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Steatocystoma Multiplex and Adolescent Development

A Case Report

Nancy M. George, Rebecca Tompkins

ABSTRACT: Steatocystoma multiplex, a benign genetic skin disorder, can have a large developmental impact on an adolescent. The prevalence of this skin disorder is unknown. What is known is that it shows no prevalence based on gender or race/ethnicity. This skin disorder is characterized by numerous small, yellow or skin-colored cystic lesions that occur primarily on the trunk and extremities. Understanding this benign condition, treatment options, both medical and surgical, in the context of adolescent development can assist nurses to intervene where appropriate. Nurses are excellent clinicians for dealing with this type of skin condition that requires not only medical and possibly surgical treatment but also psychological supportive care. For adolescents to transition successfully to adulthood, they must have the coping tools in place that help them deal with this lifelong condition.

Key words: Adolescence, Development, Genetic Skin Condition, Steatocystoma Multiplex

Homelessness in youth is a dire problem, and homeless youth with chronic health issues have difficulty accessing healthcare. In Michigan (United States), there are approximately 3,000 adolescents who are homeless and living on their own (Kids Count, 2012). A local adolescent homeless and runaway shelter provides an 8-hour weekly clinic for its residents. This clinic serves as a primary care site for the residents of the shelter. There are always a multitude of health problems seen at the clinic, but genetic skin diseases like steatocystoma multiplex are rare.

***Nancy M. George, PhD, RN, FNP-BC**, Wayne State University, Detroit, Michigan.*
***Rebecca Tompkins**, BSN, RN, MSN Student Wayne State University, Mott Children's University of Michigan Hospital.*

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Correspondence concerning this article should be addressed to Nancy M. George, PhD, RN, FNP-BC, Wayne State University, 5557 Cass, Room 306, Cohn Building, Detroit, MI 48202.
E-mail: ah1261@wayne.edu

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Although steatocystoma multiplex has long been part of the medical literature, given that it was first described in 1873 by Dr. Jamieson, the developmental and psychosocial implications of the disease are less understood (Davey, Burkhart, & Morrell, 2011; Jamieson, 1873). Steatocystoma multiplex is an inherited autosomal dominant dermatosis characterized by numerous small, yellow or skin-colored cystic lesions (Düzova & Şentürk, 2004). The disease is linked, in most cases, with defects in the keratin 17 gene and has an unknown incidence rate.

Steatocystoma multiplex is a rare disorder of the pilosebaceous unit (Kaya, Ikizoglu, Kokturk, & Tursen, 2001). Lesions are nevoid formations of abortive hair follicles at sites of attached sebaceous glands (Davey et al., 2011). The familial form of steatocystoma multiplex is caused by mutations in the keratin 17 gene that are the intermediate filament proteins, which form a cytoskeletal network within epithelial cells (Smith, 2003). These mutations are identical to those found in clients with pachyonychia congenita type 2 (Kiene, Hauschild, & Christophers, 1996). Keratin 17 is expressed in sebaceous glands, the outer root sheath of hair follicles and the outer nail bed; in fact, steatocystoma multiplex is often associated with eruptive vellus hair cysts (Davey et al., 2011). Both diseases are hereditary and share pubertal age of onset as well as similar lesion appearance. Although some believe that steatocystoma multiplex and eruptive vellus hair cysts are variants of the same disease, there are differences in how the keratin gene is expressed; thus, these are two distinct illnesses. It is thought that a hormonal trigger is responsible for the lesion growth of steatocystoma multiplex because of the relationship between the development of sebaceous glands and the disorder's presentation during puberty (Davey et al., 2011). There has been no established pattern based on gender or race, with an average age onset of 26 years (Cho et al., 2002).

CASE REPORT

Josh G., a 19-year-old African American man, presented to an inner-city clinic for homeless and runaway adolescents with a long history of acne-like lesions on his chest,

face, buttocks, and back. When he lived at home, he had received some treatment for his dermatological condition. He reported that his father had the same problem. Josh reported that, in the past, he had used a lotion that "helped," but there was never resolution of his lesions. In history interview, he said that he was concerned about his skin problem because he was being teased a lot and there was a girl he was interested in but felt that she would not be interested in him until he had his acne under control.

History

Josh was referred to the clinic by his social worker for assessment and treatment of his acne-like lesions. In the case study interview, Josh reported that, since being "kicked out" of his mother's home, he had received no medical treatment for his dermatological condition or any other health problems. Onset of lesions began at 12 years old. Josh stated that lesions were limited in size and distribution. As he aged, he developed more cysts in more places. Josh denied a history of allergies, asthma, or other dermatological problems and denied use of herbal or over-the-counter medications. Josh admitted using marijuana daily until about 4 weeks before he entered the adolescent shelter; he had quit marijuana use because the shelter does not allow any drug use. He denied alcohol consumption or other illegal drug use. Josh was unable to identify any triggers that made his condition worse. He denied being stressed, stating that living in the shelter was far better than being on the street. Josh stated he was trying to change his diet (less junk food), increasing water consumption, and changing body soaps. At the time of the interview, he was completing his GED and hoped to attend a community college for culinary arts. He worked around 10–20 hours a week in a local restaurant. He thought that, perhaps, working in the kitchen around grease was making his skin worse. He shared that he had a "friend with benefits" (someone with whom he had a casual sexual relationship) whom he really liked but felt that she did not really like his skin. He stated that "she do not touch my chest or back much." Josh, upon further questioning, admitted that some people think that he is contagious or that he does not bathe well because of his skin. He stated that he has heard this all before and it does not bother him; however, his body language during the interview contradicted what he was saying. His family history was significant for diabetes mellitus and hypertension, and his father had a similar skin problem. Josh stated that the biggest issue with the lesions is that people make comments about his "acne" and ask why he does not take care of it. He also felt that girls he had "been with" did not like to touch him because of the bumps. Josh reported that he gets depressed sometimes about it but he just tries to "forget about it."

Physical Examination

The clinical presentation for Josh was significant for numerous raised cystic-like lesions over his face, chest, neck, buttocks, and legs. The lesions were ovoid, slightly flocculent, firm, and varied in size from very small to the largest at approximately 1 cm. There was no induration, erythema, or other signs of infection for most of the cystic lesions. A few lesions appeared to have ruptured and had some erythema with no exudate on his chest. The lesions did not follow the lines of Blaschko (pathways that are believed to represent epidermal cell migration and proliferation during fetal development), and they appeared random in pattern. Although, at first glance, this condition could have been acne conglobata, the distribution and palpation of the lesions was very different (see Figure 1).

DISCUSSION

Initial Diagnosis and Treatment

The clinic's nurse practitioner (NP) made an initial diagnosis of dermoid cysts of unknown origin, drew baseline laboratories, and asked Josh to come back the next week. The NP made a personal consultation with a dermatologist, describing the lesions and showing a picture of the youth's torso. A presumptive diagnosis of steatocystoma multiplex was suspected based on familial history, onset, location, and appearance of the lesions. As a first step, the dermatologist suggested antibiotic treatment based on the inflamed cysts and the client's lack of insurance. Josh was started on tetracycline 500 mg, one pill twice daily. The baseline laboratories including liver function markers and lipid panel were completed, and all found within normal limits. As this client lacked health insurance and the NP provider was unable to book an appointment to see a dermatologist within the next 6 months, she made arrangements to have him enrolled in a special program for the uninsured in the clinic's sponsoring health system. A dermatologist agreed to fit the client into his schedule 6 months from the date of the last visit with the youth. Ideally, histopathology of the cysts would have been completed by that time.

Concern for the client's body image and his issues with his skin could not be completely addressed in the short clinic visit. Fortunately, the client was living in a shelter for homeless and runaway adolescents that had a mental health

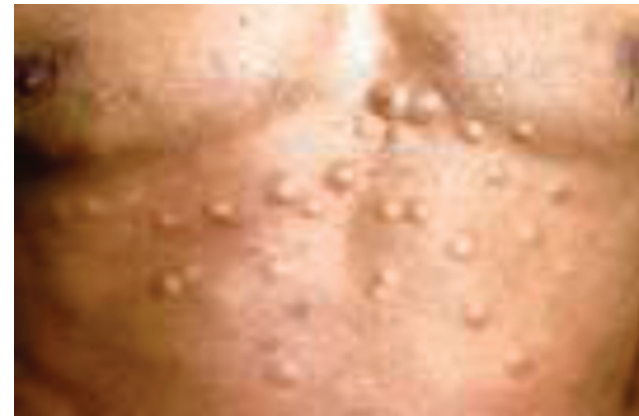

FIGURE 1. Adolescent male with steatocystoma multiplex.

counselor on staff. The client agreed to discuss Josh's feelings about his skin condition with the counselor at the weekly sessions. The NP spoke with the mental health counselor regarding concerns around Josh's expressed depression with his appearance, behaviors, and family issues. The counselor agreed to address these issues with Josh as long as he was living in the shelter.

Adolescent Development

Erickson's stage of psychosocial development associated with adolescence is identity versus role confusion (Erikson, 1968). Within this stage, adolescents truly begin to identify themselves as individuals and imagine how they will fit into society. At this stage of development adolescents become concerned with how they appear to others, with a major part of this feeling related to physical appearance. The lesions of steatocystoma multiplex are numerous and highly visible to others; this visible sign of disease can create self-esteem issues for adolescents. The adolescent may struggle with other people's perception of him or her because of the strange physical appearance. This can cause role confusion and hinder the adolescent from truly discovering his or her place in society (Erikson, 1968). The client of this study had several identified risks related to poor body image stemming from his skin appearance that impacted his development as a late adolescent/young adult. From a family perspective, the role of his mother in his life was an issue for Josh. He came from a family of divorce, with an absent father and a mother who had remarried. This statement can sum Josh's perception of his relationship with his mother: "My mom chose my stepdad over me." He described a contentious relationship with his stepfather that made his continuing to live at home impossible from his mother's viewpoint, so she "kicked me out." Josh's biological father had been incarcerated for most of his adolescence, and Josh rarely saw him, so Josh felt that his friends were his family.

From a developmental perspective, this adolescent's lack of a stable home life and his mother's conditional love combined with a genetic skin disorder had the potential for a long-term impact on his transition into adulthood. One of the milestones of adolescent development is healthy self-image and identity; however, lack of a male role model and a mother who, in the adolescent's eyes, seems more interested in a new husband can delay progress toward this stage. Add a disfiguring, although benign, skin disease to this unstable adolescence, and it is clear that transition to adulthood could be compromised. A condition like steatocystoma multiplex has an important impact on self-image and identity, as it coincides in development with puberty. For a boy who is going through pubertal changes and developing a skin disease that is difficult to treat, the damage to his self-image can be devastating. The physical, developmental, and psychological consequences of a difficult adolescence can be more dramatic and devastating for someone with steatocystoma multiplex. Adolescence is a time of challenging transition where physical development can intersect with psychological development and impact a person's self-image for life. In past research, body image or appearance for boys has not seemed important; however, more recent research suggests that adolescent boys have increasing concerns about their appearance, weight, and fitness (Jones, Vigfusdottir, & Lee, 2004). Josh's steatocystoma multiplex was an added complication for this already at-risk youth. It is easy to see how Josh's issues with self-image and relationships, especially his developing sense of intimacy, have been negatively impacted by his life circumstances.

Intimacy is another developmental milestone and impacts sexual development. Whether viewed within the framework of the biological drive theory or the social construction theory of sexuality as a socially derived and learned behavior, this period of development can be impacted by the highly visible skin changes seen with steatocystoma multiplex. Josh identified feeling uncomfortable about letting any of his girlfriends see him without a shirt on after one of the girls he was involved with made a face about the cysts on his chest. He stated that, until recently, he had not been sexually active and was reluctant to engage in intimate sexual relationships because he did not think that anyone would want to touch him. His current "friend with benefits" relationship reinforced his self-image beliefs, as his friend had not expressed an interest in touching him or his skin. Josh equated her behavior to his skin condition, although he admitted that he had never discussed with his female partner "making the relationship real."

From a developmental perspective, there are many issues to consider when working with adolescents who have steatocystoma multiplex. Consideration must be made for not only what type of physical treatments may be needed but also the possible need for psychological therapy. Thus, from a developmental perspective, there are many issues to be addressed with adolescents who have steatocystoma multiplex, including how it impacts their transition to adulthood.

Differential Diagnosis

Steatocystoma multiplex is known as an acne (conglobata and vulgaris) look-a-like. Its presentation is often confused with acne, but it rarely responds to the latter condition's treatment. The content of a steatocystoma multiplex cystic lesion is sebum and therefore very different in nature from an acne lesion. Steatocystoma multiplex beyond the dermoid cysts can have a nail component, which, when present, assists in making a differential diagnosis. Other differential diagnoses to consider are epidermal inclusion cysts, eruptive vellus hair cysts, follicular infundibulum tumor, Gardner syndrome, lipoma, milia, neurofibroma, sebaceous adenoma, sebaceous hyperplasia, and syringoma. Although many non-hereditary cases have been reported, asking the client about a family history of similar lesions may help to differentiate steatocystoma multiplex from similar diagnoses such as acne conglobata and steatocystoma simplex. Steatocystoma simplex is characterized by solitary cystic nodules with no

family history and is a rare benign cutaneous cyst arising from the pilosebaceous duct junction. It is important in making a differential diagnosis to establish age of onset, location, appearance of lesions, and mode of inheritance.

Signs and Symptoms

The cystic lesions of steatocystoma multiplex are soft, movable, palpable nodules ranging in size from 3 mm to 3 cm (Kaya et al., 2001). When punctured, the cysts secrete a creamy or oily odorless discharge. These cysts are usually asymptomatic; however, they can become inflamed, and spontaneous rupture can form abscesses (Kaya et al., 2001). The lesions can be found anywhere on the body that contain pilosebaceous units but are most commonly seen on the chest, arms, and legs.

Evaluation

Evaluation of this benign condition is based on historical account of onset, family history, and response to self-treatment aided by physical appearance of the lesions. A punch biopsy could be done to confirm diagnosis. Steatocystoma multiplex, as an inflammatory dermal cyst involving the pilosebaceous unit, will show the following indications on histopathological examination: well-encapsulated cysts with folded walls and multiple layers of epithelial cells and a thick granular layer, often a cavity filled with sebum esters (cheesy oily substance), keratin, and sometimes vellus hairs. Often, the diagnosis is based on familial and patient history and physical examination. Genetic testing can be done, but with this benign condition and the cost of genetic testing, it is not routinely performed.

Treatment

Although treatment for this condition is not necessary, clients may decide to have the lesions treated for reasons of appearance, self-image, and self-esteem. The type, number, and location of lesions determine treatment. Any inflammatory version requires antibiotics and, potentially, incision and drainage, and disfiguring lesions may require more surgical treatments. Treatment of steatocystoma multiplex has changed over the last few years with the addition of isotretinoin (Apaydin et al., 2000) to the more widely used therapy of oral antibiotics and excision and incision techniques (Cho et al., 2002; Keefe, Leppard, & Royle, 1992; Schmook, Burg, & Hafner, 2001). Surgical treatments range from cryosurgery, aspiration, and surgical excision to various methods of incisions and carbon dioxide laser (Davey et al., 2011). There is also discussion in the literature of use of a vein hook to eradicate the lesions (Lee, Choe, Park, Lee, & Kim, 2007); however, this approach may be more practical for patients with only a few lesions. Antibiotic therapy is often indicated for the inflammatory version, with use of the tetracycline class of antibiotics. A treatment regimen of tetracycline (Sumycin) 500 mg every 12 hours was given to the client to help with the few lesions he had that were inflammatory. Tetracycline derivatives have been shown to have an anti-inflammatory side effect useful in treating steatocystoma suppurativa. Isotretinoin was used to decrease the size of the sebaceous glands and their sebum production. Currently isotretinoin treatment is used more often because of its effectiveness in decreasing sebaceous gland activity; however, the drug seems to render inconsistent results with steatocystoma multiplex. Treatment with isotretinoin is used with only serious multiple lesions (adult dose: 0.5–1.5 mg/kg/day for 2 months). Discontinuation of isotretinoin is often followed by flare and recurrence of sebaceous dermoid lesions. As with all medications, tetracycline and isotretinoin should not be used if the patient has a hypersensitivity, hepatic dysfunction, or severely elevated triglycerides or if the patient is pregnant or could become pregnant. Since 2006, isotretinoin treatment requires that patients be registered in the FDA-sponsored iPledge program (https://www.ipledgeprogram.com/default.aspx) before prescribing, and only a 30-day supply can be prescribed at a time, followed by a 30-day break from the medication. Women on isotretinoin must be on two forms of birth control because of the teratogenic effect of the medication.

Disfiguring lesions can occur with this type of intradermal lesion, and a proliferation of lesions often makes a surgical approach impractical. If surgical treatment is desired, the cysts are first punctured with a surgical punch or sharp-tipped cautery point for drainage and then removed with electrodessication or clip removal (Davey et al., 2011). Low-dose antibiotics may help to prevent recurrent inflamed lesions, but this treatment is not always successful (Chu, 2003). More recent use of a series of treatments with CO_2 lasers and two complimentary nonablative lasers shows promise for isolated steatocystoma multiplex and may be an option for clients (Moody, Landau, Goldberg, & Friedman, 2012; Rossi, Cappugi, Battini, Mavilia, & Campolmi, 2003).

IMPLICATIONS FOR CLINICAL PRACTICE

Social issues complicated this adolescent's case of steatocystoma multiplex. First, Josh had no health insurance, making referral to a dermatologist difficult. Second, his homeless status made applying for any insurance assistance difficult because a permanent address was required. Finally, the psychological issues of mild depression linked to poor self-esteem and body image would require a more long-term psychological intervention, but this approach was complicated by his homeless status.

CONCLUSION

NPs in a primary care setting should suspect steatocystoma multiplex in clients presenting with intradermal cystic lesions that began in puberty and are located primarily where the pilosebaceous ducts are found and are accompanied by a family history of similar skin issues. The diagnosis is made based on the characteristic clinical features—numerous firm or elastic, flesh- to yellow-colored cystic nodules that can range from 0.2 cm to 3 cm or more in diameter. These are usually found on the trunk, proximal extremities, and

axillae as well as on the neck, scalp, abdomen, and glutei and less often on the face and genitalia. Although a benign uncommon disorder, steatocystoma multiplex can lead to other psychosocial issues for adolescents experiencing this disease; therefore, treatment needs to focus on both the physical and psychological needs of the client. ■

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