Case Report

Steatocystoma multiplex involving the face and scalp: a case report

一宗涉及面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 CO₂ laser was planned.

Keywords: CO₂ laser, face, pilar cyst, scalp, steatocystoma multiplex

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
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.
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 CO₂ 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, CO₂ laser treatment is being considered.

References