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Case Report

Steatocystoma multiplex involving the face and scalp: a case report

一宗涉及臉部和頭皮的多發性脂囊瘤案例報告

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Steatocystoma multiplex (SM) is an uncommon, familial or sporadic disorder of the pilosebaceous unit. It is usually characterised by multiple, asymptomatic, skin-coloured or yellowish, sebum-containing dermal cysts. Steatocystoma multiplex can appear anywhere, but is more common on the trunk, neck, inguinal region and proximal extremities. Cases with lesions localised on the face and scalp have also been described, but are rare. A 62-year-old man presented with multiple, yellowish, smooth, non-tender cutaneous cysts of about 2 cm in diameter on his scalp, bilateral temples and upper back. He first noticed the lesions 10 years ago. There was no family history of similar lesions. The nails, teeth and hair were normal. Histopathology revealed steatocystoma multiplex. Treatment by CO_2 laser was planned.

多發性脂囊瘤是一種罕見的家族性或偶發性毛囊皮脂腺單位病症。 其通常特徵爲多發性、無症狀、皮膚色或淡黃色的含皮脂眞皮囊腫。多發性脂囊瘤可以出現在任何部位，常見於軀幹、頸部、腹股溝區及近端四肢。限於面部和頭皮的個案也曾有論述，但屬罕見。一名六十二歲的男仕，在他的頭皮、雙側太陽穴部位和上背部有多個黃色光滑、無觸痛感的皮膚囊腫，直徑約有兩厘米。他十年前首次注意到這些病變，而指甲、牙齒和頭髮全都正常，其家族成員亦不見有此皮膚表現。組織病理學顯示爲多發性脂囊瘤。治療方案會考慮二氧化碳激光治療。

Keywords: CO_2 laser, face, pilar cyst, scalp, steatocystoma multiplex

關鍵詞：二氧化碳激光、面部、毛髮囊腫、頭皮、多發性脂囊瘤

Introduction

Steatocystoma multiplex (SM) is an uncommon disorder of pilosebaceous duct junction. A typical form of the disease is characterised by multiple, yellowish, smooth, cystic nodules which may vary in size from a few millimeters to 20 mm or more. Usually, the lesions are asymptomatic, but some lesions may become inflamed, suppurated and heal with scarring. The lesions are seen predominantly on the chest, back, proximal limbs, axilla and groin areas. On the other hand, a rare limited form of SM affecting only the face, scalp and nose, has also been described.[1-5]

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We report a rare case of typical SM with scalp and facial involvement.

Case report

A 56-year-old male patient presented with multiple, asymptomatic skin-coloured nodules involving the scalp, temples, postauricular area, neck and upper back. He had been referred from the Department of Psychiatry where he was hospitalised for organic affective disorder. He had a history of symptomatic epilepsy and parasagittal meningioma which was operated in 2011.

On examination, there were nodules ranging from 0.3-3 cm in diameter (Figure 1 and Figure 2), which had a smooth surface and were filled with a yellow, creamy/cheesy material. The nails, teeth and hair were normal. There was no family history of similar lesions. The patient stated that the lesions had appeared immediately after a traffic accident 10 years earlier and the number of the lesions had increased over the years. Histopathology of the lesion showed a cyst in the mid-dermis that was lined with stratified squamous epithelium without granular layer. The cyst was filled with amorphous keratinous material (Figure 3). These clinical and histological findings were compatible with SM.

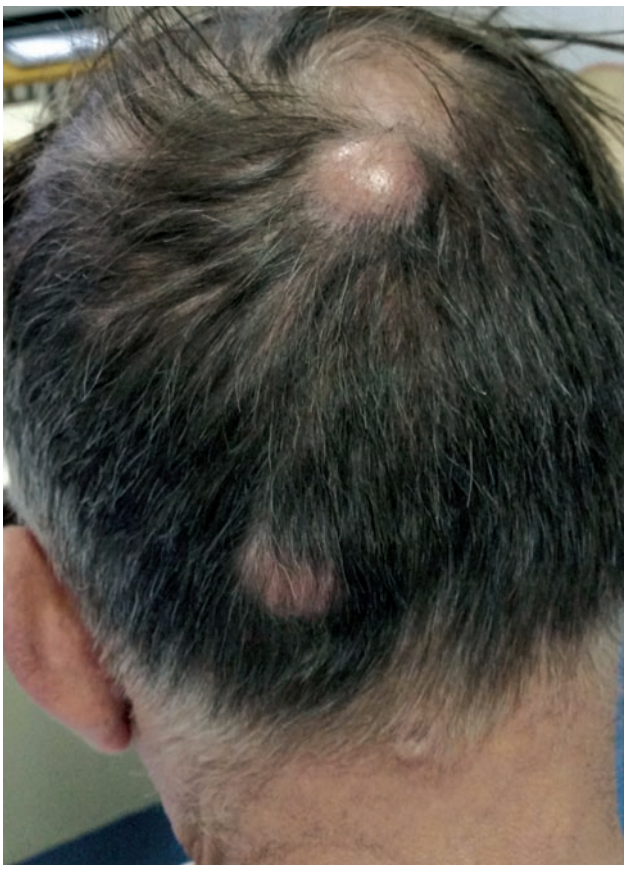

Figure 2. Steatocystoma multiplex on the scalp and neck.

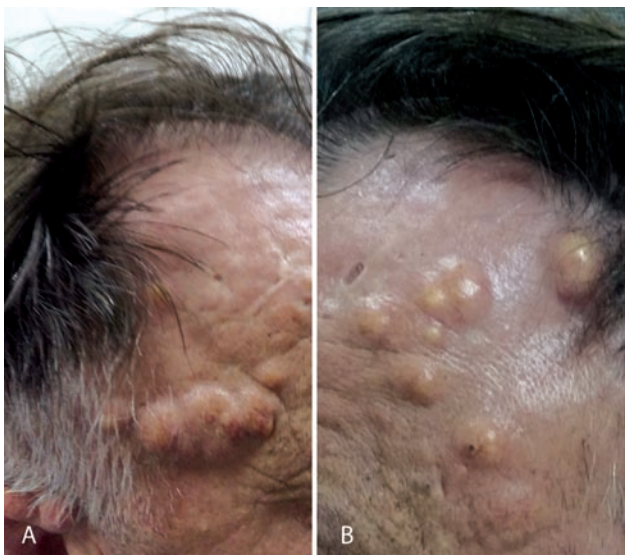

Figure 1. Cystic nodules on the temples.

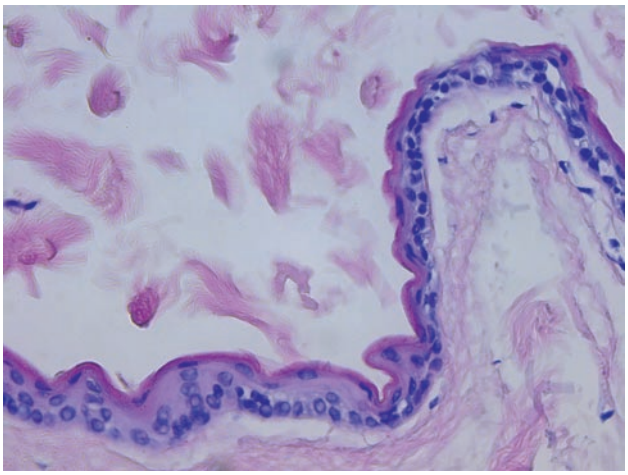

Figure 3. Histopathology revealed dermal cyst with walls lined by stratified squamous epithelium without granular layer.

Discussion

Steatocystoma multiplex is an uncommon, hereditary or sporadic, cystic cutaneous disorder, involving areas that are anatomically rich in sebaceous glands. The term was coined by Pringle in 1899, but probably the first case was reported by Jamieson in 1873, and, in 1896, Dubreuillh and Auche were the first to observe the presence of sebaceous glands within the cysts.[6]

According to the localisation and outcome of the lesions, SM may be subdivided into localised, generalised, facial, acral and suppurated forms.[7] Involvement of the scalp and face is rare, despite the fact that these regions are rich in sebaceous glands. Up to 2013, only 12 cases of SM located on the scalp have been described.[8] Kim et al have drawn a parallel between the typical SM and SM limited to the scalp. They found that the typical form is usually hereditary and appears in childhood or early adulthood, in contrast to SM limited to the scalp, which is sporadic and appears in late adulthood.[1] In accordance with this statement, Lee YJ et al and Lee D et al reported two sporadic cases of steatocystoma multiplex located exclusively on the scalp and associated with alopecia patches due to congenital causes and trichotillomania respectively.[2,9]

Our patient had typical SM involving the scalp and face and denied any history of similar lesions among family members. This is a nodular variant of the disease with typical skin-coloured asymptomatic nodules filled with oily, yellowish, non-odorous content released during the biopsy. The onset of the disease was in adulthood, when the patient was 48-year-old. Rossi et al also described sporadic, typical SM with scalp and facial involvement.[6] Furthermore, SM affecting the face and scalp has been classified into three types: a) a facial papular variant, b) sebocystomatosis, c) lesions confined to the scalp. Rare cases of papular SM exclusively on the face has been reported by Nishimura et al.[3] Papular SM localised on the face and scalp has also been reported.[10]

Linear, naevoid, congenital SM has also been reported to occur on the nose. The naevoid appearance is in accordance with current pathophysiological recognition of SM as a naevoid or hamartomatous malformation of the pilosebaceous junction.[4] In the suppurative type of the disease, the lesions become inflamed and suppurated after minor trauma. Steatocystoma simplex is the sporadic solitary tumour counterpart of SM.

Hereditary SM is inherited in an autosomal dominant manner and is often associated with mutations in the helix initiation domain (1A) of the *KR17* gene. These mutations are identical to the mutations seen in pachyonychia congenita type 2, but our patient had no features of this disease. The sporadic, non-hereditary variant is more common.

A female-to-male ratio of 1.2:1 has been reported. The majority of cases are seen in the third decade of life, although SM can present at birth and as late as 78 years of age.[11] The causative factors remain unclear, but trauma, infection, or immunological events may be responsible. In the current case, the lesions appeared immediately after trauma.

Clinically suspected SM should be confirmed on histology to exclude eruptive vellus hair cysts and epidermal inclusion cysts. Histologically, SM consists of dermal cystic lesions that are empty, partially collapsed, folded and covered with a wall formed by a flattened, stratified squamous epithelium without a granular layer and with a cellular eosinophilic cuticle over its surface. Sebaceous glands are located in the cyst wall. Riedel et al have shown that cysts of SM are calretinin-7 positive immunohistochemically, and that it is a distinct feature of the disease.[11] Nodules of SM can also be recognised by mammography and sonography.[7] There are many different methods for the treatment of SM. Surgery is the most effective treatment, but is not a desirable method when multiple lesions are present.

Treatment with Er: YAG laser followed by tetracycline ointment and CO_2 laser has been reported to be well-tolerated and cosmetically satisfactory for the treatment of multiple lesions of SM.[6]

Our patient is an extremely rare, sporadic case of typical SM affecting the scalp and face. At present, CO_2 laser treatment is being considered.

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