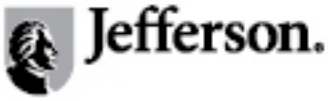

9-1-2014

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Editorial

Steatocystoma Multiplex: *Those little tumours*

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EDITORIAL

Hardly a clinician in the twenty-first century would have difficulty in identifying steatocystoma multiplex (SM). The characteristic asymptomatic yellow or skin-colored dermal papules, typically found on the trunk, upper aspects of the arms, scrotum, or chest (Figure 1) are difficult to miss, particularly when a young woman is concerned about her appearance with a low-cut blouse (Figure 2) or when a young man thinks his acne as gone astray.

DEVELOPING A CONCEPT

The first descriptions of these "innumerable number of prominent little tumours" that varied in size from "a millet-seed to a large pea" were made by James Pringle (1855-1922), better known for having delineated the adenoma sebaceum of tuberous sclerosis.[1] In 1899, he presented to the Dermatological Society of London the case of a 21-year-old man with asymptomatic "lumps" that had appeared over the previous five years.[1] Puncture of one of these lesions revealed "abundant fluid like thin skim-milk."[1] James Galloway (1862-1922) performed the histopathologic examination on a biopsy specimen taken from this patient and decided that the lesions were due to "hypertrophy of the sebaceous gland, liquefaction of its contents, and retention of its secretion."[1]

In 1898, Pier Lodovico Bosellini (1873-1945), then in Bologna, described a 40-year old man who was experiencing eight years of asymptomatic "multiple follicular cutaneous cysts."[2] He concluded that it was a retention cyst due to keratin plugging of the sebaceous glands. In Germany in 1917, Hans Günther (1884-1956), who would later distinguish himself with his work on porphyrins, analyzed the biopsy specimen of a similar patient, and suggested the term sebocystomatosis, implying the pathologic process to be analogous to lipomatosis.[i]

In 1933. Isaac Muende (1901-1987) and Archibald Cathcart Roxburgh (1886-1954) in London claimed that Pringle's nomenclature had priority to Günther's. Their description of SM is quite detailed:

> *"It was an affection of the hair follicle, just above the entrance of the gland, which prevented sebum from reaching the surface. The sebum was produced and dilated the lower part of the hair follicle, the remnants of the sebaceous gland being squeezed to one side. In the end, when it reached a certain size- that of a pea- it ceased to function."*[3]

The term sebocystomatosis continued to be used through the 1950's, as shown by a paper in the *British Journal of Dermatology and Syphilis*, that described the papules as "gelatin capsules," and defined sebocystomatosis as "a distinct clinical entity, characterized by the occurrence on various parts of the body of small cystic tumors involving the sebaceous apparatus without causing any change in the overlying skin."[4] An additional term was hereditary epidermal polycystic disease.[5]

CURRENT CONCEPTS

Today, SM is recognized as a benign nevoid malformation of the pilosebaceous apparatus that shows a mixture of keratinizing epithelium and sebaceous lobules attached to the epidermis by a thin epidermal strand.[6] A subtype of SM, known as steatocystoma multiplex suppurativa (SMS), presents with inflamed lesions that can rupture and scar.[6] On histology, SM appears as epithelial-lined sebum-filled dermal cysts, approximately 3-10 millimeters in size, with sebaceous glands present in the cyst wall (Figure 3). On the luminal side of the cyst wall is an eosinophilic cuticle with keratin, oil, and hairs in the lumen.[7] A single lesion may also occur; this entity is designated "steatocystoma simplex (SS)". In either SM or SS, the sebaceous gland tissue may be prominent or quite subtle (Figure 4).

SM can occur sporadically in an autosomal dominant pattern or as part of a syndrome, including Alagille syndrome (autosomal dominant microdeletion of the JAG1 gene on chromosome 20p causing gastrointestinal, cardiovascular, and facial defects)[7,8] SM has also been observed in association with pachyonychia congenita type II.[6] Mutations of the keratin 17 (K17) gene have been reported in familial cases of SM, although the exact origin of the spontaneous form is not fully understood. The burden of disease can vary from a handful of small cysts to more extensive disease with larger cysts. Cysts typically appear during adolescence and early adulthood, indicating a potential link to hormonal effect, and persist throughout adulthood. The classic differential diagnosis includes epidermal inclusion cysts, milia, trichilemmal cysts, and eruptive vellus hair cysts (EVHC).

Although earlier concepts placed the disease as predominantly afflicting men of Northern European descent between the ages of 19 and 28,[4] current thought has no

gender distinction or national preference. Today, the disease is recognized as having a roughly equal gender distribution, with an average age of onset of 26 years old.[9]

Treatment of SM remains unsatisfactory: cryosurgery, aspiration, excision, incision, lasersurgery, with each having its proponents. Oral retinoids and tetracycline have been tried with minimal success. Similarly, intralesional steroids have some advocates.[7]

CONCLUSIONS

What may seem merely a nuisance to some is quite bothersome to others. While SM is easily recognized, treatment options are not rewarding. The question remains how can we rid our patients of "these gelatinous capsules?"

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Figures

Figure 1: Steatocystoma multiplex. Multiple lesions on the trunk of a young man.
Figure 2: Steatocystoma multiplex. Lesions on the breasts of a young woman.

Figure 3: Steatocystoma multiplex. Photomicrograph showing diagnostic feature of sebaceous gland tissue within the epithelium of a cyst. Steatocystoma simplex shows similar histology. Hematoxylin and eosin, X 710.
Figure 4: Steatocystoma multiplex. Photomicrograph showing diagnostic features of sebaceous gland tissue within the epithelium of a cyst. Only a tiny fragment of sebaceous gland tissue is seen. Steatocystoma simplex may show similar histology. Hematoxylin and eosin, X 710

[i] Günther's paper was originally submitted in 1914, however, it could only be printed after World War I.