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Fonseca, Theresinha C.; Bandeira, Camila L.; Sousa, Bruno A.; Farias, Terence P.; Eisenberg, Ana Lúcia A.

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Proliferating trichilemmal tumor: case report

Tumor triquilemal proliferante: relato de caso

Theresinha C. Fonseca; Camila L. Bandeira; Bruno A. Sousa; Terence P. Farias; Ana Lúcia A. Eisenberg

Instituto Nacional de Câncer (Inca) José Alencar Gomes da Silva, Rio de Janeiro, Brasil.

ABSTRACT

The proliferating trichilemmal tumor is a lesion with trichilemmal differentiation, more common occurring among elderly women; it affects the scalp with sizes ranging from 2-10 cm. Microscopically, it is solid-cystic, well-defined, affecting the dermis and the subcutaneous cellular tissue. It presents trichilemmal and squamous keratinization. Pleomorphism may be present. Ghosts, apocrine and spindle cells can be observed. The differential diagnosis should be performed with malignant trichilemmal tumor and squamous cell carcinoma. Their behavior is benign and complete resection is recommended. Our goal is to report a case of proliferating trichilemmal tumor.

Key words: pathology; scalp; neoplasms; skin.

INTRODUCTION

The proliferating trichilemmal tumor (PTT) is a solid-cystic lesion showing trichilemmal differentiation present in the isthmus of the hair follicle[1]. It was first described by Wilson Jones as proliferating epidermal cysts in 1966[2].

The pathogenesis is unknown. In some cases, the human papilloma virus is present[1]. It is unclear whether PTT develops *de novo* or arises from an existing trichilemmal cyst[3-5].

PTT has several denominations, such as squamous cell carcinoma in sebaceous cyst, sub epidermal acanthoma, among others, reflecting the different interpretations from several authors, according to their histogenesis and biological behavior[1, 5-7].

PTT occur more commonly in elderly women, and 90% are located on the scalp, but can also affect the face, trunk and back[1, 8, 9]. It is presented as a solitary, nodular and exophytic tumor, and may develop from a nevus sebaceous[1, 7, 8, 10, 11]. Multiple lesions are rare. The size may vary from 2 cm to 10 cm in its largest diameter, and lesions greater than 24 cm have been described[12]. Alopecia and ulceration can be observed[1] .

Macroscopically the lesions are multinodular. On the cut surface the cysts are filled with keratin with calcification[1, 5, 6]. Microscopically, it is presented as a solid-cystic well-defined mass that affects the dermis and may extend to the subcutaneous cellular tissue. The neoplastic epithelium presents trichilemmal keratinization, which is characterized by peripheral palisade of basaloid cells and bulky squamous cell with large eosinophilic cytoplasm with abrupt keratinization. Epithelial invaginations into the cystic lumen are observed. Calcification and cholesterol crystals may be abundant. Epithelial cells vary from monotonous without atypia to pleomorphic with mitosis[1, 5, 6]. Areas with atypias may be indistinguishable from squamous cell carcinoma. Ghost cells, which are the expression of matrix differentiation, apocrine differentiation and spindle cells, can be observed[1, 5, 6, 13-16].

The differential diagnosis may include malignant proliferating trichilemmal tumor and squamous cell carcinoma, which have severe cytologic atypia and invasion of adjacent tissues[17-19].

By immunohistochemical study, PTT expresses cytokeratin of the fetal hair follicle and CK7, but this technique is not used to diagnose this lesion, nor is it useful to differentiate it from squamous cell carcinoma and malignant proliferating trichilemmal tumor[20].

PTT without atypia has a benign behavior, the complete removal of the lesion is recommended to prevent recurrences[1, 5, 6]. Tumors with cytological atypia have unknown biological behavior and may recur locally or develop metastases[21-26].

The aim of this paper is to present as a case report, the stages of diagnosis and treatment of PTT.

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CASE REPORT

Female patient, Italian, 80 years old, was enrolled in 1992 at the Instituto Nacional de Câncer (Inca) when a basal cell carcinoma of the face was resected. She returned in 2012 reporting excision of scalp lesion five years ago without histopathological diagnosis. One year ago she noticed an increase of the lesion in the same topography, painlessly. On examination she had a 15 cm tumor in its longest axis, ulcerated and friable over the scalp. There was no cervical lymphadenopathy. The hypothesis diagnosis was invasive scalp lesion. Incisional biopsy was performed and the histopathological diagnosis was actinic keratosis.

Computed tomography (CT) showed solid, expansive and lobulated formation, affecting the skin and subcutaneous cellular tissue in the left parietal region, with heterogeneous enhancement, hypodense areas of different sizes and shapes and coarse calcifications in between, measuring 8.5 × 6.3 × 4.6 cm. Thickening of the corresponding bone plate was observed (**Figure 1**).

Resection of the scalp tumor was performed with bone curettage and immediate reconstruction. The specimen was sent for intraoperative consultation (IC). Macroscopically, the specimen was represented by oval segment of skin and subcutaneous cellular tissue, measuring 14 × 11 × 7 cm, with brown and smooth epidermis, showing a central, brown-clear, lobulated lesion, measuring 10 × 7 cm. In the sections, the surface was variegated with brown-clear and smooth areas, port-wine and softened areas, and brown-green and bright areas, 0.3 cm distant from the nearest surgical margin (deep) (**Figure 2**).

On frozen sections, we observed a subepithelial expansive lesion, constituted by nests of well-differentiated squamous cells (**Figure 3**). The result of the IC was squamous neoplasia of uncertain biological behavior with tumor-free surgical margins.

Microscopically we observed an expansive solid-cystic tumor well delimited from the adjacent epidermis. The cell nests were composed of cell with large cytoplasm without atypia. Cell nests with abrupt keratinization and ghost cells were also found (**Figure 4**). The histopathological diagnosis was PTT.

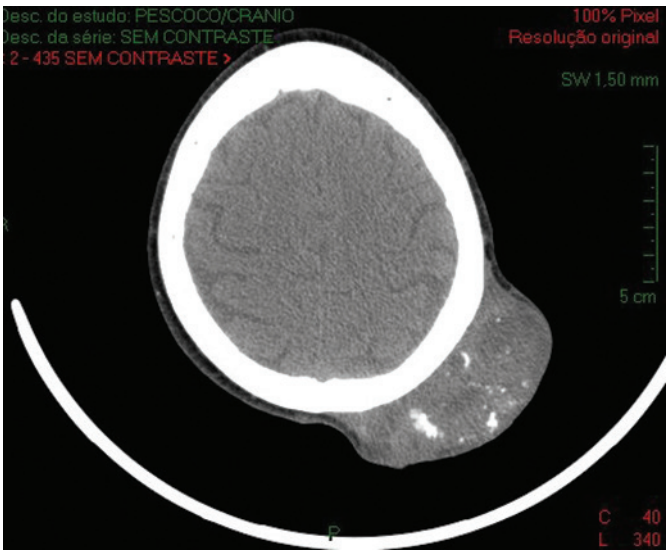

FIGURE 1 – *Computed tomography, axial view*

Hypodense scalp lesion, with calcifications without periosteal invasion, in left parietal-temporal region.
Source: PACS-Inca. Rio de Janeiro, October/2015.

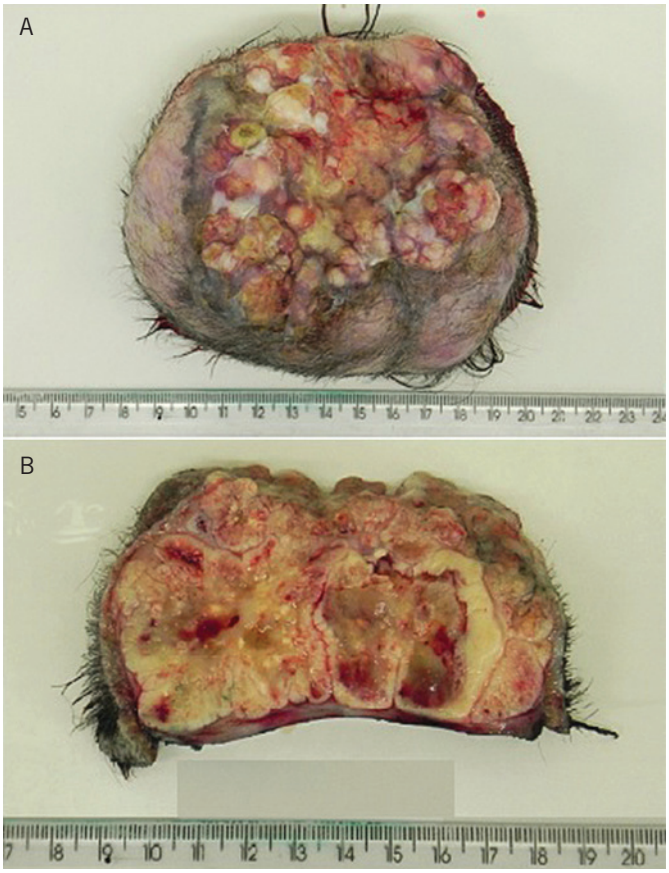

FIGURE 2 – *Macroscopic aspect*

A) skin and subcutaneous cellular tissue segment centered by vegetating bulky lesion, brown-clear with granular areas, lobulated, measuring 10 × 7 cm; B) variegated cut surface with brown-clear and smooth areas, port-wine and softened areas.

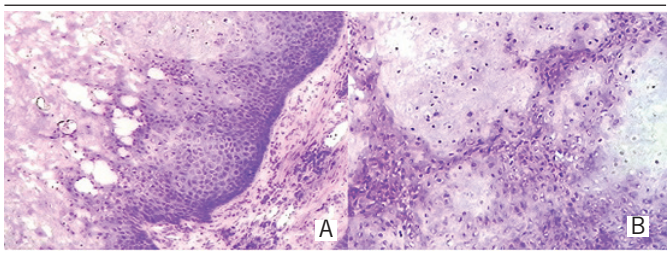

FIGURE 3 – *Intraoperative consultation*

A) frozen sections showing expansive sub epithelial lesion, consisting of nests of well-differentiated squamous cells (toluidine blue stain, 100×); B) frozen sections showing expansive sub epithelial lesion consisting of nests of well-differentiated squamous cells (toluidine blue stain, 200×).

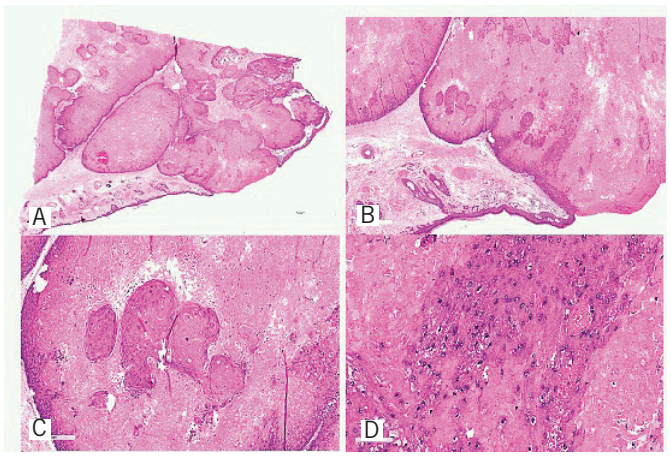

FIGURE 4 – *Microscopic aspects of proliferating trichilemmal tumor*

A) well-defined cystic solid mass, affecting the dermis (HE, 40×); B) detail of Figure A (100×); C) nests of neoplastic bulky squamous cell with extensive eosinophilic cytoplasm with abrupt keratinization (HE, 100×); D) detail of Figure C (HE, 200×).

HE: hematoxylin and eosin.

DISCUSSION

PTT is a rare tumor arising from the hair follicle epithelium, more common in elderly female patients. There are cases described in young patients[8, 9]. Ninety percent of cases are located on the scalp and are presented as a slow-growing solitary nodule. PTT may occur less frequently in other topographies, such as the vulva, trunk, face, lips, buttocks and back[1, 8, 9]. Clinically and microscopically, PTT can simulate malignancy and the correct diagnosis is essential due to the indolent biological behavior of these lesions[1, 9]. Our patient was an elderly woman with a bulky lesion in the scalp, with history of previous resection and recurrence.

In clinical examination, the lesion in our patient simulates infiltrative and relapsed tumor, however microscopically the appearance was PTT. We observed proliferation of basal cells and squamous cells with abrupt trichilemmal differentiation and ghost cells, without atypical cells, as described in the literature[1, 5, 6]. The diagnoses of squamous cell carcinoma or malignant proliferating trichilemmal tumor were discarded by the histological analysis.

In most cases, PTT has a benign biological behavior, and resection with free surgical margins is the recommended treatment[1, 26]. This was the treatment of choice for our patient.

There are reports of PTT with aggressive clinical behavior with recurrence and/or metastasis. The more aggressive behavior is more common in tumors located out of the scalp, when it is fast-growing and infiltrative, tumors larger than 5 cm, presence of atypia and mitotic activity[21-27]. Our case showed no clinical and/or microscopic aspects related to the more aggressive biological behavior, but patient's follow-up is indicated for assessment of evolution of the illness.

CONCLUSION

PTT is an uncommon neoplasm, and the reporting these lesions are important due to the good clinical evolution compared to the malignant macroscopic and microscopic feature.

RESUMO

O tumor triquilemal proliferante é uma lesão com diferenciação triquilemal, mais frequente em mulheres idosas; acomete o couro cabeludo, com tamanho variando de 2 cm a 10 cm. Microscopicamente, é sólido-cístico, bem delimitado, comprometendo a derme e o tecido celular subcutâneo. Apresenta ceratinização triquilemal e escamosa. Pleomorfismo pode estar presente. Células fantasmas, apócrinas e fusiformes podem ser observadas. O diagnóstico diferencial deve ser feito com o tumor triquilemal maligno e o carcinoma de células escamosas. O seu comportamento é benigno, sendo recomendada a ressecção completa. Nosso objetivo é relatar um caso de tumor triquilemal proliferante.

Unitermos: *patologia; couro cabeludo; neoplasias; pele.*

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CORRESPONDING AUTHOR

Theresinha Carvalho da Fonseca

Instituto Nacional de Câncer; Av. Cordeiro da Graça, 156; Santo Cristo; CEP: 20220-400; Rio de Janeiro-RJ, Brasil; Phone: +55 (21) 3207-3440; e-mail: theresinha.fonseca@inca.gov.br.