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Case Presentation

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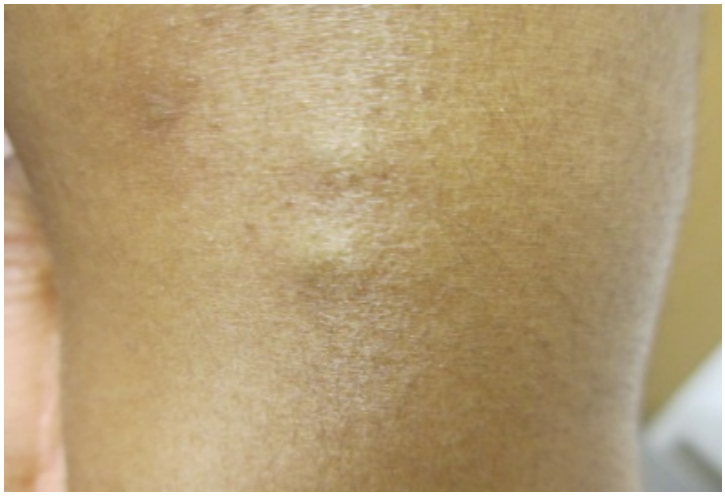
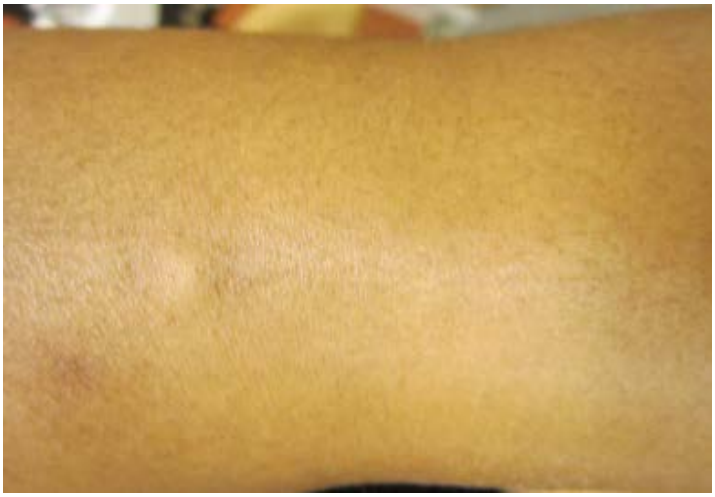
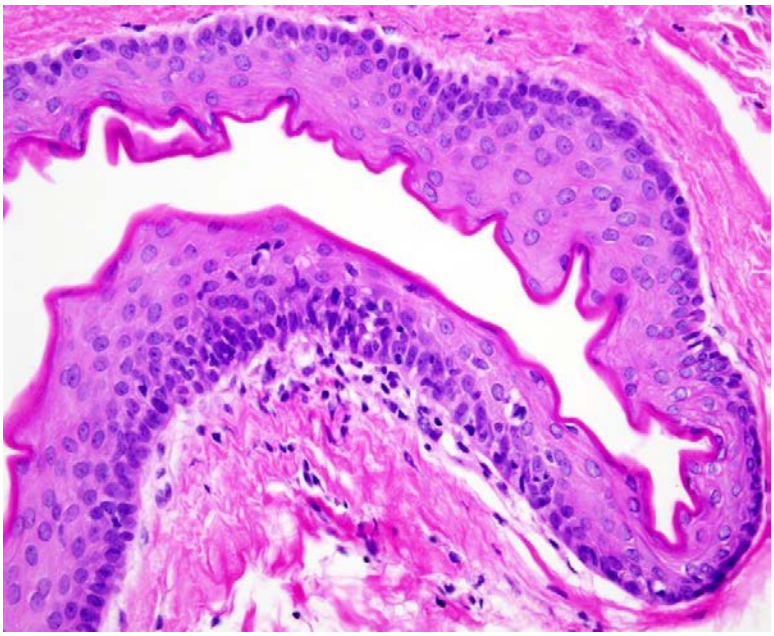
Abstract

Steatocystoma multiplex is a rare condition that is characterized by cutaneous cysts and may be inherited in an autosomal dominant manner or may occur sporadically. The pathogenesis is hypothesized to involve mutations in the keratin 17 gene. There are no internal manifestations. The lesions are usually asymptomatic. However, a suppurative variant exists in which the lesions become inflamed and suppurative after minor trauma. Treatments include cryosurgery, aspiration, surgical excision, laser therapy, and modified surgical incision techniques. This report presents a case of steatocystoma multiplex, the suppurative variant, in a 26-year-old woman with involvement of rare locations on the buttocks, groin, and extremities.







Case synopsis

A 26-year-old woman presented to the NYU Langone Medical Center's Miller Practice for evaluation of small nodules on the body. The patient began to develop tender nodules on the medial aspects of the thighs, buttocks, and groin at the age of 18. She noted numerous smaller nodules on her arms and legs, some of which had been increasing in size and would occasionally become tender and inflamed. These smaller nodules had developed over the past several months prior to presentation. She underwent incision and drainage of an abscess on the right buttock, which drained a yellow-tinged fluid. Doxycycline was started and mupirocin applied to the affected areas on the buttock and nasal passages. After resolution of the acute infection, topical clindamycin solution and benzoyl peroxide wash were started without development of further infection or suppuration of the nodules.

Past medical history was not contributory. There were no known drug allergies. There were no medications. Family history included the patient's mother developing similar nodules in her pre-pubertal years, with involvement of the breasts later in life. Dental history included horizontal wisdom teeth and braces in the patient. The family dental history included multiple dental carries. There was no history of natal teeth.

Physical Examination: There were greater than 100 subcutaneous nodules and skin-colored papules approximately 5-mm in size on the arms, breasts, and posterior aspects of the legs. There was no overlying surface change to the majority of the lesions. The lesions were only made apparent when the overlying skin was pulled taut. Isolated nodules on the buttocks, groin, right arm, and medial aspect of the left thigh were erythematous and tender to palpation. The nail plates were normal. There was no palmoplantar keratoderma. There were no oral lesions and the teeth were normal. The hair appeared normal.

Laboratory Data: Culture of nasal passages, the incision, and drainage fluid from the buttock yielded no growth.

Histopathology: Two punch biopsies were taken of subcutaneous nodules on the right and left arms. There is a cyst lined by a thin, squamous epithelium with a crenulated surface.

Diagnosis: Steatocystoma multiplex, suppurative variant

Discussion: Steatocystoma multiplex (SM) is a condition that is comprised of cutaneous cysts, which may occur in a broad distribution across the body. It is considered a benign, nevoid malformation of the pilosebaceous unit. SM may be inherited in an autosomal dominant fashion, or cases may be non-hereditary (sporadic). Classically, the lesions have appeared most commonly on the chest, axillae, and groin. Most often, the lesions of SM are mobile, non-tender dermal cysts without overlying surface change of the epidermis. There are no associated internal manifestations [1].

The largest case series of SM is a review of 64 patients for demographic, clinical, and histopathologic information. The study reported that the majority of cases were sporadic and that an average age of onset was 26 years (range 4 to 64 years-old). The most common location of lesions was on the arms (35%), chest (29%), axillae (20%), and neck (23%). Lesions were found on the legs in only 12.5% and buttocks in only 3.1% of patients; these are involved locations in our patient. A female-to-male ratio of 1.2:1 was reported [2]. The cysts of SM are usually asymptomatic. However, a suppurative variant exists, in which the cysts become easily inflamed and suppurate owing to minor trauma. This is likely the form of SM that occurred in our patient; her nodules tended to become red and tender in areas prone to trauma or mechanical factors (legs and buttocks). Appropriate treatment for the suppurative form includes incision and drainage as well as antimicrobial therapy [3].

There are reports that describe uncommon associations or locations of SM. A rare subtype of SM, which is termed acral subcutaneous SM, has been described in five patients, who have experienced lesions on the flexor surfaces of the distal aspects of the arms. A female predominance was reported in acral subcutaneous SM, with a female-to-male ratio of 4:1 [4]. SM also has been reported to occur on the scalp, which results in a concurrent acquired alopecia that may have been secondary to trichotillomania [5]. Another rare case included large, firm, disfiguring nodules that were confined to the scrotum, which revealed SM with extensive calcification that resulted in adhesion [6]. Associations have been found between SM and pilar cysts, preauricular sinuses, natal teeth, and trichoblastomas [7-9].

The pathogenesis of SM may involve mutations in the keratin 17 (KRT17) gene, which is a type 1 basic keratin that is expressed in epidermal appendages that include sebaceous glands and hair follicles. This is the same mutation (along with another mutation in keratin 6b) that is found in pachyonychia congenita type 2 (PC-2), which is also an autosomal dominant disorder that is characterized by nail dystrophy, palmoplantar keratoderma, oral leukokeratoses, foillicular keratoses, and epidermal inclusion cysts. SM, natal teeth, pili torti, and vellus hair cysts also are sometimes found in PC-2 [10].

A study analyzed the genetic composition of five patients with SM and found heterozygous missense mutations in KRT17 in all affected individuals. The missense mutations were in the same genetic region or the exact mutations as found in PC-2. This finding raises the question of whether SM and PC-2 are two separate entities or whether they represent different phenotypic expressions along a spectrum of the same disorder [11].

Treatments for SM have included cryosurgery, aspiration, and time-consuming and scarring surgical excisions. Reports have detailed modified techniques for surgical removal of these lesions, which are often numerous, even in the hundreds. Two patients were treated with a radiofrequency device to make 1-to 2-mm incisions followed by expression of the cyst and removal of the cyst wall with forceps. No sutures were used, no scars occurred, and there was no recurrence after five months of follow up [12]. Alternatively, five patients were treated by 2-to 3-mm incision with a blade followed by removal of the cyst using a vein hook (used in ambulatory phlebectomy). The procedure was successful in all five patients, with no side effects and no recurrence over follow-up periods of 14 to 22 months [13]. Lasers have been utilized in the treatment of SM. One patient experienced substantial clearance of lesions after two treatments using the diode laser to target the superficial sebaceous glands and the fractionated erbium-doped fiber laser to target the cystic component in the dermis [14]. Another report details the successful treatment of SM in a patient with PC-2 with the erbium:yttrium-aluminum-garnet laser to create a punctum allowing for drainage of the lesions. There was no scarring or recurrence in three months of follow-up [15]. The carbon dioxide laser has been used for cyst opening, drainage, and wall vaporization, with good cosmetic result and no recurrence over two years of follow up [3].

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