A rare ossifying trichilemmal cyst in a young female patient: a case report and literature review

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Abstract

Trichilemmal cysts (TCs) constitute the second most common cutaneous cysts and are mostly presented on the scalp of middle-aged women. Therefore, it is unusual for a young person to have a TC and it is extremely rare for a TC to be ossified. In the literature, only 8 cases of TCs with concomitant ossification have been described. We report the case of a 22-year-old female who presented with a scalp nodule and was treated via surgical excision of the lesion. The pathology examination of the surgical specimen revealed a lesion consisting of a multilayered squamous epithelium of slightly eosinophilic maturing keratinocytes. There was no granular layer, whereas the core of the lesion was occupied by mature bone tissue with calcium deposits. The definite diagnosis of the pathology report was ossifying TC. The aim of this report is, to enlighten clinicians about this rare pathological entity.

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Introduction

Trichilemmal cysts (TCs), also known as pilar cysts, are benign lesions that originate from the outer root sheath of the hair follicles and occur predominantly on the scalp of women aged between 50-75 years.[1] In a study of 2438 cases of cutaneous cysts, the TCs were the second most common, presented in 23.8% of patients, of whom 56.8% were female.[2] Greater than 90% of TCs occur on the scalp and the color of the skin has not been associated with their appearance.[1,2]

TCs can be presented either sporadically or in an autosomal dominant pattern of inheritance, with incomplete penetrance.[3,4] In the case of heredity, there are multiple cysts all over the body that appear at a young age, and *PLCD1* has been recognized as the responsible gene.[3,4]

Furthermore, TCs are externally smooth and mobile.[1] They are composed of a stratified squamous epithelium with dense keratin and without an inner granular layer.[1,5,6] The presence of foci of calcification in 25% of the cases has been reported, but the appearance of complete bone tissue is extremely rare.[1,5-10]

We report the case of an ossifying TC in a young female patient, and we aim to further educate clinical doctors on this rare entity.

Case Report

We present the case of a 22-year-old caucasian female with a non-painful, slowly growing, clearly visible nodule in the parietal region of the scalp. The patient detected the lesion one year before her admission to the hospital. She was taking no medication and had no other underlying disease. Moreover, her medical history was free of any previous surgical interventions. The patient underwent excision of the nodule through a 4 cm fusiform incision under local anesthesia.

The pathology examination of the surgical specimen revealed complete excision of a well-described 2 cm lesion that was extending to the epidermis (Figures 1 and 2). The lesion was formed by multiple layers of squamous slightly eosinophilic ker-

atinocytes. Furthermore, a granular layer was not detected. Calcium deposits were evident in the core of the lesion, as well as foci of mature bone tissue with sub-cellular marrow. Cartilaginous tissue was not recognized. The above findings were compatible with an ossifying trichilemmal cyst with rupture of the cystic wall.

Table 1. Comparison of the 8 cases with ossifying cysts, regarding the age and the sex of the patients, and the site of the lesions.

Author	Site of lesion	Age and sex of the patient
Pusiol *et al.*[1]	Scalp	46 years - female
Civatte *et al.*[7]	Left mammary region	57 years - female
Mommers *et al.*[8]	Scalp	61 years - female
Bar *et al.*[9]	Scalp	39 years - female
Bulut[10]	Scalp	40 years - female
Baldovini *et al.*[5]	Scalp	74 years - female
	Scalp	48 years - female
	Upper back	53 years - male

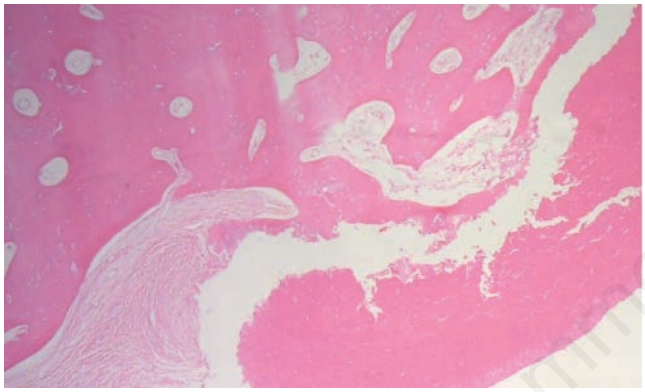

Figure 1. Stratified squamous epithelium of the trichilemmal cyst with mature bone tissue within it (Hematoxylin and Eosin stain 2.5x). The image was processed with the use of MS Paint, edition 6.1, 2009 Microsoft Corporation.

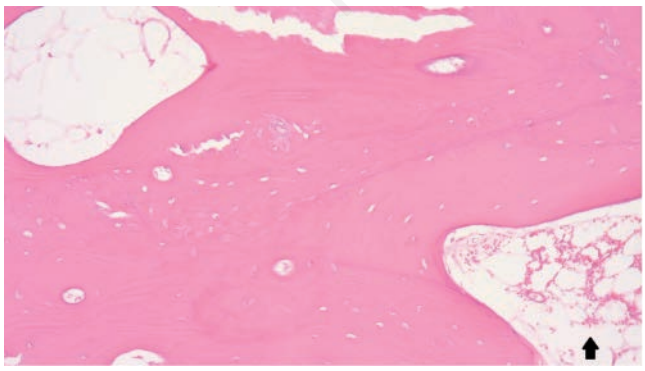

Figure 2. Closer view of the mature bone tissue in the core of the trichilemmal cyst (Hematoxylin and Eosin stain 10x). The arrow is demonstrating the bone tissue with sub-cellular marrow. The image was processed with the use of MS Paint, edition 6.1, 2009 Microsoft Corporation.

The patient underwent regular follow-up meetings. Her last follow-up was three months after her admission to the hospital, in which she was found in good clinical condition without signs of recurrence.

Discussion

TCs are usually asymptomatic unless they calcify or rupture their contents leading to the inflammatory process and pain in the affected site.[6,11] They constitute the second most common cutaneous cysts and are diagnosed mainly clinically.[2,11] Imaging modalities such as ultrasound and computerized tomography can provide additional information regarding the location and characteristics of the cyst, whereas diagnostic confirmation is made via pathology examination of the lesion.[12]

Radical surgical excision of the trichilemmal cyst is the gold standard treatment.[1,6] Recurrence may occur in case of incomplete removal of the cystic wall.[11] Furthermore, inflammation of the cyst should lead to postponement of the surgical removal until the inflammatory signs subside.[11]

Although most TCs are benign lesions, there are rare examples of TCs appearing malignancy.[13-15] In 2% of TCs, single or proliferating cells lead to tumors called proliferating trichilemmal cysts which may be locally aggressive.[13-15] Metastases to regional lymph nodes and distant organs have been rarely observed.[13-15]

Moreover, it is extremely rare for a TC to be ossified.[1,5-10] In the literature, only 8 cases of TCs with concomitant ossification have been reported (Table 1).[1,5,7-10] The rupture of the cystic wall is a possible explanation for the presence of ossification.[1,5,7-10] Dystrophic calcification following the rupture of the cystic wall may lead to the formation of bone. Another explanation for the appearance of cutaneous bone is that, it may have been directly formed from osteogenic stromal elements without a cartilaginous precursor.[1,5,7-10]

Conclusions

Ossifying TCs are extremely rare. Nevertheless, clinicians should consider this rare entity in the differential diagnosis of dermal cysts. It should always be diagnosed and treated appropriately to avoid recurrence.

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