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Case Study

Proliferating Trichilemmal Tumor: A Rare Tumor of Unpredictable Evolution

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Abstract

The trichilemmic cyst (TC), also known as the hair cyst, is a rare adnexal tumor, developed from the isthmic portion of the hair follicle. More aggressive forms, such as proliferative trichilemmal tumors (PTT), and malignant proliferative trichilemmal tumors (MPTT) can occur, but they are much less common. PTT is generally benign, some cases with malignant transformation are described in the literature. Therefore the criteria of malignancy (clinical and histological) have been widely discussed. The real challenge is to distinguish the benign form from the malignant one.

Herein, we report the case of a 71-year-old patient who presented a scalp parietal tumor that had been evolving for 19 years, whose histological study was related to a proliferating trichilemic tumor, without sign of malignancy.

Keywords: Proliferative Trichilemmal Tumor; Trichilemmic Cyst; Malignant; Aggressive

Introduction

Trichilemmal cyst (TC), also known as pilar cyst, is a rare keratin-filled adnexal tumor developed from the isthmic portion of the pilar follicle [1]. These cyst can affect up to 10% of the population. Other more aggressive forms, such as proliferative trichilemmal tumors (PTT) and malignant proliferative trichilemmal tumors (MPTT), can also occur, but are much less frequent [2].

The current hypothesis considers that there is a "continuum" spectrum of simple trichilemmal cysts followed by benign PTT and then low-grade and high-grade malignant PTT. The real challenge is to distinguish the benign from the malignant form [3]. We report a case of a 71-year-old patient with a scalp tumor evolving since 19 years corresponding to a proliferating trichilemmal tumor.

Observation

It was a 71 year old patient, chronic smoker weaned 30 years ago, without any similar case in the family. He had consulted for a scalp tumor evolving since 19 years. The clinical examination revealed a patient in good general condition, with a polylobed tumor, well limited, flesh colored, dotted with fine telangiectasias, with alopecia, without ulcerations or necrosis zone, measuring 9/7 cm, painless, with firm consistency, mobile with respect to the deep plane, located in the parietal region of the scalp (Figure 1). The examination of the lymph nodes was normal. The cerebral CT scan showed a parietal scalp tumor respecting the bones, enhancing after injection of PDC, with a mixed component, liquid, tissue with calcifications, without involvement of the brain parenchyma (Figure 2). Surgical removal of the cyst without deterioration was performed (Figure 3). The histological study of the excisional speci-

men showed a proliferating trichilemmal cyst or Wilson Jones tumor, with no sign of malignancy. The postoperative course was simple. The patient was subsequently seen in consultation without local recurrence after 3 months.

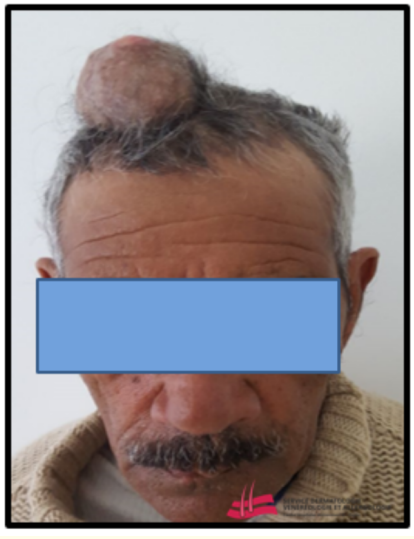

Figure 1: Well limited parietal tumor, flesh colored, dotted, with alopecia, without ulcerations or necrosis zone, measuring 9/7 cm.

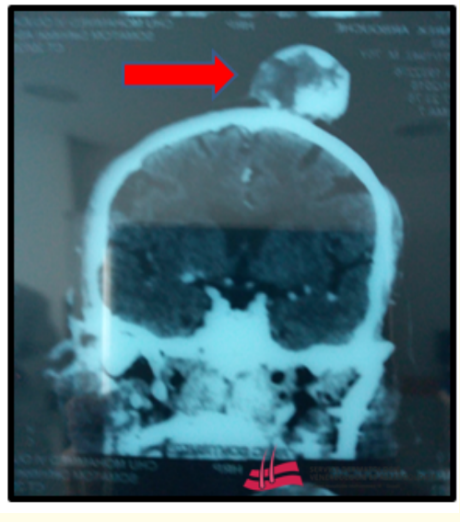

Figure 2: Mixed tumor containing liquid, tissue and calcifications, without involvement of the bone or brain parenchyma.

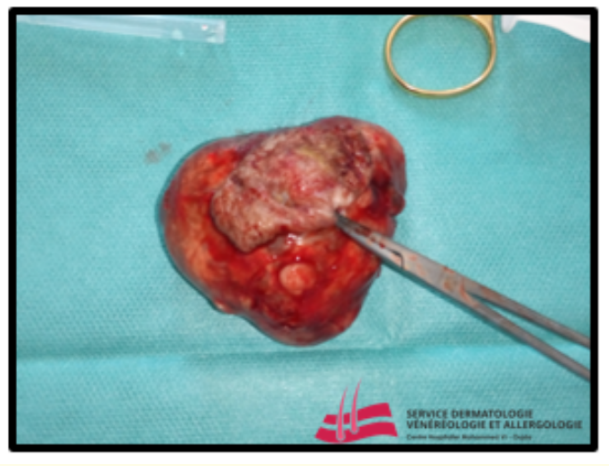

Figure 3: Tumor removed without deterioration.

Discussion

PTT was first described by Wilson Jones in 1966 as an entity with the histologic ability to mimic squamous cell carcinoma [4]. A female predominance has been reported, with a sex ratio F/M: 2.5 [1] and a mean age of onset at 60 years [2]. It is thought to arise from pre-existing TC following inflammation or previous trauma [2]. A genetic component has been reported in the literature [4].

Clinically it is often solitary [5], paradoxically multiple lesions in the same patient have been reported in the literature. It affects the scalp in 90%, rarely the face, ear, upper limbs, genital region, hips and lower limbs [6]. This tumor is not visible in the bald scalp [4]. It usually presents as a round, smooth, painless, slow-growing mass of variable size ranging from 0.2-10 cm. Alopecia on the lesion and ulceration can be detected [1].

Histologically, PTT are characterized by the presence of trichilemmal keratinization [2]. Several authors have proposed a hypothetical evolution of TC in three stages: first adenomatous, then epitheliomatous, and finally carcinomatous [3]. MPTT are very rare and have an invasive component with the classic appearance of trichilemmal keratinization. The criteria for malignancy have been widely discussed in the literature. For most authors, only the presence of stromal infiltration confirms malignancy [3]. A clinicopathological study of 76 cases divided PTT into three groups, on the degree of stromal invasion and the level of cellular atypia, benign, low-grade and high-grade malignancy [4].

Immunohistochemistry could be of considerable help. Indeed, benign PTT would strongly express CK10 and involucrin, whereas in the malignant portion there would be a strong expression of nuclear cell proliferation antigen, as well as CK16 with loss of CD34 expression [3].

The main differential diagnosis of PTT is squamous cell carcinoma (SCC), because of the histological and sometimes evolutionary similarities. Other authors have even considered PTT as a follicular variant of SCC [3]. The differential diagnosis of PTT may also include squamous cell cyst, lipoma, pilomatrixoma, keratoacanthoma, dermatofibrosarcoma, basal cell tumor, and angiosarcoma [1].

PTT is usually benign in course, with a few cases with malignant transformation with fatal outcome [7]. Criteria for malignancy have been defined such as rapid growth of the cyst, appearance of superficial ulcerations, invasion of adjacent tissues, presence of areas of necrosis in histology with high mitotic activity and marked cytonuclear atypia [3]. In our case, the slow evolution of the tumor,the CT scann image, clinical and histological presentation excluded the malignant nature of the tumor.

At the same time, Noto, in view of the frequency of recurrence and the occurrence of some metastatic cases, considers that all PTT should be classified as low-grade adnexal carcinomas [5].

Surgical removal of the cyst wall without damage is sufficient for the treatment [1]. Considering the general condition of our case, as well as the absence of suspicious signs, a surgical removal of the cyst without deterioration was performed. The most aggressive PTT may require wide local excision and have a higher risk of recurrence, for which additional measures such as lymph node dissection, chemotherapy and radiotherapy may be necessary [2]. Mohs micrographic surgery can be undertaken to reduce recurrence and metastasis rate after tumor resection [4].

Conclusion

If this entity is widely recognized by the literature, it still escapes to a nosological, diagnostic and consequently prognostic and therapeutic consensus. The ambiguity and especially the aggressiveness of this tumor (PTT) pushes to carry out studies in order to establish diagnostic and prognostic criteria as well as to guide the therapeutic management [3]. The purpose of our presentation is to draw the practitioners' attention to the unpredictable evolution of this tumor imposing a monitoring of the patient.

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