Case report

MULTIPLE PILOSEBACEOUS CYSTS

M. Lomuto, G. Ditano and V. Nirchio

ABSTRACT

The term "multiple pilosebaceous cysts" has recently been proposed to describe lesions currently classified as steatocystoma multiplex as well as those named eruptive vellus hair cysts. This terminology reflects the concept that both types of cyst are expressions of a single pathological process involving the epithelium of the hair follicle. We report a 23-year old female with cystic lesions of the abdomen, axillae, lateral neck and right leg that had been present for 6 years. The clinical aspects of the disease were typical of steatocystoma multiplex, but histopathologic findings were suggestive of both steatocystomas and eruptive vellus hair cysts. This case illustrates the close relationship between these two types of cysts that has already been suggested by several authors.

KEY WORDS

steatocystoma multiplex, eruptive vellus hair cysts, multiple pilosebaceous cysts

INTRODUCTION

Some authors (1 -3) consider steatocystoma multiplex and eruptive vellus hair cysts as manifestations of the same pathological process, Ohtake et al (4) therefore recently proposed the term "multiple sebaceous cysts" to describe both conditions.

CASE REPORT

A 23-year-old housewife was referred to our hospital for evaluation of multiple cutaneous cysts that were subjectively asymptomatic. The lesions had first appeared in the presternal region and on the abdomen (fig. 1) when the patient was 16 years old and had gradually extended to involve the axillae

the back, the lateral part of the neck and the right leg. The cysts-like lesions (ranging in diameter from 2 mm to 1 cm) were roundish with a firm, elastic consistency. The overlying skin was smooth and somewhat yellow. Some of the cysts were raised while others were embedded within the cutaneous tissue. The patient reported that the lesions sometimes became inflamed and suppurative; these episodes resolved without treatment leaving retracted brownish scars.

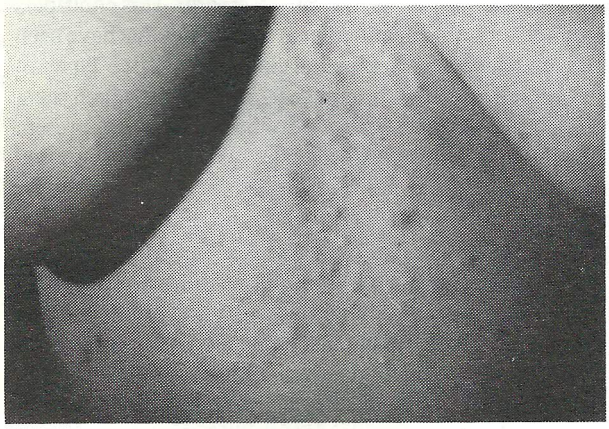
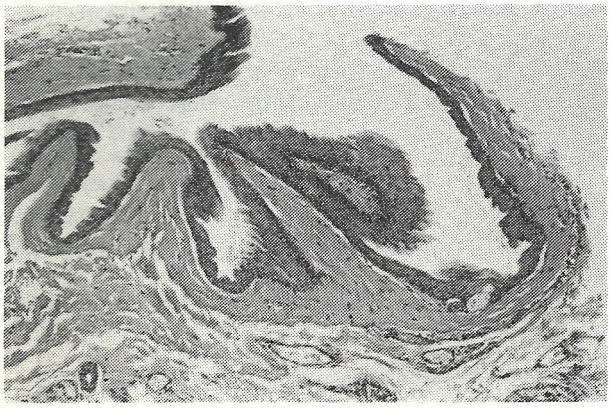


Fig. 1 Multiple cutaneous cysts in the presternal region of a 23-year old female patient



Firg. 2 Histopathology of a cyst from the axilla: apparently empty intricately folded cystic formation lined with squamous epithelium; the cells in the basal layer are arranged in palisades.

She also noted that her brother had similar lesions on the trunk for several years.

Both the patient and her father had reportedly been suffering from Raynaud's syndrome for several years.

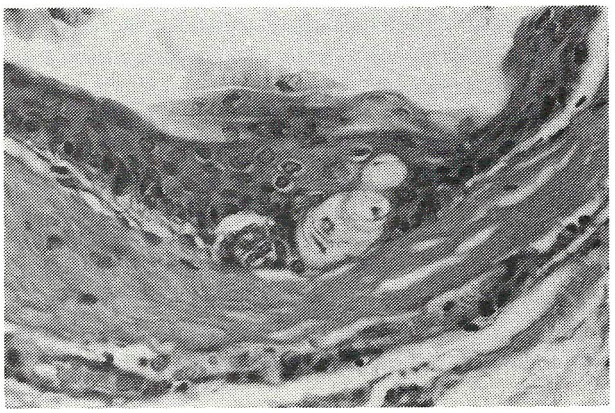


Fig. 3 The innermost layer of the cyst wall is composed of a homogeneous eosinophilic cuticle; within the wall some cells show signs of sebaceous differentiation

Laboratory findings (blood chemistry, hematology, immune parameters, urinalysis) and complete skeletal x-rays were within normal limits. An ultrasound examinations of the lesion performed with a 20-MHz probe showed features that were typical of epidermal cysts.

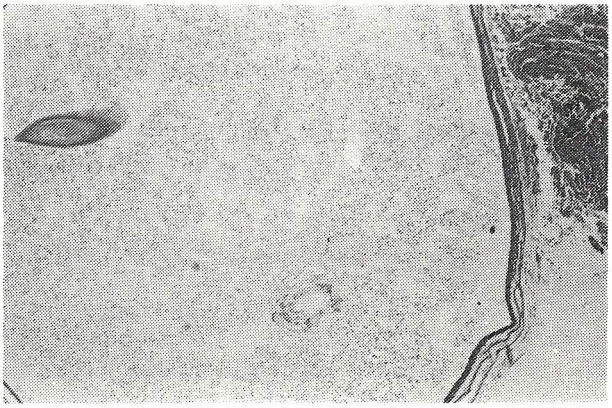


Fig. 4 A cyst from the presternal region containing a lanugo-like hair

Histopathologic examination of a biopsy specimen from the axilla (fig. 2) revealed an apparently empty intricately

folded cystic formation lined with squamous epithelium. The cells of the basal layer were arranged in palisades; the stratum granulosum was absent. The remaining cells appeared swollen and intercellular bridges were absent. The innermost layer of the cyst wall was composed of a homogeneous, eosinophilic cuticle suggesting a differentiation towards apocrine secretion. Within the walls of the cyst some cells showed signs of sebaceous differentiation (fig. 3).

A second cyst from the presternal regions was found to contain lanugo-like hairs (fig. 4). The wall of this cyst was lined with 2-3 layers of squamous epithelial cells covered by a thin, homogeneous eosinophilic cuticle. There were no signs of sebaceous glandular tissue within the lining of the cyst, the stratum granulosum was missing. The cyst did not appear to be in communication with the skin surface or with other components of the pilosebaceous unit.

DISCUSSION

In the late 19th century, Jamieson (5), Dubreuilh and Auche (6) and Boselli et al. (7) described an eruptive dermatosis characterized by cystic structures arising from the sebaceous glands. This condition is currently referred to as "steatocystoma multiplex", a term that was first used by Pringle in 1899 (8). These multiple (steatocystoma multiplex) or isolated (steatocystoma simplex) cysts generally evolve during puberty, but onset has also been described in children and elderly subjects. Scalp and facial involvement is apparently rare, but has been reported in both simplex (9) and multiplex (10) forms of the disease. In males the cysts are often distributed within a diamond-shaped area between the xiphoid process of the sternum and umbilicus; in females the most common sites of involvement are the center of the chest, the axillae, the groin and the vulva. Steatocystoma multiplex appears to be transmitted as an autosomal dominant trait. Suppuration is not uncommon and usually causes scarring.

In 1977, Esterly et al. (11) used the term "eruptive vellus hair cysts" to describe a condition whose clinical features and inheritance pattern are quite similar to those of steatocystoma multiplex. The cysts described by these authors can be histopathologically distinguished from those of steatocystoma by the vellus-type hairs they contain. Steatocystoma multiplex and eruptive vellus hair cysts are currently considered by several authors (1-4) to be morphologically different manifestations of a single pathological process in which the hair-follicle epithelium undergoes benign neoplastic transformation. This concept is supported by the histopathological similarities that have been observed between the epithelial lining of the two types of cysts as well as by the differences they present, which seem to reflect the distinct types of epithelium that are present within the hair follicle. Ohtake et al. (4) have thus proposed the term "multiple pilosebaceous cysts" to refer to both conditions based on the segment of the pilosebaceous unit that is involved in the cystic process. The following types can be observed: 1) Infundibulum: infundibular type of pilary form; 2) Infundibular-isthmic junction: infundibular-isthmic type of pilary form; 3) Isthmus: isthmic type of pilary form; 4) Pilosebaceous duct junction: pilosebaceous form; 5) Sebaceous duct: sebaceous form.

The case described here supports the view that steatocystoma and eruptive vellus-hair cysts are expressions of the same pathological process. The distribution and macroscopic features of the cysts, the histopathologic features of the axillary cyst, the onset of disease during adolescence, the absence of sexlinked familiarity, and the suppurative-cicatrical episode reported by the patient are all typical of steatocystoma multiplex, but the presternal lesion showed clearly an eruptive vellus hair cyst. In our view for these reasons we feel that the term "multiple pilosebaceous cysts" is more appropriated for conditions like the one reported here.

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AUTHORS' ADDRESSES

Michele Lomuto M.D.,professor and chairman, Department of dermatology,
Casa Sollievo della Sofferenza-IRCCS Hosp., I-71013 San Giovanni Rotondo, Italy
Giuseppina Ditano M.D., dermatologist, same address
Vincenzo Nirchio M.D., pathologist, Service of anatomopathology,
Casa Sollievo della Sofferenza-IRCCS Hospital I-71013 San Giovanni Rotondo, Italy