Ultrasound Morphologic Features of Steatocystoma Multiplex With Clinical Correlation

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The ultrasound features of 87 steatocytoma multiplex (SCM) lesions detected in 9 patients are reported. Steatocytoma multiplex is a hamartomatous condition derived from the pilosebaceous duct junction that generates multiple cutaneous cystic lesions. It appeared as clusters of well-defined hypoechoic nodules with mild posterior enhancement in 100% of cases, with both dermal and subcutaneous locations in 67%. No calcification foci were detected within or at the periphery of the lesions. Fifty-six percent of the cases showed signs of hypervascularity in the edge of the nodules, and 44% of the lesions were associated with another dermatologic condition, most frequent being hidradenitis suppurativa (75%), followed by vellus hair cysts (25%). Steatocytoma multiplex shows ultrasound features that allow discrimination from other common cutaneous entities.

Key Words—dermatologic ultrasound; hidradenitis ultrasound; skin ultrasound; steatocytoma multiplex ultrasound; steatocytoma ultrasound

Steatocystoma multiplex (SCM) is a rare hamartomatous condition derived from the pilosebaceous duct junction that generates multiple cutaneous cystic lesions.[1] These can occur sporadically or under a genetic form. An autosomal dominant inheritance pattern[2,3] and a mutation located in the keratin 17 gene have been identified.[4,5]

Clinically, this entity is characterized by dome-shaped papules and asymptomatic pink or yellowish nodules, most often located in the upper extremities (35%), followed by the thorax (29%), armpits (20%), and neck (23%).[3] On pathologic specimens, steatocystomas have an eosinophilic cuticle with no granular layer and a variable amount of hairy tracts, keratin, sebum, sebaceous glands, and fragments of smooth muscle.[3]

To date, the use of ultrasound (US) in steatocystomas has been described in a few case reports, mainly in breast locations.[6,7] The objective of this work was to analyze the US appearance of SCM and look for US patterns that can support its diagnosis.

Materials and Methods

We retrospectively reviewed the clinical and US data of 9 patients with a confirmed histopathologic diagnosis of SCM from January 2010 to December 2017. All examinations were performed with color Doppler US equipment (LOGIQ E9 XD Clear; GE Healthcare, Milwaukee, WI) using linear and compact linear (hockey stick) variable-frequency transducers, with upper ranges of

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Abbreviations

HS, hidradenitis suppurativa; SCM, steatocystoma multiplex; US, ultrasound

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15 and 18 MHz, respectively. All examinations were performed by the same radiologist, who was trained in dermatologic US, and according to the published protocol for dermatologic US examinations.[8]

The diameters of the lesions were measured in millimeters, and their volumes were calculated by the ellipsoid method based on the formula volume = length × width × height × 0.52, obtained from images showing the lesions in two perpendicular axes. A descriptive analysis of the findings was performed. All cases were studied under the Helsinki principles of medical ethics. The Institutional Review Board (Hospital Clinico Universidad de Chile) waived the need for signed informed consent; nevertheless, according to the protocol for all of the US examinations, all patients signed permission for the publication of their images.

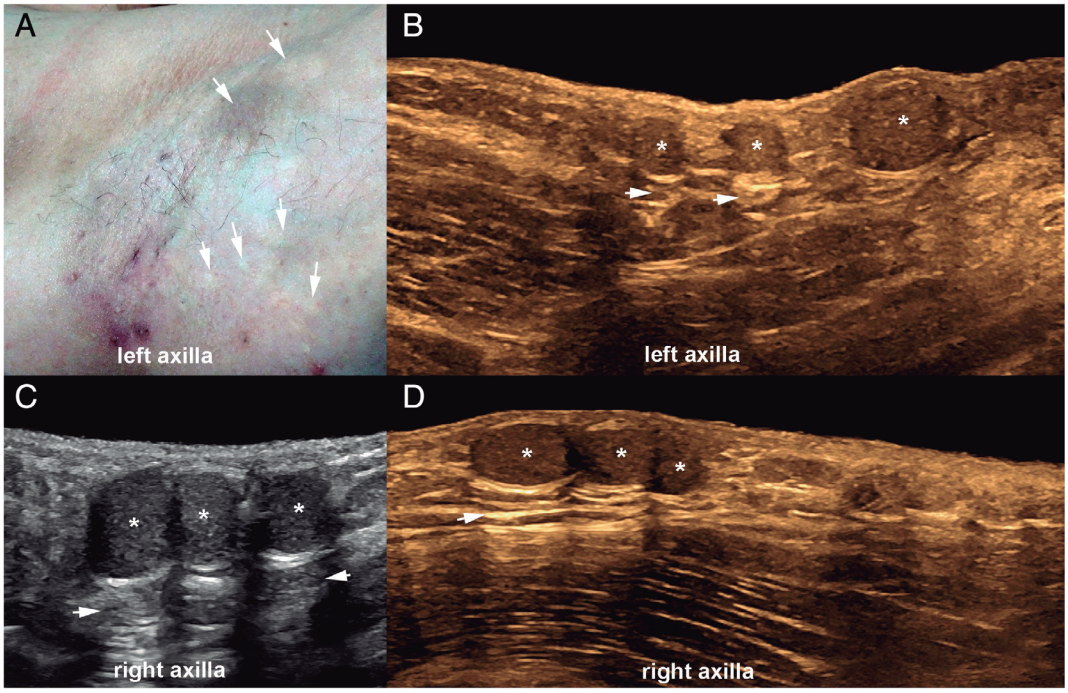
Results

The images of 9 patients with histologically confirmed SCM with a total of 87 lesions were analyzed. The average number of steatocystomas found per patient was 9.7. Sixty-seven percent of the patients were female, and 33% were male (mean age, 33 years; range, 16–49 years; SD, 10.2 years). The most frequent location was the axillary region, which was present in 44.4% of the sample, whereas the cervical, perineal-genital, intermammary, intergluteal, and anterior chest regions were 11.1% each.

On US imaging, SCM appeared as well-defined hypoechoic nodules with mild posterior acoustic enhancement in 100% of cases (Figure 1). In 67%, they were located in the dermis and subcutis; in 11.1%, they were only dermal; and in 22.2%, they were only subcutaneous. A more detailed description of the cases is shown in Table 1.

The average volume of the lesions was 70 mm^3 (range, 12–198 mm^3; SD, 59.9 mm^3), and 100% of cases showed clusters of 2 or more adjacent nodules in the same body region (Figures 2 and 3). No calcification foci were detected inside or at the periphery of the lesions.

Figure 1. Steatocystoma multiplex at the axillary region (female, 16 years old). **A**, Clinical image (left axillary region) shows several yellow and brown nodules (vertical arrows). **B–D**, Left axillary region (**B**) and right axillary region (**C** and **D**) show clusters with well-defined subcutaneous hypoechoic nodules (asterisks) that slightly protrude into the dermis in **B**. Notice the posterior acoustic reinforcement (horizontal arrows) suggestive of cystic lesions.



Regarding vascularization, signs of hypervascularization were observed in the periphery of steatocystomas in 55.6% of the patients (Figure 4). The mean thickness of the peripheral vessels was 1.1 mm (range, 0.7–1.5 mm; SD, 0.25 mm), and the mean peak systolic velocity of these vessels was

Table 1. Clinical and Ultrasound Characteristics of SCM Cases

Case	Age, y	Sex	Body Location	Lesions	Echogenicity	Extension	Volume, mm^3	Vascularization	Associated Condition
1	45	Female	Perineal-genital	5	Hypoechoic	Dermal and subcutaneous	54	Increase, periphery	HS
2	49	Female	Intermammary	15	Hypoechoic	Dermal and subcutaneous	14	No increase	None
3	27	Male	Posterior neck	12	Hypoechoic	Dermal and subcutaneous	12	Increase, interior and periphery	None
4	16	Female	Axillary	12	Hypoechoic	Subcutaneous	28	Increase, interior	None
5	32	Male	Intergluteal	10	Hypoechoic	Dermal	24	No increase	Vellus hair cyst
6	37	Female	Axillary	7	Hypoechoic	Dermal and subcutaneous	72	No increase	None
7	39	Female	Axillary	6	Hypoechoic	Dermal and subcutaneous	198	Increase, periphery	None
8	23	Female	Axillary	5	Hypoechoic	Subcutaneous	88	Increase, periphery	HS
9	25	Female	Anterior chest	15	Hypoechoic	Dermal and subcutaneous	140	No increase	HS

Figure 2. Steatocystoma multiplex at the anterior chest (male, 25 years old). **A**, Clinical image shows yellow and erythematous nodules (arrows). **B**, Grayscale US with a color filter shows a cluster of 3 dermal and subcutaneous hypoechoic nodules (asterisks). **C**, Color Doppler US shows slight hypervascularity (in colors) in the periphery of one of the nodules.

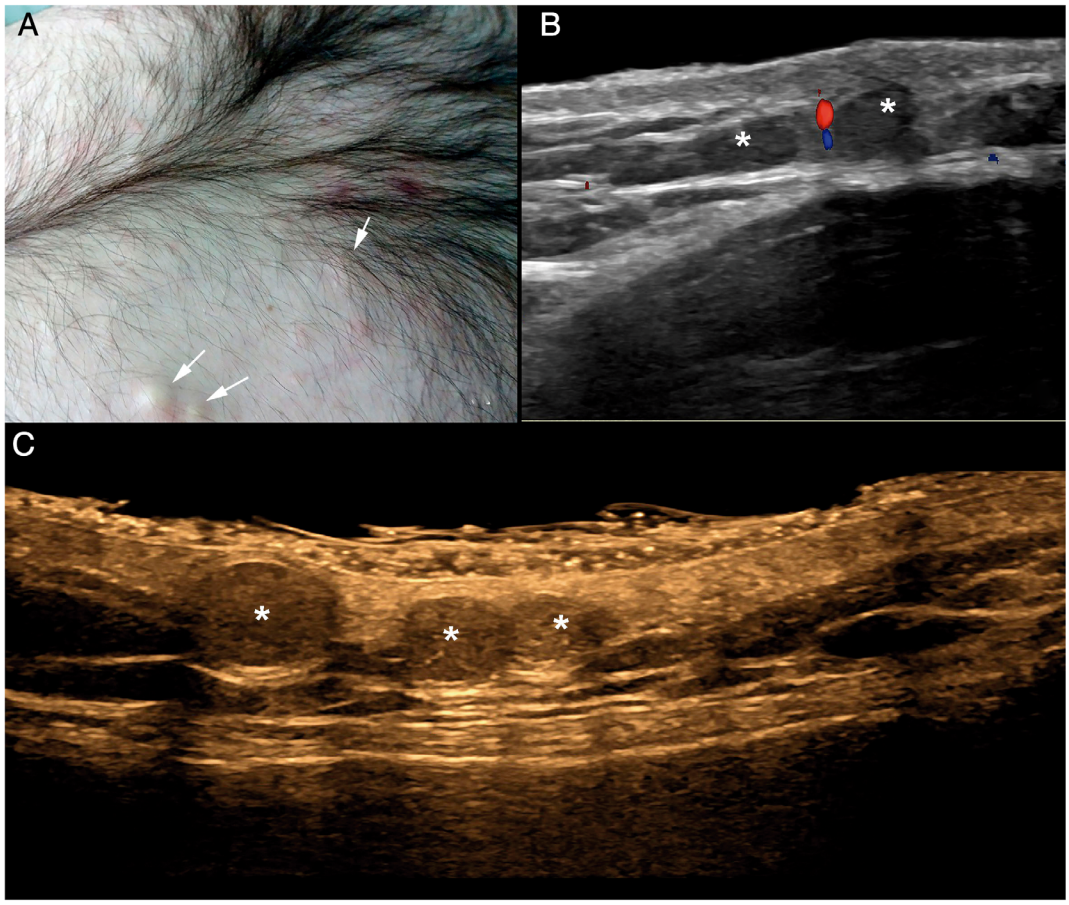
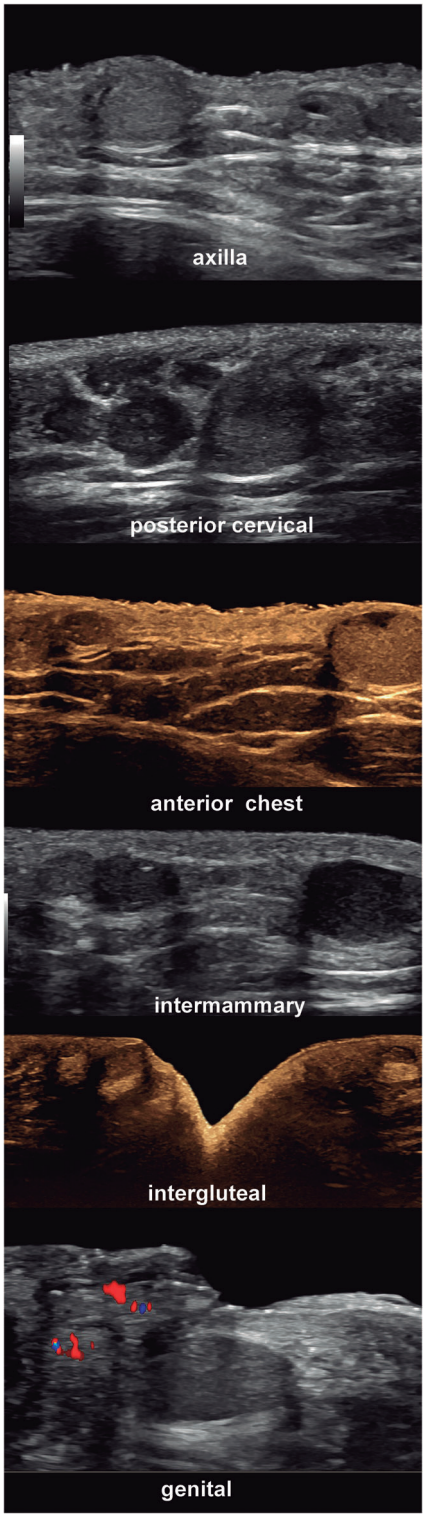
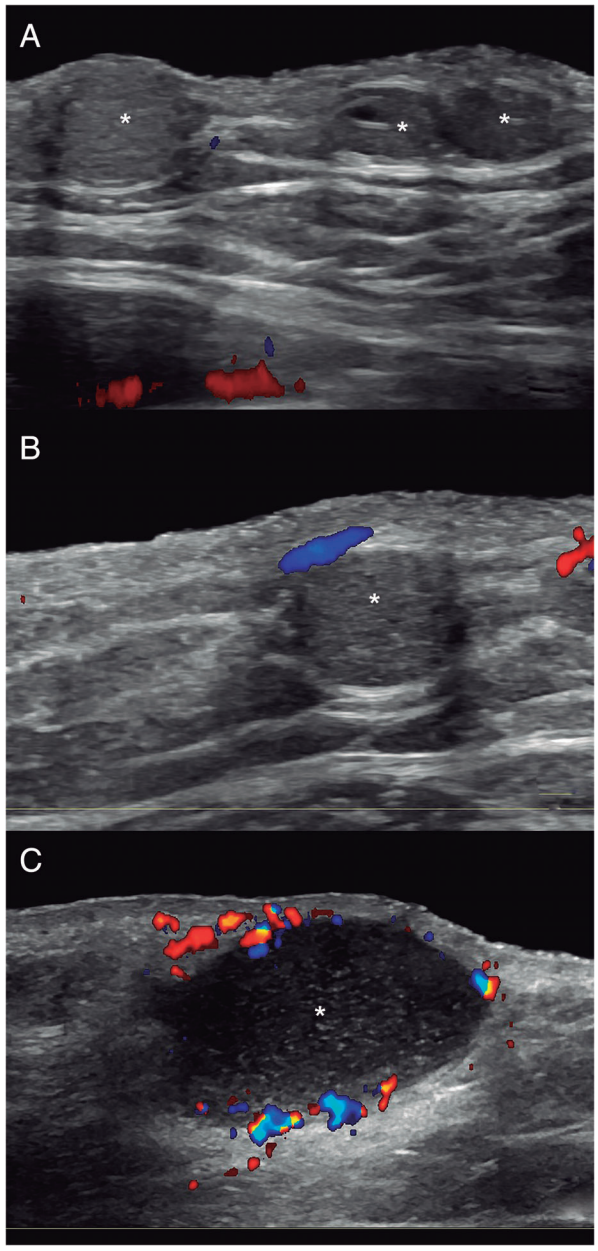


Figure 3. Steatocystoma multiplex at different corporal locations.



8.6 cm/s (range, 3.1–21.7 cm/s; SD, 6.7 cm/s). In total, 44.4% of cases were associated with another dermatologic condition investigated by US; the most frequent was hidradenitis suppurativa (HS; 75%), followed by vellus hair cysts (25%).

Figure 4. Steatocystoma multiplex (asterisks) with variable degrees of peripheral vascularity going from low (**A**) to high (**C**).



Discussion

To our knowledge, this is the largest series of SCM to date, and the anatomic information provided by US was essential to confirm this condition and rule out other dermatologic entities. In SCM, US can be relevant to define management strategies and eventually to decide whether a US examination is also needed for other family members. Unlike some histologic descriptions that report an only dermal location of SCM, our series demonstrates that steatocystomas frequently compromise subcutaneous tissue, which can be relevant to plan the surgery properly, since the incision should be deeper.[9,10] The presence of hypervascularization in the periphery of SCM is associated with inflammation, which could be idiopathic or secondary to trauma: eg, chronic friction in intertriginous areas, such as the axillary or perineal folds.

Unlike a study by Park et al,[6] we did not find isoechoic nodules, whereas SCM lesions were examined in 5 women in that study, and 17% of them were isoechoic lesions. Our case series coincides with the dermal and subcutaneous locations reported by Wan et al,[7] who described the US characteristics of SCM in a 59-year-old woman presenting with multiple dermal and subcutaneous small nodules in the chest wall, armpits, and forearm. One of the key US features of the SCM is its cluster type of presentation, which differs from the US appearance of other types of cutaneous cysts, such as epidermal or trichilemmal cysts.

The main differential diagnoses of SCM should be established with other cystic and adnexal tumors, which can present a similar clinical appearance. As an example, epidermal cysts, unlike SCM, usually present as a single well-circumscribed round or oval dermal and subcutaneous structure that can show a communicating tract to the epidermal plane called a punctum.[11] The cavity of epidermal cysts contains keratin, commonly without fragments of sebaceous glands or muscular fibers. Additionally, the presence of compact hyperechoic laminae of keratin, already reported in epidermal cysts, was not observed in our SCM cases.[12]

Trichilemmal cysts commonly show a well-defined oval structure and are located in the dermis and subcutaneous tissue.[13] In a study of 50 patients with trichilemmal cysts, 72% were hypoechoic, and 26% were mixed hypoechoic and anechoic. In contrast to SCM, calcifications are common in trichilemmal cysts, being reported in 65% of cases, and their most common location is the scalp.[14]

Pilomatrixoma is another differential diagnosis that can generate a clinical challenge. This is a benign adnexal tumor derived from the matrix of the hair follicle. This benign tumor appears on US imaging as a well-defined dermal and subcutaneous nodule.[15] Its most common form of presentation is the target type, with a hypoechoic rim and a hyperechoic center that contains a variable amount of hyperechoic spots that correspond to calcium deposits, which may show different intensities of a posterior acoustic shadowing artifact.[16] In SCM, no internal calcium deposits were observed in our cases or reported in the literature. On color Doppler imaging, pilomatrixomas can show a variable degree of vascularity both in the periphery and internally.[15] In our SCM series, the hypervascularity only showed a perilesional pattern.[17]

Interestingly, HS was identified as an associated lesion in 33% of cases (3 of 9). In these patients, the US alterations already described in HS were observed. These included a widening of the regional hair follicles, decreased dermal echogenicity, dermal and subcutaneous pseudocysts, fluid collections, and fistulous tracts in the intertriginous regions.[18–21] The association of SCM and HS has been previously described in the literature.[22–24] Hollmig and Menter[22] reported a case of SCM and HS in the context of a family association.

Moreover, an inflammatory variant of SCM has been described, which is called suppurative SCM. This type of SCM presents as lesions that evolve with rupture, inflammation, and the consequent healing of the areas involved, showing clinical lesions similar to HS.[25,26] Furthermore, Atzori et al[24] reported 3 cases of SCM associated with HS, confirmed with a biopsy, supporting the coexistence of the two diseases.

One of the limitations of our work was the small number of cases. However, this is the highest number of cases reported in the literature, which could be explained by the fact that, in comparison to other dermatologic conditions, SCM is rare. Further research may be needed to compare these US morphologic features with other larger samples of SCM.

In conclusion, SCM has US characteristics that allow discrimination from other common cystic and solid dermatologic entities. These US features can support an earlier and more accurate diagnosis as well as better surgical planning and management.

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