CASE REPORT

Malignant Proliferating Trichilemmal Tumour of the Scalp with Intra-Cranial Extension and Lung Metastasis—a Case Report

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Abstract Malignant proliferating trichilemmal tumour (MPTT) is a rare cutaneous tumour predominantly affecting the scalp, eyelids, neck and face of elderly women. It is a large, solitary, multilobulated lesion that may arise within a pilar cyst. These tumours are largely benign, often cystic and are characterized by trichilemmal keratinisation. However at times, the tumour has an aggressive clinical course and a propensity for distant metastasis. We report a case of a 29-year-old male who presented with a scalp swelling. Histology revealed malignant proliferating trichilemmal tumour of scalp. CT thorax showed multiple nodules in both lungs suggestive of lung metastasis. The patient underwent palliative radiotherapy and chemotherapy and expired 2 months after palliation. To the best of our knowledge, malignant proliferating trichilemmal tumour of the scalp with intra-cranial extension and lung metastasis 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 with intra-cranial extension and lung metastasis with relevant discussion at the same time.

Keywords Malignant proliferating trichilemmal tumour · Scalp · Lung metastasis · Proliferating pilar cyst · Intra-cranial extension

Abbreviations

MPTT Malignant proliferating trichilemmal tumour
PTT Proliferating trichilemmal tumour

Introduction

Proliferating trichilemmal tumours (PTTs) were first described by Jones in 1996 using the term “proliferating epidermoid cyst” [1]. These tumours are rare cutaneous neoplasms derived from the outer root sheath of the hair follicle [2]. Their characteristic histological finding is the sudden compact amorphous keratinisation of the epithelial cells that cover the cyst wall without a granular layer, and this phenomenon is called trichilemmal keratinisation. PTTs commonly exhibit benign behaviour and rarely present a malignant course. When they invade neighbouring tissues accompanied by anaplasia and necrosis, it is described as a malignant proliferating trichilemmal tumour (MPTT) [2, 3]. MPTTs are invasive and metastatic tumours that demonstrate biologically aggressive behaviour [3]. Squamous cell carcinoma (SCC) should be eliminated by a differential diagnosis due to the similarity of disease presentation [3]. Treatment of MPTTs is controversial because only a limited number of cases are reported in literature. We report a case of malignant proliferating trichilemmal tumour with intra-cranial extension and lung metastases in a 29-year-old male (Figs. 1 and 2).

Case Report

A 29-year-old male presented with a scalp swelling since 7 years, which was insidious in onset and rapidly

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Fig. 1 a A clinical picture of malignant trichilemmal tumour of the scalp. **b** CT of the head revealed a large soft tissue swelling in the left frontoparietal region with destructive and aggressive bony changes and intra-cranial extension suggestive of neoplastic aetiology

increased in size since 1 year with no other symptoms. On physical examination, there was a single irregular swelling measuring 30×20 cm on the left side of scalp. Swelling was non-tender, fixed and nodular with variable consistency. Trucut biopsy was suggestive of malignant proliferating trichilemmal tumour of scalp. X-ray skull was reported as a large lobulated radiopaque soft tissue swelling of the scalp over the left frontoparietal region, and intra-cranial extension cannot be commented upon advised CT of the head. CT of the head revealed a large soft tissue swelling in the left frontoparietal region with destructive and aggressive bony changes and intra-cranial extension suggestive of neoplastic aetiology. Chest X-ray showed multiple cannon ball opacities noted in both the lungs suggestive of metastases to the lungs. CECT thorax revealed multiple tiny soft tissue nodules in both lungs suggestive of lung metastases. CECT of the abdomen and pelvis was reported as normal. We report a case of malignant proliferating trichilemmal tumour of the scalp with intra-cranial extension and lung metastasis. Patient underwent palliative radiotherapy followed by chemotherapy (6 cycles of

cisplatin and 5-fluorouracil) and expired 2 months after palliation. We report this case because of its rarity.

Discussion

A MPTT is an unusual neoplasm originating from the outer root sheath of hair follicle [4]. The common sites include scalp, forehead and neck. Women over 40 years are most commonly affected [4]. Grossly, the tumours have been described as exophytic, ulcerative, polypoid or nodular lesions that may be keratotic. Histologically, a MPTT shows a severe nuclear atypia, marked cellular pleomorphism with atypical mitosis, dyskeratotic cells and infiltrating margins. A trichilemmal tumour has a more indolent clinical course. It can rarely undergo a malignant transformation in a stepwise manner starting with an adenomatous stage of PTT to an epitheliomatous stage of PTT evolving into a carcinomatous stage of MPTT. MPTT usually arise from a pre-existing trichilemmal cyst. MPTTs have a more aggressive and biological behaviour, and these lesions present a significant morbidity and potential mortality [4]. This tumour lacks a distinctive histological or immunohistochemical marker to suggest malignant transformation [4]. The real incidence of MPTT is unknown due to its rarity and also because of its inconsistencies in nomenclature and misclassification as squamous cell carcinoma [2, 3]. The treatment of choice for MPTT is surgery with periodic surveillance without adjuvant therapy [4]. The differential diagnosis of MPTT includes basal cell carcinoma, sebaceous carcinoma, squamous cell carcinoma and clear cell hydradenocarcinoma [5]. The treatment of MPTT without metastasis is wide local excision with 1 cm of normal tissue [2, 3]. In our case, MPTT with intra-cranial extension and lung metastasis carries a very poor prognosis. Palliative chemotherapy (cisplatin and 5-fluorouracil) followed by palliative radiotherapy is

Fig. 2 Microscopy shows tumour cells arranged in sheets, cords and clusters with areas of keratinisation seen. Tumour cells have scanty amount of cytoplasm, enlarged round to oval nuclei which are hyperchromatic containing coarse chromatin

the treatment of MPTT with distant metastasis. Clinically, the enlargement of a long-standing nodular scalp lesion and histological evidence of significant abnormal mitosis, marked cellular pleomorphism, infiltrating margins and aneuploidy reflect a malignant transformation. Our case possibly represents the event of malignant transformation in a proliferating trichilemmal tumour.

Conclusion

To conclude, MPTT with intra-cranial extension and lung metastasis is presented because of its rarity. MPTT can occur in young people especially in an individual with pre-existing PTT. Since it follows an aggressive course, it is essential to distinguish it from other similar looking neoplasms. MPTT with intra cranial extension and lung metastasis is a rare entity and carries a poor prognosis.

Compliance with Ethical Standards

Conflict of Interest The authors declare that they have no competing interests.

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