Steatocystoma multiplex - case report

Anca E Chiriac*,¹, Tudor Pinteala¹, Piotr Brzezinski²

¹ "Grigore T. Popa" University of Medicine and Pharmacy, Iasi, Romania, ² Department of Dermatology, 6th Military Support Unit, Ustka, Poland

Abstract

Steatocystoma multiplex is a rare, benign, sporadic or familial disorder (autosomal dominant mutation in Keratin 17). Diagnosis is based on clinical grounds: multiple cysts, localized in axilla, groin, neck, scrotum and proximal extremities. We describe a case of Steatocystoma multiplex in a 28 year old male patient with a family history of similar lesions.

Keywords: *Steatocystoma multiplex, familial disorder, scrotum*

Case report

A 28 year-old Caucasian male patient was seen in the Dermatology Unit for multiple nodular lesions, localized on the scrotum, observed by the patient a few months prior to consultation (Fig. 1).

Fig. 1. Round cysts of different size, on the scrotum

On clinical examination numerous, whitish and of different size (ranging from a few mm to 3 cm) cystic tumors were detected on the scrotum, not accompanied by any symptoms. The patient was in good health status, he denied any past personal medical history and drug intake, but he admitted that similar lesions were reported by his father. Based on simple clinical observation, a diagnosis of Steatocystoma multiplex was made, the patient refused skin biopsy. The patient was reassured by the benignity of his disease, different therapeutical options have been proposed and genetic counseling was advised.

Discussions

Steatocystoma multiplex is also known as epidermal polycystic disease and sporadic or familial disorder (autosomal dominant mutation in keratin 17) [1].

Usually lesions appear during adolescence or around twenties (maybe a hormonal trigger), but are fully expressed and diagnosed in adulthood or even later. No gender prevalence has been reported.

Diagnosis is mostly clinical: multiple dermal cysts, of different size, involving areas with high density of sebaceous units: axilla, groin,

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*Corresponding author: Anca E. Chiriac, "Grigore T. Popa" University of Medicine and Pharmacy Iasi, Romania. Email: chiriacut@yahoo.com.

neck, scrotum and proximal extremities; but rare forms have described the lesions on the face [2], generalized [3], scalp [4].

A classification can be made: localized, generalized; facial, acral, and the suppurative types; solitary lesions are known as Steatocystoma simplex [4].

Steatocystoma multiplex can be associated with pachyonychia congenita type 2 (which is characterized by hypertrophic nail dystrophy, focal keratoderma, multiple pilosebaceous cysts, and myriad conditions associated with

ectodermal dysplasia) or eruptive vellus hair cysts [1, 5].

The diagnosis is confirmed by histopathological report (Table 1).

Treatment options are: cryotherapy, carbon dioxide laser, surgical excision, oral tretinoin or simple surveillance.

Particularities of the present case: a familial case of Steatocystoma multiplex in a 28 year-old male patient with localized lesions on the scrotum.

Table 1. Differential diagnosis based on histopathological findings.

Eruptive Vellus Hair Cysts	Dermal cysts lined by stratified squamous epithelium, with granular layer, contained vellus hairs; no sebaceous glands [6]
Epidermoid cysts	Cysts lined by stratified epithelium, with granular layer, lamellar keratin inside [7]
Milia	Small cysts in the superficial dermis coated by epithelium with granular cell layer' contains lamellar keratin [7]
Trichilemmal cysts	Cysts lined by squamous epithelium without granular layer, filled with homogenous keratin [7]
Steatocystomas	Cysts lined by stratified squamous epithelium without granular layer and vellus hairs inside the cavity; sebaceous lobules close to the cystic wall [7]

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