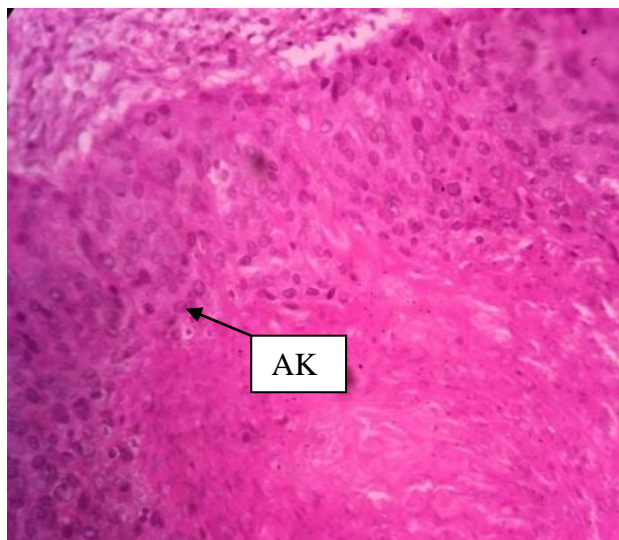
**Figure 1:** Initial two papular lesion over upper perigenital area (a) and large tumorous plaque lesion of size approximately 12 x 5 cm with overlying brownish black crust over outer border and oozing of yellowish purulent discharge at places in lower peri-genital area (b)

Two cutaneous and one mucosal biopsy were undertaken. Cutaneous samples from papulonodular lesion, tumorous plaque and mucosal cervical biopsy were sent for histopathological examination. First cutaneous biopsied lesion grossly, the nodule was firm with embedded hairs and had a margin of normal tissue all around. On histopathological examination, nodule showed a well differentiated keratinized stratified squamous epithelium with proliferation of epidermis into the dermis. In the dermis, there were interlacing bands and lobules of squamous epithelium which were well demarcated and separated by fibrocollagenous septa. These

bands were made of squamous epithelium showing abrupt keratinisation. (Figure 3) Mild atypia was seen in the lobules. These features were consistent with PTC. (Figure 2)



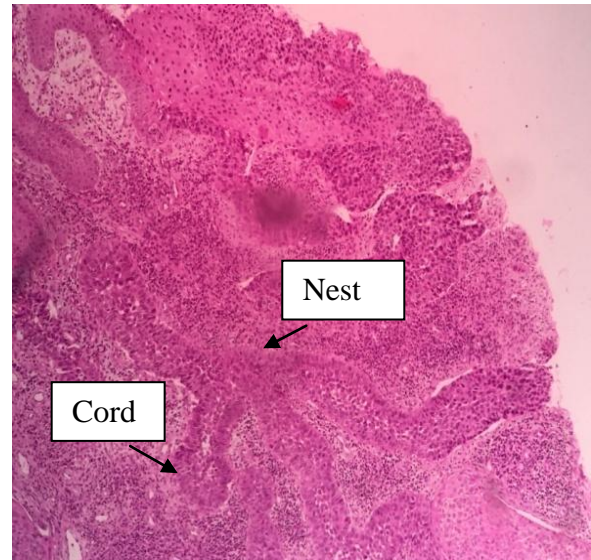
**Figure 2:** A histopathology section that shows proliferation of epidermis into the dermis. Interlacing bands and lobules of squamous epithelium well demarcated by fibrocollagenous septa (FCS).



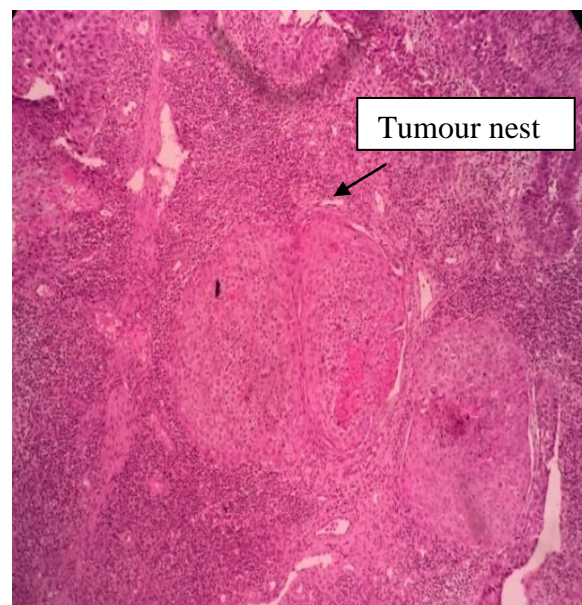
**Figure 3:** A histopathology section that shows Bands of squamous epithelium with abrupt keratinisation (AK).

Histopathological examination of tumorous growth as well as cervix showed infiltration of nest and

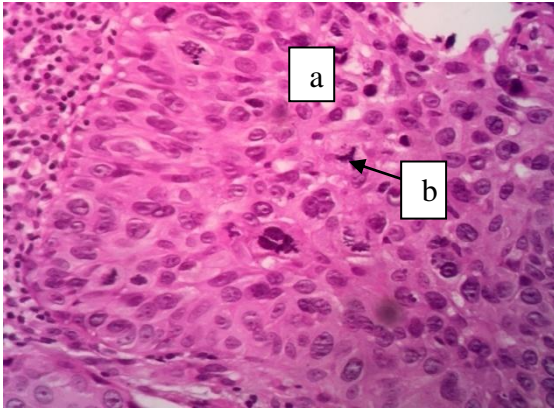
corde of squamous epithelial cells into the dermis. These tumour nests were surrounded by desmoplastic stroma and chronic inflammatory cells. Pleomorphic tumour cells revealed abnormal keratinisation with atypical mitosis. These features were consistent with moderately differentiated squamous cell carcinoma. (Figure 4, 5 and 6)



**Figure 4:** A histopathology section that shows infiltration of nest and cords of squamous epithelial cells into the dermis.



**Figure 5:** A histopathology section that shows tumour nest in the deeper dermis surrounded by desmoplastic stroma and chronic inflammatory cell infiltration



**Figure 6:** A histopathology section that shows Pleomorphic tumour cells (a) revealing abnormal keratinisation (centre) with atypical mitosis (b).



**Figure 9:** After 3 months all lesions healed



**Figure 7:** CECT showing heterogenous, bulky cervix

A final diagnosis of PTC with peri-genital SCC and cervical SCC (Stage IVB) was made and considering the poor prognosis patient was initially planned for prophylactic treatment and was given single fraction radiotherapy (SFRT) in a dose of 1000 cGy. After 3 weeks, patient was started on chemotherapy with Paclitaxel, Cisplatin and 5-FU along with radiotherapy. Significant improvement in lesions was observed (Figure 9) after 3 months of follow-up



**Figure 8:** After radiotherapy

**Discussion**

The intradermal lesions which show trichilemmal keratinisation can be the trichilemmal cyst, proliferating trichilemmal cyst and malignant proliferating trichilemmal cyst (MPTT).<sup>[2]</sup> Trichilemmal keratinisation is characterized epithelial cells, which generally lose their nuclei and keratinize without the formation of keratohyaline granules i.e abrupt transition of a nucleated epithelial cell to an anucleate, keratinized cell without the formation of a granular layer.<sup>[2]</sup> Under normal circumstances such keratinisation is observed in that part of the isthmus of the anagen hair where the outer root sheath is no longer covered by the inner root sheath. Similar keratinisation also appears in the trichilemmal sac, surrounding the lower end of catagen hair, thus producing the telogen bulb.<sup>[4]</sup>

Approximately 90% of patients with TCs present with scalp lesions which presents as a single lesion 1 to 10 cm in size, elastic firm to soft in consistency, over the scalp, although lesions up to 25 cm have been reported.<sup>[3]</sup> About 84% of the patients are greater than 50 years of age with predominance of females. PTC's tend to occur after the age of 60years<sup>[5]</sup> but they have been reported in individuals as young as 18 years.<sup>[6]</sup> Uncommonly present with ulcerated, fungating growth<sup>[1]</sup> and at atypical sites like 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.<sup>[2]</sup>

PTC is thought to originate from the trichilemmal cyst (TC) following some trauma and inflammation.<sup>[1, 3]</sup> PTC's can occur de novo. Although PTC is considered biologically benign, they may be locally aggressive with potential of recurrences and malignant transformation with metastasis to lymph nodes and internal organs.<sup>[7]</sup> A meta-analysis of 185 cases showed a local recurrence rate of 3.7%.<sup>[5]</sup> It is often difficult to differentiate between SCC and PTC. For the same reason, Ackerman and co-workers<sup>[8]</sup> called PTC a proliferating follicular cystic neoplasm (PFCN), and later they came to consider that PTC is a kind of squamous cell carcinoma and called it proliferating trichilemmal cystic squamous cell carcinoma (PTCSCC) because it displays architectural, cytopathologic, and biologic characteristics of a malignant neoplasm. Areas of malignant change are characterized by increased cellularity, atypia, mitoses, and most importantly invasion of the surrounding stroma.

Brownstein et al<sup>[3]</sup> studied 56 cases of PTC (and 50 cases of TC as comparison). In many PTCs, areas indistinguishable from uncomplicated TC were found and some uncomplicated TC contained foci of hyperplasia indistinguishable from PTC. Thus a spectrum was observed from TC with minimal hyperplasia to full blown proliferating TC. Similarities between TC and PTC, identification of intermediate stages between the two, and the occasional association of these two conditions suggest that PTC is a complication of TC; it would seem likely that trauma and inflammation induce occasional TC to proliferate.

Saida et al<sup>[9]</sup> described the three stages in the oncological development of trichilemmal tumor. Trichilemmal cyst, being the adenomatous stage, proliferating trichilemmal cyst, the epitheliomatous stage, and malignant proliferating trichilemmal tumor, the carcinomatous transformation of PTC

Cassarino et al<sup>[7]</sup> classified SCC arising from pre-existing PTC in a category of tumours with high risk behaviour i.e. tumours with high risk of malignant conversion and metastasis. Sau et al<sup>[5]</sup> reported a case series of 63 patients with PTC and observed that 10 cases had features of infiltrative

carcinoma i.e. 15.8% risk of malignant conversion. In our case also, the SCC which developed from pre-existing PTC metastasize to regional lymph nodes as was evident on CECT abdomen.

The morbidity and mortality associated with these tumors is represented by their tendency to recur, which was documented in the literature to be around 3.7%, their tendency to metastasize, the difficulty of differentiating them from squamous cell carcinoma, and the recommendations to deal with it as a low-grade malignant carcinoma.<sup>(10)</sup>

In our case PTC was present over peri-genital area which is a rare site and it underwent malignant transformation into a moderately differentiated SCC (which is even rarer) which involved the cervix as well and metastasized later to regional lymph nodes. Though it is difficult to differentiate without immunohistochemistry between MPTC and SCC, presence of co-existing cervical SCC strongly points towards transition of PTC into SCC of the cutaneous area as well as it's spread to cervix and further metastasis. If we had considered cervical SCC as the primary site with cutaneous metastasis, opinion against this view is that patient had no symptoms of cervical carcinoma as such and it was just an incidental finding.

### Conclusion

PTC should be regarded as a benign cystic squamous neoplasm, with a potential for recurrences and progression to SCC. Hence careful close clinical follow up with histological examination is necessary to detect focal areas of malignancy. Patients with histopathological diagnosis of PTC should be monitored for recurrences after excision, malignant changes or metastasis.

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