Malignant Proliferating Trichilemmal Tumour of the Scalp(MPTT)

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ABSTRACT

Malignant proliferating trichilemmal tumour also known as malignant proliferating pilar tumour is a very rare malignant hair follicle tumour , commonly occurring on the scalp, eyelids, neck and face of elderly women. The exact cause is unknown ,but most commonly starts as a benign trichilemmal cyst and then progressed to a proliferating trichilemmal cyst before becoming malignant.These tumours are largely benign, often cystic and are characterized by trichilemmal keratinization. However at times ,the tumour has an aggressive clinical course and a propensity for distant metastasis.We report a case of a 70 year old female who presented with an swelling on the scalp since 4 years.Anexcional biopsy was done with adequate margins and revealed a malignant proliferating trichilemmal tumour of the scalp with positive margins.A medical oncologist opinion was sort for adjuvant therapy who suggested no further treatment inview of the multiple co-morbities,old age, and patient's refusal for further treatment . The patient was investigated for any local and distant metastasis ,which were negative.To the best of our knowledge, malignant proliferating trichilemmal tumour of the scalp is a rare condition and only a handful of cases are reported till date .We report a case of malignant proliferating trichilemmal tumour of the scalp without metastasis with relevant discussion on the same.

Key words: *Malignant proliferative trichilemmal tumour; scalp; proliferating pilar cyst; trichilemmal keratinization; metastasis.*

INTRODUCTION

It is a rare cutaneous tumour predominantly affecting the scalp, eyelids, neck and face of elderly women[1].It is an unusual neoplasm originating from the outer root sheath of hair follicles[1].It was first described by Jones in 1996 using the term "proliferating epidermoid cyst"[2].Their classical histological finding is sudden compact amorphous keratinization of the epithelial cells that cover the cyst wall without a granular layer,this phenomenon is *called trichilemmal keratinizatio*n[1]. Proliferating trichilemmal tumour commonly exhibit benign behaviour and rarely present a malignant course,when they invade the neighbouring structures accompanied by anaplasia and necrosis,it is described as a malignant proliferating trichilemmal tumour (MPTT)[1]. Malignant proliferating trichilemmaltumor are invasive and metastatic tumours that demonstrate biologically aggressive behaviour[3]. Squamous cell carcinoma (SCC) should be eliminated by a differential diagnosis due to similarity of disease presentation[1]. The management of malignant proliferating trichilemmal tumour is controversial because only a limited number of cases are reported in literature. We report a case of malignant proliferating trichilemmal tumour without metastasis.

CASE REPORT

A 70 year old female presented with a scalp swelling since 4 years, which was insidious in onset and rapidly progressed in size since the last 6 months with ulceration at the apex of the swelling.On physical examination ,there was a single oval swelling measuring 4.5 cms X 4.5 cms on the scalp. Swelling was non-tender, mobile, nodular with variable consistency. An

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excional biopsy was done with adequate margins and revealed a malignant proliferating trichilemmal tumour of the scalp with positive margins.A medical oncologist opinion was sort for adjuvant therapy who suggested no further treatment inview of the multiple co-morbities,old age, and patient's refusal for further treatment . Metastatic work –up was negative in view of this tumour having a propensity for local and distant metastasis. We report a case of Malignant proliferating trichilemmaltumor without metastasis.

LEGENDS OF ILLUSTRATION

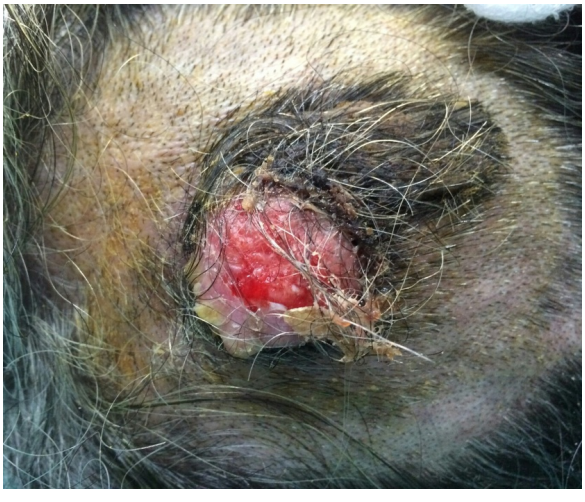

FIGURE 1-A clinical picture of a malignant trichilemmal tumour of the scalp (MPTT).

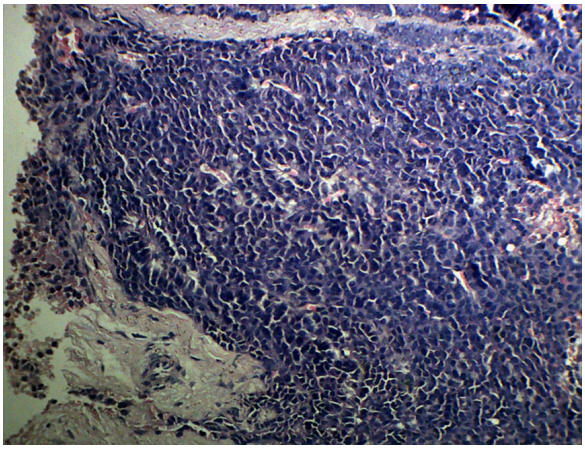

FIGURE 2- Microscopy shows tumour cells arranged in sheets or cords with areas of keratinization. Tumour cells have scanty amount of cytoplasm,enlarged round to oval nucleus which are hyperchromatic containing coarse chromatin.

DISCUSSION

It is a rare cutaneous tumour occurring on the face,scalp,eyelids and neck of elderly women as in our case. Grossly, MPTTS can be exophytic,ulcerative,polypoid or nodular lesions[1].Histologically ,a MPTT shows severe nuclear atypia, marked cellular pleomorphism with atypical mitosis, dyskeratotic cells and infiltrating margins[1].The differential diagnosis of a MPTT include basal cell carcinoma, sebaceous carcinoma, squamous cell carcinoma, clear cell hydraadenocarcinoma[1].Due to inconsistencies in nomenclature and misclassification as squamous cell carcinoma,the real incidence of MPTT is unknown[1].Malignant transformation of a trichilemmal tumour occurs in a stepwise manner starting with an adenomatous stage of PTT evolving into a carcinomatous stage of MPTT[1,2].The treatment of choice for MPTT is surgery with periodic surveillance without adjuvant therapy[1].The treatment of choice for MPTT without metastasis is wide local excision with 1 cm of normal tissue as we did in our case,however a MPTT with metastasis ,the treatment of choice is palliative chemotherapy(cisplatin and 5-fluoro-uracil) followed by palliative radiotherapy however it carries a very poor prognosis[1,3].Clinically ,enlargement of a scalp lesion and histological evidence of significant mitosis,marked cellular pleomorphism,infiltrating margins and aneuploidy suggests a malignant transformation[1] .In our case possibly reflects a malignant transformation in a proliferating trichilemmal tumour.

CONCLUSION

To conclude,MPTT is a rare clnical entity with under 50 cases reported in literature.the treatment of a MPTT without metastasis as of now is wide local excision with 1 cm of normal tissue without adjuvant therapy.under periodic surveillance MPTT follows an aggressive clinical course hence it is important to distinguish from other similar looking neoplasms.

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Be more concerned with your character than with your reputation. Your character is what you really are while your reputation is merely what others think you are.

~ Dale Carnegie