

# Malignant proliferating trichilemmal tumour of the scalp.



Ann. Ital. Chir., 2006; 77: 179-182

## Report of a case and a short review of the literature

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## Malignant proliferating trichilemmal tumour of the scalp. Report of a case and review of the literature

**BACKGROUND:** Malignant proliferating trichilemmal tumour is a rare skin tumour that originates from the root sheath of the hair and usually arises in the sun-exposed areas of elderly women. It mimicks poorly differential squamous cell carcinoma and its biological behaviour is unpredictable, because rarely can produce distant metastases.

**CASE REPORT:** The Authors report on a case of a 54-year-old female patient who proceeded with a cystic lesion in the scalp. The lesion was removed with wide excision. The histological examination of the lesion revealed a malignant proliferating trichilemmal tumour. Two years after the excision the patient is free of disease.

**DISCUSSION:** Only few cases (about seventy) of malignant proliferating trichilemmal tumours have been reported in the Bibliography, in some of the patients with malignancy, the tumour produces distant metastases. In the reported case, although the high risk of distant metastasis, two years after the tumour resection the patient is free of disease. It is also reported a short review of the literature.

**CONCLUSIONS:** Malignant proliferating trichilemmal tumours are rare malignant lesions that affect mainly older women. The appropriate treatment includes wide resection and close postoperative follow-up of the patient to facilitate the early diagnosis of distant metastases.

**KEY WORDS:** Malignant proliferating trichilemmal tumour, Skin tumours.

## Introduction

Trichilemmal cysts are benign lesions that usually located in the scalp of elderly people. Malignant proliferating trichilemmal tumours are atypical clear cell neoplasms of the adnexal keratocytes adjacent to epidermis or to follicular epithelium, which are associated with skin exposure to the sun. They derived from the outer root sheath of the hair follicle and may simulate a squamous cell carcinoma<sup>1-3</sup>.

Less than seventy cases of malignant proliferating tumours have been reported in the bibliography<sup>4</sup>. The diagnosis of the malignancy needs to be established histologically. The biological behaviour of the tumour is unknown because malignant trichilemmal tumours can rarely

produce metastases. Close follow-up is essential for the early diagnosis of tumour metastases<sup>5,6</sup>.

Malignant proliferating trichilemmal tumour represent slow-growing lesions and most cases arise on existing benign trichilemmal cyst. The cysts are solitary in 30% and multiple in 70% of cases and present a familial pattern in 75% of the Caucasian people<sup>7,8</sup>.

## Case report

A 54 years-old female farmer presented with a 3 years history of a cystic nodule in the scalp. The patient was a farmer, and thus exposed for a long time in the sun. Clinically the nodule was dome-shaped, elastic hard in consistency, with no surface abnormalities. The diameter of the lesions was 2x3 cm, and it was situated in the left forehead about 1 cm above the frontal hairline. Although initially clinical diagnosis suggested benign lesion, probably sebaceous cyst, the lesion was widely resected. The histological examination of the specimen

Pervenuto in Redazione Marzo 2005. Accettato per la pubblicazione Luglio 2005.

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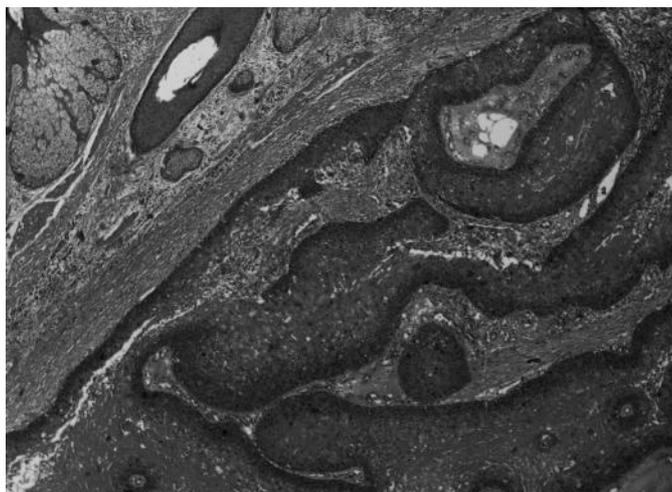


Fig. 1: Photo of the histological examination of the specimen, which indicates proliferating malignant trichilemmal tumour of the scalp. The margins of the excision were free of disease.

suggested malignant proliferating trichilemmal tumour (Fig. 1). The margins of excision was clear. The lesion was a deeply invasive lobular tumour in continuity with the epidermis, part of which was cystic and filled with keratin. It was encased in cystic wall, which was composed of clear atypical cells with moderate differentiations and multiple mitoses. In some areas micro-invasions of the dermis lymphatic plexus were identified (Fig. 2). The malignant cells were strongly positive in p53 stains (3+) and in proliferation index factor ki-67. The existence of micro-invasions of the dermis suggested increases risk if distant metastases development, and the patient set in close follow-up. Computed Tomography

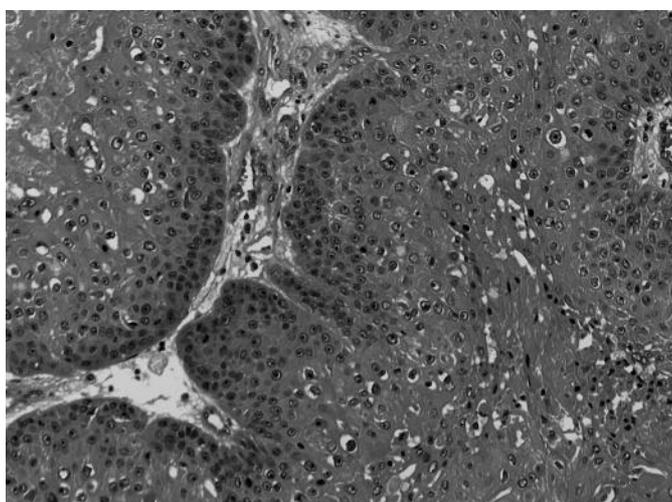


Fig. 2: The trichilemmal tumour was a deeply invasive lobular tumour in continuity with the epidermis, part of which was cystic and filled with keratin. The lesion was encased in cystic wall, which was composed of clear atypical cells with moderate differentiation and multiple mitoses.

of the head-neck and the chest failed to reveal distant metastases. Two years after the initial treatment the patients remains free of disease. The follow-up continue for three more years in annual basis.

## Discussion

Malignant proliferating trichilemmal tumour is a rare cutaneous carcinoma. It primarily affects elderly women and arises in skin sites that are exposed to sun. Most common sites occurrence of the tumour are the forehead, the scalp, the neck, and the upper thorax. Rarely the tumour can arise in the upper and lower extremities <sup>9,10</sup>. In this case the tumour occurred in a woman 54 years-old, in the scalp above the forehead. The woman worked as farmer and is exposed for a long time to the sun. Although the clinical presentation of malignant proliferating trichilemmal tumour varies, usually it appears as a solitary solid or cystic lesion, nodular, ulcerative or as keratotic plaque. It is difficult to establish the diagnosis clinically, and the tumour can be misdiagnosed as basal cell carcinoma, malignant melanoma, squamous cell carcinoma, and sebaceous or trichilemmal cyst <sup>11</sup>. The colour of the lesion can also varies. Some lesions have normal skin colour like our patient, while others are pigmented, brown or black like melanoma.

The description of the histological characteristics of malignant proliferating trichilemmal tumour belongs to Headington <sup>2</sup>. Atypical cells of moderate differentiation resembling the cells of the outer root of the hair sheath, which also present peripheral palisading. The malignant cells are characterized by high mitotic rate, atypical mitoses, nuclear polymorphism, invasion of the adjacent tissues or epidermis, and foci of keratinization <sup>1,12</sup>. Plump et al report on several cases of malignant proliferating trichilemmal tumour that did not express keratin. In our case the tumour expressed strongly keratin (3+) <sup>8</sup>.

The pathogenesis of malignant proliferating trichilemmal tumour still remains mysterious and poorly understood. Many Authors though that is based mainly in the distribution of the lesions and suggest that exposure in the sun light may possess a significant role in the pathogenesis of the disease <sup>4</sup>.

The biological behaviour of the tumour and the clinical course of the patient are unpredictable. Although in most of cases the tumour didn't produces metastases. Several other cases with distant metastases have been reported. Wide resection of the tumour and close postoperative follow-up for several years is essential to ensure the absence of distant tumour metastases. In cases with incomplete resection where the margins of the excision are involved, additional resection in clear margins is indicated <sup>3,9</sup>. Although the clinical diagnosis in our patient suggested benign cystic lesion we removed it with wide resection, thus no additional intervention was neces-

sary. The histological examination of the specimen revealed multiple micro-invasions of the epidermis lymphatic plexus, which increases the potential risk for late distant metastases. Two years after the initial treatment our patient remains free of disease.

### Riassunto

**INTRODUZIONE:** Il trichilemma maligno proliferante è un raro caso di tumore cutaneo, che origina di solito dal bulbo del follicolo capillare ed abitualmente si riscontra nelle aree del cuoio capelluto esposte al sole, e più facilmente nelle donne. D'età avanzata. Simula il carcinoma a cellule squamose a basso grado di differenziazione ed il suo comportamento biologico si potrebbe definire imprevedibile, in quanto, anche se raramente, può dare delle metastasi a distanza.

**CASO CLINICO:** Viene presentato il caso di una donna 54enne che venne a controllo per una lesione cistica del cuoio capelluto. La lesione venne asportata chirurgicamente, usando larghi margini di escissione. L'esame istologico della lesione asportata rivelò un trichilemma maligno proliferante. La paziente dopo due anni dall'escissione, risulta libera da malattia.

**DISCUSSIONE:** Sono riportati in letteratura solamente alcuni casi (circa 70) di trichilemma maligno proliferante. In alcuni dei pazienti portatori della forma a basso grado di differenziazione, il tumore ha prodotto delle metastasi a distanza. Nel caso studiato, nonostante l'elevato rischio per la presenza di metastasi a distanza, la paziente a due anni distanza risulta libera di malattia.

Viene anche riportata una breve rassegna della bibliografia internazionale correlata a tale raro tipo di tumore.

**CONCLUSIONI:** Il trichilemma maligno proliferante è un raro tumore cutaneo che colpisce prevalentemente le donne d'età avanzata. L'appropriato tipo di trattamento per tale tipo di tumore include da una parte una radicale escissione chirurgica della lesione e, all'altra, uno

stretto e continuo follow-up del paziente nel postoperatorio al fine di poter scoprire, precocemente e tempestivamente, possibile metastasi a distanza del tumore.

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