Editor's key points

- Steatocystoma multiplex is a benign disorder that presents as papules or nodules on areas with densely concentrated pilosebaceous units including the trunk, arms, axillae, face, thighs, scalp, and less commonly the genitals and breasts. Although the lesions are asymptomatic, with no malignant potential, patients often seek treatment options owing to the psychological distress caused by the cosmetically undesirable lesions.
- Despite the benign and often asymptomatic nature of this disease, an inflammatory variant known as *steatocystoma multiplex suppurativa* can present. The inflamed lesions of steatocystoma multiplex suppurativa pose a great risk of infection, purulent discharge, and scarring.
- Patients should be informed that effective treatment options are time-consuming and can result in scarring, and recurrence is common. Unrealistic expectations and risks of unnecessary treatment should be discussed in depth with this patient population.

Numerous asymptomatic dermal cysts

Diagnosis and treatment of steatocystoma multiplex

Jorge R. Georgakopoulos Arvin Ighani Jensen Yeung MD FRCPC

Abstract

Objective To provide family physicians with the information needed to recognize, diagnose, and discuss available treatment options for steatocystoma multiplex (SM).

Sources of information A comprehensive PubMed search using *steatocystoma multiplex* as either a text word or a MeSH term was conducted, and articles reporting on treatment outcomes were included.

Main message Steatocystoma multiplex is a benign disorder often characterized by numerous asymptomatic dermal cysts on the trunk, arms, axillae, face, thighs, and scalp. Psychological distress due to these undesirable lesions is not uncommon for this condition. A literature review identified the following SM treatments, all of which were associated with limitations: carbon dioxide laser, modified surgical techniques, cryotherapy, and medical management. Steatocystoma multiplex is challenging to treat and, at this time, effective management is most often achieved through patient education.

Conclusion Family physicians play a critical role in the early diagnosis and management of SM. Education about treatment options and managing patient expectations might greatly alleviate the psychosocial implications of this disease.

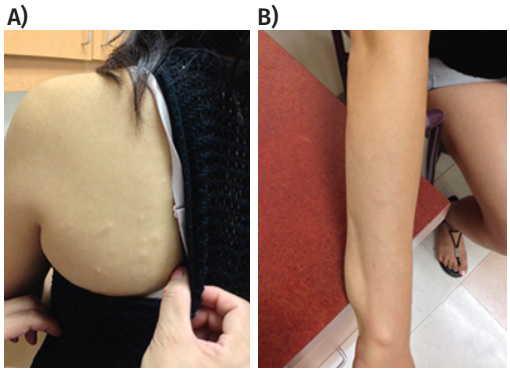
Steatocystoma multiplex (SM) is a benign disorder of the pilosebaceous unit, manifesting as multiple asymptomatic dermal cysts. Although sporadic presentation is most often reported, it is believed to have an autosomal dominant pattern of inheritance involving a keratin 17 mutation.[1] The following described case and review of treatment options can be used as a learning tool for how to recognize, diagnose, and manage SM. Early disease recognition and patient education might play a critical role in alleviating psychological implications associated with this disease.

Case description

A 22-year-old woman presented to an academic dermatology clinic concerned about numerous bumps on her arms, thighs, and trunk. She was referred by her family doctor to confirm the diagnosis of SM and discuss treatment options for these cosmetically bothersome lesions. Physical examination revealed numerous yellow- to skin-coloured, firm, freely mobile papules and nodules on her arms, thighs, and trunk that had slowly enlarged and increased in number over 5 years (**Figures 1A** and **1B**). Her family history was not relevant for similar lesions and no other cutaneous findings were identified at the time of presentation.

While her history and physical examination findings strongly suggested the diagnosis of SM, the patient requested a biopsy to obtain a definitive answer. Two punch biopsies were taken of subcutaneous nodules using a 4-mm full-thickness punch at 2 separate sites. Histopathology revealed

Figure 1. Steatocystoma multiplex: *Numerous skin-coloured, firm papules and nodules on A) the back and B) the forearm of a female patient.*



encapsulated cysts lined by several layers of epithelial cells folded in an irregular manner. The cystic lumen was lined with a thick eosinophilic band. These findings confirmed the diagnosis of SM. The contents of the cyst could not be elucidated by the pathologist; however, vellus hairs and hair follicles are commonly observed under microscopy.[2]

Sources of information

A review of the PubMed database was conducted and relevant studies were retrieved using *steatocystoma multiplex* as either a text word or a MeSH term on March 20, 2017. The initial search yielded 231 articles for screening (**Figure 2**). After abstract and full-text review, primary articles (ie, case reports or case series) reporting treatment outcomes for SM were identified for inclusion in this review of treatment options. In addition, manual reviews of citation lists for relevant articles were performed.

Main message

Common presentation. Steatocystoma multiplex papules or nodules commonly present on areas with densely concentrated pilosebaceous units including the trunk, arms, axillae, face, thighs, scalp, and less commonly the genitals and breasts.[3-6] It most frequently presents in adolescence or early adulthood, with a mean age of 26 years at the time of diagnosis, and has no predilection for sex.[7] Although the lesions are asymptomatic, with no malignant potential, patients often seek treatment options owing to the psychological distress caused by the cosmetically undesirable lesions. The case described here represents the classic presentation of SM and highlights the important role family physicians can play in early disease recognition.

Diagnosis. Although SM shares similar characteristics with other diseases, its diagnosis is often made through history and physical examination alone.[8] Pathologic analysis of sebaceous cysts, such as those seen in SM, provides little value when there is a low index of concern for malignancy. For this reason, a diagnosis of SM based on clinical findings is often warranted unless clinical suspicion suggests otherwise.[8] If a biopsy is required, multiple samples should be taken because variations in the classic histopathologic features of SM have been described.[2] A 4-mm full-thickness punch biopsy is recommended.

Evaluation and differential diagnosis. Despite the benign and often asymptomatic nature of this disease, an inflammatory variant known as *steatocystoma multiplex suppurativa* (SMS) can present.[2] This is seen in a male patient who presented to the clinic with numerous SM lesions on his chest, some of which were inflamed (**Figure 3**). The inflamed lesions of SMS pose a great risk of infection, purulent discharge, and scarring.[9] A mixed presentation of asymptomatic and inflamed lesions is common. Additionally, SM might mimic other common skin disorders, sometimes leading to misdiagnosis and unnecessary treatment. The clinical appearance of SM can resemble acne vulgaris, vellus cyst, epidermoid or dermoid cyst, hidradenitis suppurativa, milia, follicular infundibular tumours, and lipomas.[10]

Treatment options. Although SM is difficult to treat, having a strong understanding of all available treatment options, with a focus on recurrence and cosmetic outcomes, is crucial for patient education and disease perception. A total of 37 relevant publications describing treatment outcomes of SM patients were identified and are summarized in **Table 1**.[3-5,9,11-43] Goals of SM treatment include substantial reduction of cyst size, prevention of recurrence, good cosmetic outcome, and patient satisfaction. At this time, no treatment can prevent the formation of new lesions. Positive outcomes are often challenging to achieve, as the described treatment methods are associated with limitations. Effective management is most often achieved through patient education. However, recent advanced techniques include the following.

Carbon dioxide laser: Carbon dioxide laser has recently been shown to successfully treat multiple lesions in a single session with good cosmetic outcomes. However, this procedure might not be suited for larger cysts and is not easily accessible to all patients.[11,12] This laser method is comparable to excising the lesion, as the wavelength of the laser ablates skin tissue.

Modified needle aspiration: Needle aspiration with gentle extirpation of cystic contents has proven successful, with excellent cosmetic outcomes.[5,13,14] Despite good results, this treatment requires a skilled operator and does not work well on very large (>15 mm in diameter)

Figure 2. Flowchart of study selection process: *Review of treatment options for steatocystoma multiplex.*

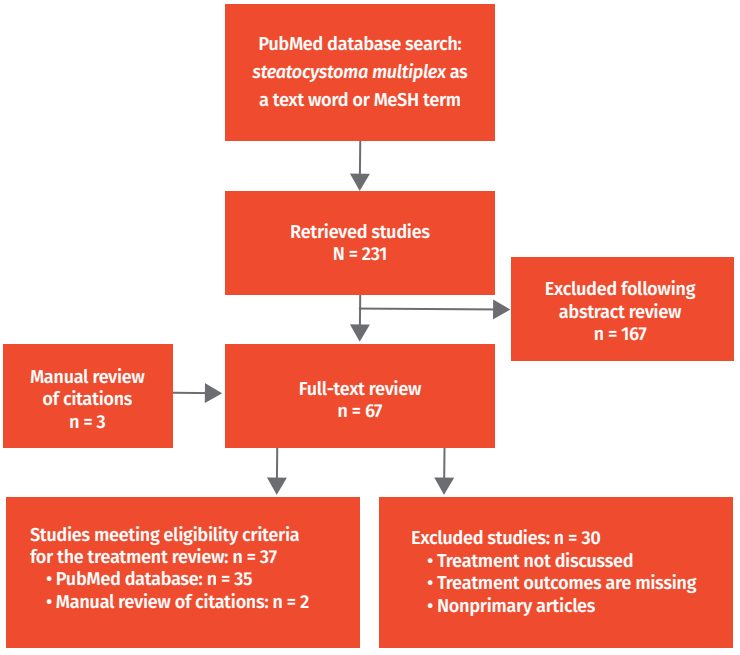
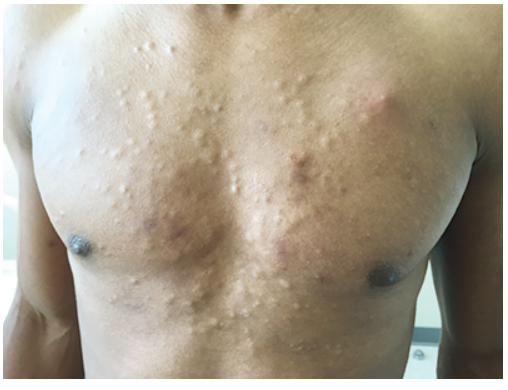


Figure 3. Steatocystoma multiplex suppurativa: *Mixed presentation of steatocystoma multiplex with suppurativa inflammatory variant involvement on the chest of a male patient.*



or small (<3 mm in diameter) cysts. Recurrence rates are extremely high for this treatment method.

Modified surgical techniques: Surgical techniques, including fine incision followed by cyst wall extraction with forceps, vein hooks, or curette, show excellent cosmetic results but are time-consuming and invasive.[4,15-17]

Cryotherapy: Cryotherapy allows for the treatment of multiple lesions in a single session; however, it is greatly limited owing to the cosmetic disfigurement it causes and its extremely low efficacy.[9,18]

Medical management: Medical management with oral isotretinoin is the preferred treatment for SMS and it provides great reduction in inflammation.[9,19,20] However, results are often not seen for months and recurrence following discontinuation has been reported.[21,22] Mixed variants of both SM and SMS might require combination therapy.[9,18] Isotretinoin has no effect on noninflamed lesions.[20] A short course (2 to 4 weeks) of oral tetracycline, topical clindamycin, or benzoyl peroxide wash (ie, antibiotics with anti-inflammatory properties) might be considered for management of noninfectious inflammatory lesions.[21,36]

Case resolution

Two weeks after the biopsies were taken, the patient was seen in the clinic for consultation regarding her confirmed diagnosis of SM. After educational guidance, she was aware that this condition posed no medical risk and that treatment was not required or recommended. Owing to the extensive number of lesions and evidence showing that effective treatment often leads to scarring, the patient elected to forgo therapy at this time. She was instructed to return for follow-up should any changes occur, including extensive growth, discoloration, or severe inflammation suggesting infection.

Table 1. Summary of available publications describing treatment outcomes for SM, by treatment

REFERENCE	NO. OF PARTICIPANTS	SM OR SMS	LOCATION OF CYSTS	TREATMENT DESCRIPTION	BENEFITS	LIMITATIONS
				Laser		
Kassira et al, 2016[11]	1	SM	Face	Fractionated ablative CO_2 laser	• Led to spontaneous expression of cystic content • No recurrence at 3 y • Less time-consuming procedure • Minimal risk of scarring	• Treated cysts were small and localized to the temple
Bakkour and Madan, 2014[12]	8	SM	Chest, back, and axilla	CO_2 laser incision and cyst removal with Volkmann spoon	• Good clinical improvement • Minimally invasive, allowing for multiple treatments in 1 session	• Minimal scarring and little recurrence
Moody et al, 2012[13]	1	SM	Abdomen and lower chest	Targeted laser treatment of the sebaceous glands and dermal cysts	• Good improvement (75% reduction) • Noninvasive • Allows for treatment of multiple lesions	• None reported
Varshney et al, 2011[14]	1	SM	Head and neck	CO_2 laser ablation	• Good cosmetic results • No recurrence at 18 mo	• None reported
Mumcuoğlu et al, 2010[15]	1	SM	Chest, forehead, axillae, and knees	Er:YAG laser and drainage	• Good cosmetic results with no scarring • No recurrence at 3 mo	• None reported
Madan and August, 2009[16]	1	SM	Back, chest, and abdomen	CO_2 laser incision and cyst removal with Volkmann spoon	• Minimally invasive	• Postinflammatory hyperpigmentation
Riedel et al,* 2008[17]	1	SM	Forehead and cheeks	Laser and surgical: incision, cyst removal with sharp spoon, followed by CO_2 vaporization of remaining content	• No recurrence at 8 mo • Good clinical improvement	• None reported
Rossi et al, 2003[18]	1	SM	Forehead, eyelids, and neck	CO_2 laser and cyst removal with forceps	• Minimally invasive with quick healing • Good cosmetic results with no recurrence at 2 y • Can treat multiple lesions without anesthesia	• None reported
Krähenbühl et al, 1991[19]	1	SM	Trunk	Focused CO_2 laser incision followed by defocused laser therapy of cyst wall	• Good clinical improvement • Quick healing	• Scar formation
				Surgical		
Kumar et al, 2014[20]	1	SM	Malar region	Wide local surgical excision	• Total excision and no recurrence	• Scar formation • Requires anesthesia • Limited to treating localized regions
Gordon Spratt et al,* 2013[21]	1	SMS	Thigh, buttocks, groin, arms, and legs	Surgical and antimicrobial therapy: incision and drainage followed by topical clindamycin solution and benzoyl peroxide wash	• Can treat grossly enlarged and infected nodules	• None reported

Table 1 *continued on page 896*

Table 1 *continued from page 895*

REFERENCE	NO. OF PARTICIPANTS	SM OR SMS	LOCATION OF CYSTS	TREATMENT DESCRIPTION	BENEFITS	LIMITATIONS
				Surgical		
Choudhary et al, 2010[4]	2	SM	Scrotum	Incision using a radiofrequency instrument, drainage, and cyst extraction with forceps	• Complete removal and no recurrence at 5.5 mo • No scarring or postinflammatory hyperpigmentation • Provides a bloodless field • Able to treat multiple cysts in 1 sitting	• None reported
Lee et al, 2007[22]	5	SM	Not reported	Incision and vein hook cyst removal	• Complete removal and no recurrence at 14-30 mo • Faster than other surgical techniques (1 min/cyst)	• Mild transient hyperpigmentation: resulted in satisfactory cosmetic outcome
Ichikawa et al, 2006[23]	1	SM	Face	Forehead flap and cyst extraction	• Complete removal • Scar formation hidden by hairline	• Recurrence 16 mo postoperatively • Invasive and surgical risks
Düzova and Şentürk, 2004[24]	2	SM	Face	22-gauge needle aspiration	• Very good cosmetic outcome • No recurrence at 10 mo	• Challenging to extirpate the dense contents of larger cysts • Risk of hematoma
Kaya et al, 2001[25]	1	SM	Chest, neck, axilla, inguinal folds, and inguinal regions	Puncture with sharp-tipped cautery point, drainage, and cyst removal with forceps	• Good clinical improvement • No recurrence at 14 mo	• Hypopigmented macules and superficial depressions • Requires multiple sessions
Schmook et al, 2001[26]	5	SM	Not reported	Incision, drainage, and cyst wall removal with curette followed by forceps	• Virtually unnoticeable scarring • No recurrence at incision site	• None reported
Adams et al, 1999[27]	1	SM	Chest and neck	Incision and cyst removal with small artery forceps	• No visible scarring • No recurrence at 4 mo	• Requires multiple sessions
Oertel and Scott, 1998[5]	3	SM	Arm, forearm, chest, neck, axilla, and breast	22-gauge needle aspiration	• No scar formation • Minimally invasive • Inexpensive procedure	• Tedious, requiring precision and a skilled technique
Pamoukian and Westreich, 1997[28]	7	SM	Head and neck	Incision and cyst removal with mosquito hemostat	• Good clinical improvement	• Recurrence of 10% of lesions • Time-consuming • Requires anesthesia
Kanekura et al, 1995[29]	1	SM	Scalp, forehead, and chest	3-mm biopsy punch, drainage, and cyst removal with forceps	• Complete removal • Wounds healed in 10 d • No recurrence at 1 y	• Time-consuming and not feasible to remove all lesions in 1 sitting (only 2 removed)
Sato et al, 1993[30]	1	SM	Head, neck, trunk, and upper extremities	Aspiration and scraping with a syringe connected to an 18-gauge needle	• Reduction in number and size of cysts • Much improvement in psychological condition	• Aspiration does not work on smaller cysts

Table 1 *continued on page 897*

Table 1 *continued from page 896*

REFERENCE	NO. OF PARTICIPANTS	SM OR SMS	LOCATION OF CYSTS	TREATMENT DESCRIPTION	BENEFITS	LIMITATIONS
				Surgical		
Keefe et al, 1992[31]	1	SM	Neck, forearms, behind the ears, over the scapula, chest	Surgical blade puncture, drainage, and cyst removal with forceps	• Good clinical improvement • Minimal recurrence	• Time-consuming, requiring multiple surgeons • Requires anesthesia • Scar formation
Feinstein et al, 1983[32]	1	SM	Scalp and forehead	Excision and skin graft	• No recurrence at 15 y	• Inadequate cosmetic outcome
Holmes and Black, 1980[33]	1	SM	Face, trunk, and axillae	Hairline flap and cyst extraction	• No recurrence at 4 y	• Incomplete clearance • Invasive and surgical risks
Egbert et al, 1979[34]	1	SMS and SM	Entire body surface	Incision, drainage, and electrocautery	• None reported	• Required anesthesia and multiple visits to the operating room
				Medical management		
Lima Santana et al, 2016[35]	1	SMS	Axillary regions, inguinal region, trunk, lower limbs, antecubital fossae, face, and scalp	Isotretinoin	• Stabilized condition with no new or worsening lesions at 3 mo	• Had minimal effect in decreasing the size or number of lesions
Adams and Shwayder, 2008[36]	1	SMS	Face, scalp, trunk, and extremities	Tetracycline	• Cleared infected lesions on legs	• Noninfected lesions persisted
Moritz and Silverman, 1988[37]	1	NA	Not reported	Isotretinoin	• Shrinkage of lesions that persisted at 6 mo	• Delayed response: 2 mo after discontinuing
Friedman, 1987[38]	1	SM	Not reported	Isotretinoin	• None reported	• Provided no improvement
Rosen and Brodkin, 1986[39]	1	SMS	Not reported	Isotretinoin	• Inflamed cysts markedly improved	• Recurrence of cysts 8 wk into therapy
Statham and Cunliffe, 1984[40]	3	SMS and SM	Trunk and limbs	Isotretinoin	• Substantial improvement in inflamed lesions	• No effect on noninflamed lesions
Schwartz and Goldsmith, 1984[41]	1	SMS	Not reported	Isotretinoin	• Abscesses involuted and inflamed cysts decreased in size • Persisted at 10 wk after discontinuing therapy	• Cysts returned after discontinuing treatment
				Other		
Kamra et al,* 2013[3]	1	SM	Chest, breast, axilla, inguinal region, and extremities	Radiofrequency probe and isotretinoin	• None reported (suggested to provide a bloodless field)	• None reported

Table 1 *continued on page 898*

Table 1 *continued from page 897*

REFERENCE	NO. OF PARTICIPANTS	SM OR SMS	LOCATION OF CYSTS	TREATMENT DESCRIPTION	BENEFITS	LIMITATIONS
			Other			
Fekete and Fekete,* 2010[42]	1	SMS and SM	Entire body surface	Cryotherapy and isotretinoin	• Slight regression and healing of lesions	• Local disfigurement, hyperpigmentation, and unpleasant scars
Apaydin et al,* 2000[9]	1	SMS and SM	Entire body surface	Cryotherapy and isotretinoin	• Isotretinoin cleared inflamed lesions, which did not reappear	• Cryotherapy caused scarring and hypopigmentation
Notowicz, 1980[43]	NA	SM	Not reported	Cryotherapy: necrotic tissue and cyst content removed with pressure 3-4 d later	• Treatment of numerous lesions in 1 sitting	• Extensive scar formation

CO_2—carbon dioxide, Er:YAG—erbium:yttrium-aluminum-garnet, NA—not available, SM—steatocystoma multiplex, SMS–steatocystoma multiplex suppurativa. *Combination therapy.

Conclusion

Owing to limited evidence for effective treatment methods, SM is a therapeutically challenging diagnosis. The ability to identify multiple asymptomatic dermal cysts and recognize the characteristic pattern of multiplication and progressive growth seen in SM can allow for early education about the benign nature of this disease. Furthermore, a concrete understanding of treatment options, including risks and benefits, can play a critical role in managing the psychosocial implications of SM. Patients should be informed that effective treatment options are time-consuming and often result in scarring, and recurrence is common. Unrealistic expectations and risks of unnecessary treatment should be discussed in depth with this patient population, as these individuals might desire unattainable outcomes.

Mr Georgakopoulos is a medical student in the Schulich School of Medicine and Dentistry at Western University in London, Ont. **Mr Ighani** is a medical student at the University of Toronto in Ontario. **Dr Yeung** is a lecturer and staff dermatologist in the Division of Dermatology at the University of Toronto.

Contributors
All authors contributed to the literature review and interpretation, and to preparing the manuscript for submission.

Competing interests
None declared

Correspondence
Dr Jensen Yeung; e-mail **jensen.yeung@utoronto.ca**

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