

Rare case of a Malignant Proliferating Trichilemmal Tumor of Vulva: A Case Report

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Abstract: Proliferating Trichilemmal tumors (PTT) are rare neoplasms of the external root hair sheath that are largely benign. The potential for PTT to undergo transformation to a Malignant Proliferating Trichilemmal Tumor (MPTT) is very rare. In this case report we discuss the diagnosis, histo-pathological characteristics, and surgical treatment of such tumors.

Keywords: Malignant Proliferating Trichilemmal tumor; Proliferating Trichilemmal tumor; Squamous cell carcinoma

1. Introduction

Proliferating Trichilemmal tumors (PTT), also known as proliferating pilar tumors, are rare neoplasms of the external root hair sheath that are largely benign, cystic in nature, and characterized as containing trichilemmal keratin. These rare exophytic tumors are mainly confined to the scalp and back of the neck and most often reported in middle age females [1]. We here by present a rare case of a 83 year-old female patient with a rapidly growing swelling on the vulva initially suspected as a squamous cell carcinoma and later classified as malignant proliferating trichilemmal tumor.

2. Case Report

A 83 year-old female presented with a swelling on the vulva. The painless swelling was present since 1 month with uv prolapse. The patient was otherwise healthy with no significant past medical history. Patient was menopausal since 34 years. No history of post menopausal bleeding was present. On examination UV prolapse was present with Extension of growth reaching upto lower 1/3^r of vagina on right side along with Cystocele and rectocele. Grossly we received an already cut open specimen measuring 3.5x2x1cm covered by skin. Fleshy in appearance with irregular outlines reaching upto the epidermis. Cut surface shows grey white areas and Focal haemorrhagic areas. (Figure 1)



Figure 1: Specimen measuring 3.5x2x1cm

Histopathological examination showed tissue fragments covered over by stratified squamous epithelium revealing Hyperkeratosis, focal parakeratosis, variable moderate acanthosis, neutrophil exocytosis.

The dermis reveals a circumscribed nodular lesion comprised of closely packed lobules of tumour cells separated by thin fibrovascular septae. The tumour cells have moderately pleomorphic round to oval nuclei with fine chromatin, variably prominent nucleolus and moderate amount of eosinophilic, focally vacuolated cytoplasm with scattered mitoses and focal dyskeratosis. (Figure 2) Periphery of tumour nodules show intersecting cords and nests of epithelial cells with focal peripheral palisading. (Figure 3)

Few scattered multinucleated tumour giant cells, some of the lobules reveal abrupt central keratinization with keratinized spindle cells in the center of the lobule.

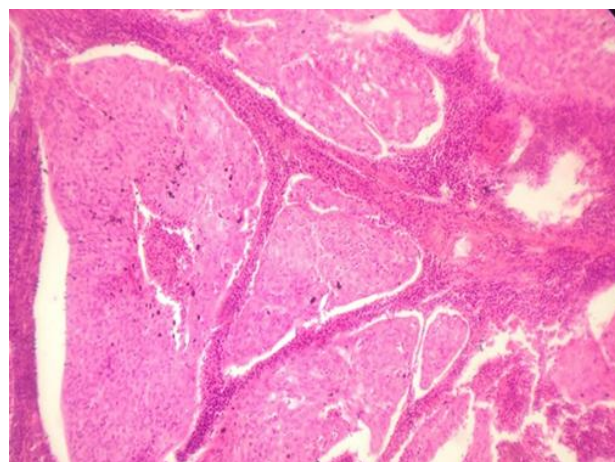


Figure 2: Dermis and subcutaneous tissue showing cellular tumor comprised of lobules of squamous cells with focal dyskeratosis (H and E stain, ×400)

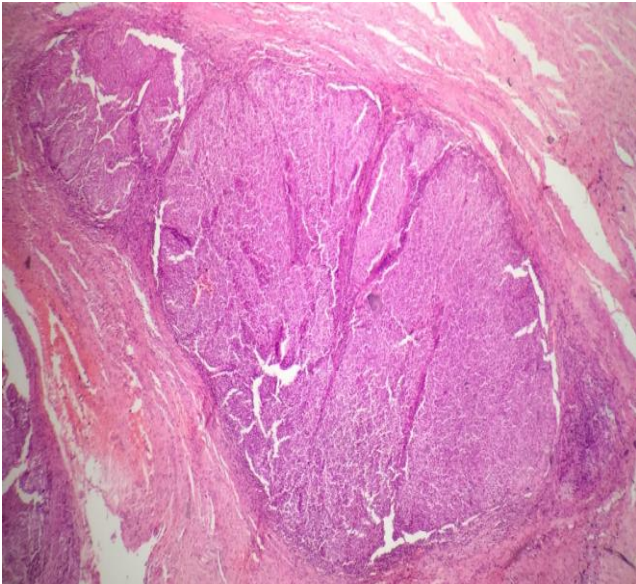


Figure 3: Periphery of tumour nodules with intersecting cords and nests of epithelial cells with focal peripheral palisading (H and E stain, $\times 100$)

We could not find the recent status of the patient as she was didn't show up for follow-up.

3. Discussion

Proliferating trichilemmal tumors (PTT) of the vulva are relatively uncommon. These tumors were first named as proliferating epidermoid cysts by Wilson Jones in 1966² and have been commonly misinterpreted both clinically and histologically as squamous cell carcinomas. They are five times more likely to occur in females and have a median age of occurrence of 65-year-old³. Although a predominantly benign condition, PTT has been reported to undergo a malignant transformation. The potential for PTT to undergo malignant transformation is unknown and is an exceedingly rare occurrence.

Histological examination of malignant proliferating trichilemmal tumors reveal a well-circumscribed tumor composed of a proliferation of malignant - appearing squamous cells in which there is extensive areas of amorphous keratinization. The tumor cells overall are not well differentiated with mitotic figures scattered throughout and small areas of calcifications present. Grossly, MPTT usually presents as a mobile, firm, smooth and rounded nodule. Larger lesions may be lobular and multiple cysts are commonly found. Features which favored malignancy and a change of our initial diagnosis of squamous cell carcinoma included the large size, location, infiltrative borders, abrupt central keratinization, nuclear pleomorphism, rapid change in growth, and increased mitoses.

The differential diagnoses of malignant proliferating trichilemmal tumors includes; squamous cell carcinoma, trichilemmal carcinoma, proliferating trichilemmal tumors, sebaceous cyst, and angiosarcoma. It has also been stated that the tumor has tendency to recur and metastasize more frequently than squamous cell carcinoma⁴. It is essential to properly diagnose due to the differences in the course and aggressiveness, the likelihood of recurrence and metastases,

and the difference in recommended management of these differentials.⁵

The current recommended management of malignant proliferating trichilemmal tumors is local excision with margins of normal tissue and a work-up to exclude metastases along with routine long-term follow-up⁵.

4. Conclusion

Proliferating trichilemmal tumor (PTT) is a benign tumor originating from the outer root sheath of a hair follicle. Malignant transformation in case of PTT is very rare and unusual finding, pose a diagnostic dilemma for the pathologist. It is usually confused with squamous cell carcinoma both sharing many common features. So the identification of malignant PTT is very essential. Wide surgical excision should be considered as the primary modality of treatment while alternative therapies require further evaluation.

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