Malignant Proliferating Trichilemmal Tumour: A Case Report

Abstract
Proliferating trichilemmal tumour (PTT) is an uncommon neoplasm that has been derived from the outer root sheath of hair follicle. We describe a case of a 73 year old female who presented with solitary friable nodular mass on scalp measuring 3.5x2x1cm. She underwent local excision of this growth. Histologically, tumour shows trichilemmal keratinisation without interposed granular layer which was characterized by poor circumscription with invasion of surrounding tissue with marked nuclear atypia, hyperchromasia and atypical mitotic activity.

Introduction
Proliferating trichilemmal tumour (PTT) is a rare skin neoplasm. It was first described as "proliferating epidermoid cyst" by Wilson and Jones in 1966 [1]. These lesions are the cutaneous neoplasms that are derived from the outer root sheath of the hair follicles [2]. PTT present as solitary nodules on scalp in elderly women. In rare instances, malignant transformation of proliferating trichilemmal cyst takes place [3-5]. This is indicated clinically by rapid enlargement of the nodule. Histopathologically, tumour shows trichilemmal keratinisation without interposed granular layer, invasion associated with nuclear atypia, giant cell nuclei and tumour necrosis that indicate malignancy which is described as malignant proliferating trichilemmal tumour (MPTT) [6].

Case Report
A 73 year old female presented to the hospital with a solitary friable nodular mass on scalp measuring 3.5x2x1cm. She underwent local excision of this growth. Grossly, specimen consists of single piece of partly skin covered friable nodular mass 3.5x2x1cm. Histologically, tumour shows trichilemmal keratinisation without interposed granular layer which was characterized by poor circumscription with invasion of surrounding tissue with marked nuclear atypia, hyperchromasia and atypical mitotic activity (Figure 1 & 2).

Figure 1: Tumour islands of pleomorphic squamous cells with keratin pearls in dermis (X40).

Figure 2: Trichilemmal keratinisation without interposed granular layer (X40).
Discussion

PTT are the rare cutaneous neoplasm that occurs as solitary nodular mass which are most commonly found on scalp of the elderly women [4]. The other less common sites are neck, trunk, armpits, groin, lower and upper limbs, upper lip and buttocks [4,7,8]. Malignant change in trichilemmal cyst is rare. These tumours develop in the course of time from the foci of proliferating epithelial cells in the trichilemmal cysts because of factors such as trauma or chronic inflammation [9]. Histologically, malignancy is characterized by poor circumscription with invasion of surrounding tissue with marked nuclear atypia, hyperchromasia and atypical mitotic activity[10]. Different reports have mentioned local or lymph node spread but hematogenous spread has been rarely reported [11-15].

Squamous cell carcinoma(SCC) should be considered as differential diagnosis but trichilemmal keratinisation is important to differentiate SCC from MPTT [2,4]. Immunohistochemistry such as Ki67, p53 and CD34 plays a significant role for the diagnosis and differential diagnosis of PTTs and morphological characteristics . Ki 67 and p53 were used when MPTT cases were compared with PTT [2,16,17]. CD34 is an important marker to differentiate SCC from MPTT [2]. The higher risk of metastasis is present when the lesion occurs in areas other than scalp, when it is fast growing and infiltrative, is over five centimeters in diameter and shows cytological atypia and mitotic activity[18]. The treatment of choice is surgical resection. However, due to local recurrence rate, Mohs micrographic surgery would be a therapeutic option [19].

Conclusion

MPTT appears as diagnostic dilemma for the surgeon as well to the pathologists. It is crucial to distinguish MPTT from SCC due to similar morphological characteristic which may affect the treatment approach for the patient. Hence, immunohistochemistry is useful in differential diagnosis and determining morphological characteristics.

References
