

# An Atlas of Lumps and Bumps, Part 45: Steatocystoma Multiplex

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## Steatocystoma Multiplex

Steatocystoma multiplex is a hamartomatous malformation of the pilosebaceous glands characterized by multiple skin-colored sebum-containing dermal cystic papules and nodules.<sup>1-3</sup> The condition frequently presents in adolescence and early adulthood.<sup>3-6</sup> Occasionally, it can appear earlier only to worsen at puberty, when the activity of the sebaceous glands is elevated.<sup>4</sup> Steatocystoma multiplex has been described at birth and in patients in their eighth decade.<sup>2,3,7</sup> There is no sex predilection.<sup>2,6,8</sup>

Steatocystoma multiplex is a rare skin condition; the exact incidence is not known. Familial cases are often caused by mutations in exon 1 of the keratin 17 (*KRT17*) gene located on chromosome 17q21.2 and have an autosomal dominant mode of inheritance.<sup>9-12</sup> The majority of cases are sporadic.<sup>8,13,14</sup> Androgenic stimulation of the sebaceous glands, genetic predisposition, along with environmental factors influence the onset of steatocystoma multiplex. The condition has been reported following the use of ustekinumab which might have unmasked a genetic predisposition.<sup>15</sup> Hypertrophic lichen planus, eruptive vellus hair cysts, pachyonychia congenita type 2 (Jackson-Lawler syndrome), hypohidrosis, congenital alopecia, acrokeratosis verruciformis, hidradenitis suppurativa, hypothyroidism, preauricular sinuses, and natal teeth occur with increased frequency in patients with steatocystoma multiplex.<sup>5,16-19</sup>

Clinically, steatocystoma multiplex is characterized by multiple, asymptomatic, smooth, round, soft to firm, often freely movable, yellow to skin-colored papules and nodules (**Figure 1**).<sup>1,7,8</sup>



**Figure 1.** *Steatocystoma multiplex* is characterized by multiple, asymptomatic, smooth, round, soft to firm, often freely movable, yellow, skin-colored papules and nodules.

Lesions tend to be a few millimeters to a centimeter in diameter and are slow growing.<sup>1</sup> Superficial lesions are usually yellowish while deeper lesions skin-colored (**Figure 2**).<sup>1,8</sup> The overlying epidermis is normal with no central punctum present.<sup>1,8,20</sup> The content is usually oily or creamy. Sites of predilection include the trunk (especially the presternal area), neck, axillae, proximal extremities, and groin where high numbers of sebaceous glands are found.<sup>1,4,7,21</sup>



**Figure 2.** Sites of predilection for *steatocystoma multiplex* include the trunk, particularly the presternal area, as well as the neck, axillae, proximal extremities, and groin where high numbers of sebaceous glands are found.

Rarely, the face, scalp, breasts, and genital area are affected.<sup>1,2,9,20-22</sup> There is usually little or no nail or hair involvement. There are no systemic manifestations.<sup>1,14</sup>

Several variants exist. In the suppurative variant, the lesions become inflamed and suppurative after minor trauma.<sup>14</sup> In the acral variant, the lesions appear on the distal upper or lower extremities.<sup>7,15</sup> The female to male ratio is 4:1 in the acral variant.<sup>3</sup> Localized forms of *steatocystoma multiplex* have also been described, such as cases with a cephalic or genital localization.<sup>23</sup>

The diagnosis is usually clinical, based on typical physical findings. There are no abnormal laboratory findings. Skin biopsy should be considered if the diagnosis is in doubt.

*Steatocystoma multiplex* is cosmetically unsightly and may have an adverse impact on quality of life.<sup>18</sup> Occasionally, the lesions may rupture, become infected and painful, and heal with scarring.<sup>6,9,24</sup> Rarely, the lesions may calcify.<sup>8</sup> Malignant degeneration has rarely been described.<sup>25</sup> Without treatment, the lesion tends to persist and may slowly enlarge.

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**EDITOR'S NOTE:**

This article is part of a series describing and differentiating dermatologic lumps and bumps. To access previously published articles in the series, visit: <https://www.consultant360.com/resource-center/atlas-lumps-and-bumps>.

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