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CASE REPORT

A case with a giant trichilemmal cyst

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Abstract: Trichilemmal cyst, also known as pilar cyst, was first reported by dermatologist Hermann Pinkus in 1969. It is the most common cyst in the parietal and occipital regions of the scalp. Herein we report a case presented with a giant trichilemmal cyst and review the literature regarding the differential diagnosis of this lesion.

Keywords: trichilemmal cyst; pilar cyst; case report; diagnosis

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Introduction

Trichilemmal cyst, also known as pilar cyst, originates from the outer root sheath of the deep hair follicle and is filled with keratin[1]. It occurs in areas of high follicular density, most commonly in elderly women (80%) and in the scalp (90%)[2]. Complete loss of the activity of the P53 tumor suppressor gene may lead to more aggressive forms of the trichilemmal cyst, such as proliferative trichilemmal tumor (PTT) and malignant proliferative trichilemmal tumor (MPTT)[1,3]. They are often confused with sebaceous cysts, but the punctum contents of these cysts make trichilemmal cysts different from others.

Case report

A 55-year-old woman was admitted from a nephrology clinic to our clinic, with a six-year history of nontender growing swelling on her scalp. The patient, who lived in rural area, had dialysis treatment for five years for chronic renal failure (CRF) due to hypertension.

On physical examination, there was a solid, fixed and painless ulcerated subcutaneous nodule with dimension of 8×7×5 cm on the left parietooccipital region of the scalp (**Figure 1**). The lesion was totally excised in the local operation room, considering the general condition of the patient. The defect was closed with a rotation flap.

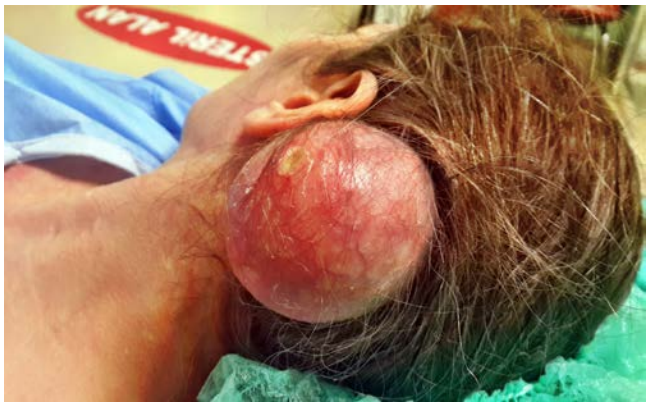

Figure 1. A giant ulcerated nodule in the parietooccipital region

On the macroscopic examination, a piece of cystic tissue with smooth surface and lumen keratin was observed in the 8×7×5-cm nodule. Histopathologic examination revealed a cyst surrounded by mature multi-layered flat epithelium with no granular layer and wet keratin in its lumen (**Figure 2**). The dysplasia or proliferation was not observed in the cystic epithelium and the case was reported as "trichilemmal cyst".

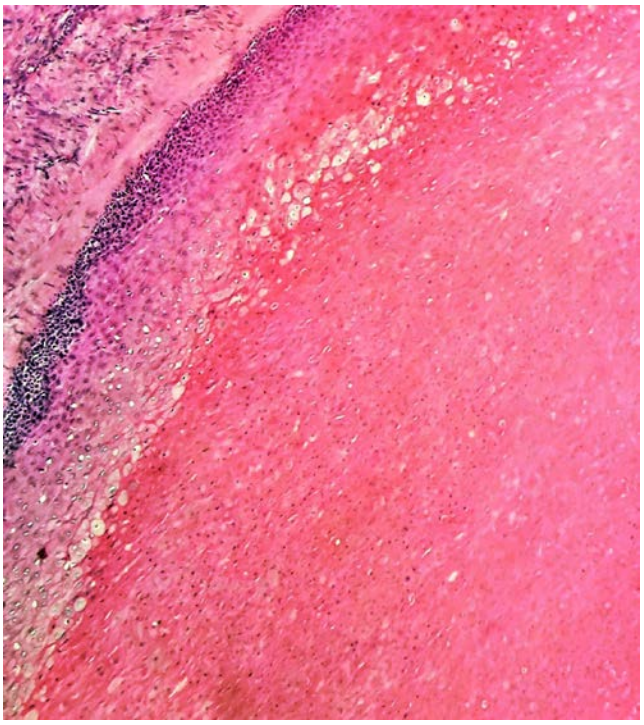

Figure 2. A cyst surrounded by mature multi-layered flat epithelium with no granular layer and wet keratin in its lumen (H&E 20×)

Discussion

Pilar cyst was named as trichilemmal cyst (TC) by Pinkus in 1969[4]. It is the most common cyst in the parietal and occipital regions of the scalp. Keratin-filled cysts originate from the outer root sheath of the deep hair follicle. They are often misdiagnosed as sebaceous cysts, but differently from these, they do not contain the punctum. They are typically round, flat, solid, mobile, solid, slow-growing, painless and with varying sizes. Browstein *et al.* reported a 90% scalp localization and 84% female predominance in 50 PTT cases[5]. Sau *et al.* found that the most frequent observed localization of TC was the scalp and the ratio of female to male was 2.5:1[6]. Although the most common localization of cysts has been reported to be the scalp, rare cases have been observed in the face, ear, upper limbs, genital area, hips and lower limbs[7]. Very rarely, local recurrence and metastasis of the tumor have been reported[7,8].

The proliferative trichilemmal tumor, first reported by Jones, is less common and originates from TC[9]. Both lesions are histologically characterized by the presence of trichilemmal keratinization, the absence of granular layering, the presence of palate in the basal layer similar to the outer root sheath of the hair follicle, and the presence of dense keratin in the cyst cavity extending into the subcutis[1,5,10]. According to Brownstein, simple TCs showed proliferation and transformation to PTT due to trauma or inflammation. In recent reports, loss of p53 tumor suppressor gene activity was implicated in this process[1,3]. More aggressive PTTs can mimic squamous cell carcinoma, usually with a wider and atypical appearance, although they show the same histologic features as TC. MPTTs are very rare and have invasive features with classical trichilemmal keratinization appearance[3]. PTTs that are thought to develop from the TC base are less common.

Older women are more common among patients with TC. These lesions are exophytic nodular and multilobular lesions. Alopecia on the lesion and ulceration may be detected. TC and PTT can be seen in sizes ranging from 0.2–10 cm[1,11]. PTTs contain more atypia than TCs. Involvement of surrounding tissues is observed in PTTs showing atypia in the middle and high position. However, MPTT is referred to in cases of severe atelectasis and peripheral involvement. Nuclear pleomorphism, atypical mitotic activity, necrosis and infiltrative margins are seen in MPTT[1,12]. PTTs can be confused with invasive squamous cell carcinoma if they show marked atypia and pleomorphism that is microscopically indistinguishable when infiltration is seen[1,13].

Approximately 20% of epithelial cysts are TCs while 80% are epidermoid cysts[1]. Epidermoid cyst, lipoma, pilomatrixoma, keratoacanthoma, dermatofibrosarcoma protuberans, cilindroma, basal cell tumor, squamous cell tumor and angiosarcoma should be considered in the differential diagnosis of trichilemmal cysts[14]. Considering the general condition of our case, the mass was excised in the local operation room after dialysis. Surgical removal of the cyst wall without deterioration is sufficient in the treatment of TCs. If invasion or malignancy can be seen in PTTs, excision with 1-cm solid margin is recommended to prevent recurrence. In MPTTs, more aggressive applications such as lymph node dissection and chemoradiotherapy may be needed in addition to local excision[1,13].

The characteristic of our case was that the lesion size was larger than the published cases in the literature.

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Histological examination of the specimens showed that there was no evidence of trichilemmal keratinization, no granular cell layer, no mitosis and no cellular atypia at the center of the cyst epithelial layer and cell islands, which showed marked keratinization.

Conflict of interest

The authors declare no potential conflict of interest with respect to the research, authorship, and/or publication of this article.

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