

## BRIEF ARTICLES

**Steatocystoma Multiplex: Case Report and Review of Treatment**Mina Amin BS<sup>a</sup>, Peter Hashim MD MHS<sup>b</sup><sup>a</sup>University of California, Riverside, School of Medicine, Riverside, CA<sup>b</sup>Department of Dermatology, Icahn School of Medicine at Mount Sinai, New York, NY**ABSTRACT**

Steatocystoma multiplex, an uncommon autosomal dominant disorder, is characterized by numerous asymptomatic skin-colored cystic nodules. The leading hypothesis regarding the pathophysiology of this disease involves a defect at the pilosebaceous duct junction, which leads to cyst formation. Herein, we describe a case of a male patient that presented with steatocystoma multiplex. We examine the cosmetic outcomes of different techniques used in his treatment as well as review the varied therapeutic options available to such patients, including surgical removal, needle aspiration, cryotherapy, isotretinoin, and CO2 laser treatment.

**INTRODUCTION**

Steatocystoma multiplex (SM) is a rare cutaneous disorder characterized by multiple asymptomatic skin-colored nodules.<sup>1</sup> Although SM can involve any portion of the body, areas with a high number of sebaceous glands, including the neck, trunk, axilla, groin, scalp, and proximal extremities are commonly affected.<sup>1,2</sup> The inheritance pattern is considered to be autosomal dominant, although sporadic cases of SM have also been reported. The incidence of SM is evenly distributed across genders.<sup>2</sup> Given that SM typically appears during adolescence, variations in hormone levels during puberty may trigger the development of the disease.<sup>2</sup> In a subtype of SM, referred to as steatocystoma multiplex suppurativa, cysts can become inflamed and

subsequently rupture, leading to secondary infection and abscess formation.<sup>1</sup> Keratin 17 is an intermediate filament found in the epithelial cells of the nail bed, hair follicle, and sebaceous gland.<sup>3</sup> Pachyonychia congenita (specifically, pachyonychia congenita KRT17, formerly known as pachyonychia congenita type 2) and SM have both been associated with a mutation in the gene encoding keratin 17. Moreover, SM has been associated with eruptive vellus hair cysts and some authors consider these two entities to be on the same spectrum of pathologic disease processes.<sup>4</sup>

## CASE REPORT

A 26-year-old male presented with cysts involving multiple areas of the body since 12 years of age. The cysts first appeared on his chest, and then subsequently involved his neck, waist, and abdomen. No treatment was used in the past. A biopsy performed five years to his presentation revealed a diagnosis of steatocystoma multiplex. His past medical history was noncontributory. There was no family history of similar lesions. On examination of the skin, multiple, firm, mobile, skin-colored nodules varying

from 0.5-3 cm in size were observed on the chest, neck, abdomen, and arms (Figures 1 & 2).

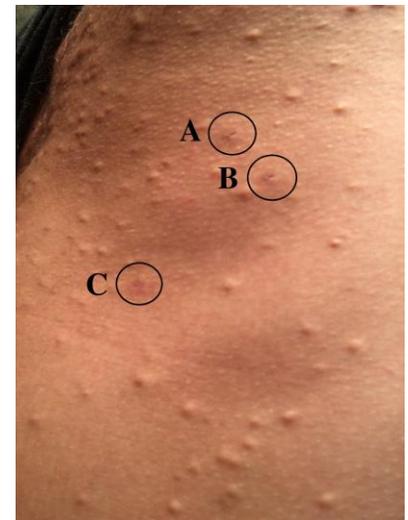
Three of the cysts were chosen to each undergo a different treatment: simple incision and drainage (labeled A), incision and drainage followed by electrodesiccation (labeled B), and incision and drainage followed by the application of trichloroacetic acid (labeled C). The patient presented for follow-up at 2 weeks, and his healing wounds were evaluated for cosmetic outcomes (Figure 3).



**Figure 1.** Multiple soft cysts on the neck and chest.



**Figure 2.** Numerous cysts are located on the lateral side of the chest.



**Figure 2.** Cyst removal by incision and drainage was followed by (A) no further treatment, (B) electrodesiccation, or (C) trichloroacetic acid treatment, respectively. At two week follow-up, all three lesions were healing well.

## DISCUSSION

Multiple different treatment options have been described for SM. Cryotherapy has been associated with scarring, blistering, hyperpigmentation, hypopigmentation, and poor cosmetic outcomes.<sup>1-2</sup> However, a combination of cryotherapy to non-suppurative lesions in a patient with SM suppurativa has also been reported with successful cosmetic results.<sup>1</sup>

Inflamed and suppurative lesions respond well to isotretinoin; however, non-inflamed cystic lesions remain unchanged after isotretinoin treatment.<sup>5</sup> The anti-inflammatory effect of isotretinoin likely accounts for this finding. One exacerbation of SM with severe inflammation after isotretinoin use has also been reported.<sup>6</sup> Nevertheless, patients with inflamed cystic lesions could potentially benefit from isotretinoin treatment.<sup>5</sup>

The extent of involvement of the condition can make surgical removal of multiple lesions challenging. Madan et al. used CO<sub>2</sub> laser to perforate the cyst wall.<sup>7</sup> A Volkmann's spoon was used to evacuate the cyst and the cyst healed by secondary intention and the cosmetic outcome was favorable. Rossi et al. also utilized CO<sub>2</sub> laser to perforate the cyst wall of lesions on the face, with subsequent mechanical evacuation of the cyst contents or the cyst walls of small cysts were vaporized.<sup>8</sup> CO<sub>2</sub> laser therapy is a minimally invasive option for treating multiple lesions that provides favorable aesthetic outcomes and a low recurrence rate.<sup>8</sup>

Choudhary et al. described the use of a radiofrequency instrument to create tiny incisions into the cysts under local anesthesia.<sup>9</sup> Subsequently, the content of

the cysts were manually expressed with the thumb and the index finger. Forceps were then used to grip the wall of the cyst and the sac of the cyst was extracted through small incisions. The lesions healed by secondary intention without any scar or postinflammatory hyperpigmentation. Follow-up approximately 6 months later demonstrated favorable cosmetic outcome.

Lee et al. utilized a No. 11 blade to make a small incision in the cyst, a vein hook was then used to pull the cyst out, followed by the use of mosquito forceps to completely remove the contents of the cyst.<sup>10</sup> Transient post-inflammatory hyperpigmentation was present but recurrence did not occur.

Oertel et al. reported the use of fine needle aspiration in three patients with no scarring.<sup>11</sup> Egebert reported a severe case with extensive cysts in which hospitalization, oral antibiotics, incision and drainage, and electrocautery under general anesthesia.<sup>12</sup>

According to the literature, this is the first report that directly compares the outcomes of three different treatments performed for a patient with SM. In our case, cyst removal by simple incision and drainage (A) and incision and drainage followed by electrodesiccation (B) both yielded the best cosmetic outcomes at two-week follow up (Figure 3). The patient noted that the lesion treated with trichloroacetic acid application (C) after incision and drainage was associated with increased irritation relative to the other two lesions (Figure 3). Further follow-up will be needed to evaluate for possible recurrences using these different techniques.

For the remaining lesions, multiple treatment options were discussed with the patient. Incision and drainage of all of lesions was considered impractical due to the extent of

involvement of his skin lesions and potential risk for scarring. As a result, the patient decided not to undergo surgical removal of his remaining lesions. Instead, he intends to receive treatment for his remaining lesions with CO2 laser therapy.

Despite the benign nature of SM, the quantity and extent of lesions can negatively impact the quality of life of these patients. Multiple treatment options are available, but there lacks a consensus on optimal treatment. Disease burden and patient preference remain important factors in pursuing therapeutic strategies. Thus, a discussion with patients regarding the benefits and limitations of each therapeutic option available is essential before formulating a personalized treatment plan that is ultimately tailored to the individual patient.

**Conflict of Interest Disclosures:** none.

**Funding:** none.

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