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Mammographic and sonographic findings of steatocystoma multiplex presenting as breast lumps

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ABSTRACT Steatocystoma multiplex (SM) is an uncommon cutaneous disorder characterised by multiple intradermal cysts distributed over the trunk and proximal extremities. This condition affects both genders and is often inherited as an autosomal dominant trait, although sporadic cases have been described. This report describes the mammographic and sonographic features of the cysts, which presented as breast lumps, for evaluation. The cysts appeared as numerous well-circumscribed, radiolucent nodules with thin radiodense rims on mammography. On sonography, the cysts could be hypoechoic, isoechoic or demonstrate mixed echoes containing debris-fluid levels, depending on the amount of clear oily liquid and keratinous material. SM can be diagnosed based on a clinical setting of multiple asymptomatic small intradermal nodules over the trunk and proximal extremities, positive family history and imaging findings.

Keywords: cysts, mammography, sonography, steatocystoma multiplex
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INTRODUCTION

Steatocystoma multiplex (SM) is an uncommon cutaneous disorder secondary to hamartomatous malformation of the pilosebaceous duct junction.[1] The condition manifests as numerous small intradermal cysts distributed primarily over the trunk, axillae, neck, groin and proximal extremities.[2] It tends to start in adolescence or early adulthood and affects both genders, although some authors reported a higher rate in males.[3,4] SM is often inherited as an autosomal dominant trait,[3,5] but sporadic cases have been reported.[5,6] This case report presents the mammographic and sonographic features of the cysts, which presented as palpable breast lumps, for investigation.

CASE REPORT

A 59-year-old Chinese woman was recalled to the breast assessment clinic for further assessment following a routine BreastScreen Singapore (BSS) screening mammogram. Although her mammogram was read as normal/benign, she was recalled due to her complaint of palpable breast lumps, in accordance with BSS protocol. At the breast assessment clinic, she underwent further imaging with breast ultrasonography and clinical review by a breast surgeon. She was found to have superficial palpable lumps with benign sonographic and clinical features. The patient had brought along an old report from her rheumatologist, which stated her history of systemic lupus erythematosus (SLE), Raynaud's phenomenon, Sjögren's syndrome, hypertension and SM.

The patient had had the lumps for many years with no associated pain, overlying skin changes or discharge. Physical examination revealed several small, superficial, mobile, smooth and soft nodules over the anterior chest wall, breasts, axillae and forearms. She has no known family history of a similar cutaneous condition. On mammography, the lumps appeared as superficial, well-defined, round or ovoid radiolucent nodules with thin, smooth and slightly radiodense rims with or without posterior acoustic enhancement. There was no associated architectural distortion, spiculation or microcalcification (Fig. 1). On sonography, the nodules were well-circumscribed, exhibiting either iso- and/or hypoechogenicity. No colour Doppler signal is detected within the nodules, which is in keeping with their avascular cystic nature (Fig. 2). The findings from the imaging studies and physical examination were in

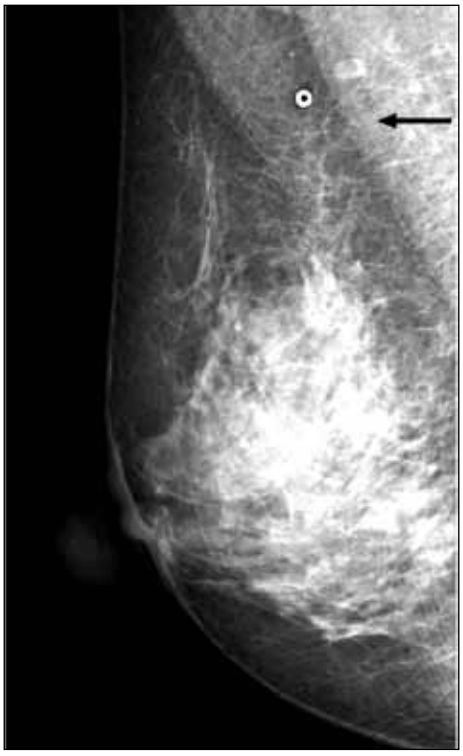

Fig. 1 Mammograph shows right breast mediolateral oblique view with a surface bead marker over the clinically palpable lump, which corresponds to a well-defined radiolucent nodule with a thin rim (arrow).

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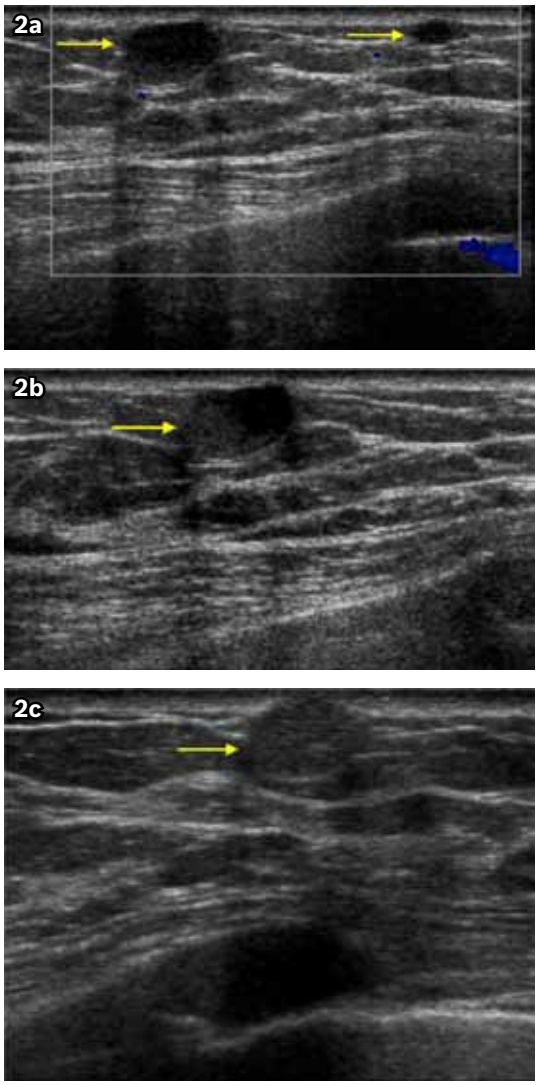

Fig. 2 Colour Doppler images show (a) hypoechoic; (b) mixed-echoic; and (c) isoechoic cystic nodules with well-circumscribed margins in the dermal plane (arrows), displaying the avascular nature of the nodules.

concordance with the patient's documented diagnosis of SM, and thus referral to a dermatologist or further investigation was not required.

DISCUSSION

SM is a rare autosomal dominant disorder with sporadic cases described in the literature. Covello et al reported keratin 17 mutations to be an important factor in the formation of familial SM, but they could not find any correlation between genotype and phenotype, nor could they detect any keratin 17 mutations in sporadic cases, thus suggesting a multifactorial basis (both genetic and environmental) for the condition.[7] SM is usually distributed over the anterior chest, epigastrium, axillae, groin, neck and proximal extremities, and less frequently, on the back, abdomen and external genitalia, and rarely confined to the face/scalp.[6,8] Acral SM, in which involvement of the extremities is more prominent than the trunk, is an uncommon form of SM described in previous case reports.[9,10] There is no known association between SM and autoimmune diseases such as SLE, Sjögren's syndrome and Raynaud's phenomenon.

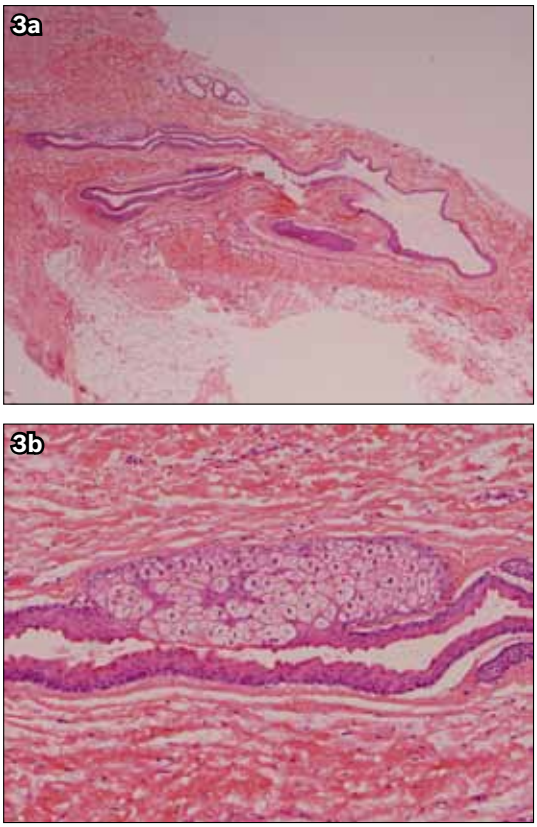

Fig. 3 Photomicrographs show the typical histological appearance of a steatocystoma multiplex nodule (of another patient), with the undulating cyst wall composed of stratified squamous epithelium and sebaceous gland within the cyst wall (Haematoxylin & eosin, × 20 and × 100, respectively).

The histopathological findings of SM consist of cysts in the mid/deep dermis, which sometimes extend to the adjacent subcutaneous plane. They are composed of three to five layers of squamous epithelium,[5] and are lined by flattened sebaceous glands and acellular eosinophilic cuticles.[6] Cysts can contain clear oily liquid, small amounts of keratinous material, and rarely, lanugo hair.[4] The radiolucent appearance at mammography is due to the presence of its oily content. On sonography, the presence of a clear oily liquid usually results in a hypoechoic appearance, and less commonly, isoechoic appearance. If both the oily liquid and echogenic keratinous material are present, a debris-fluid level can be seen. The sizes of the cysts are all less than 1.5 cm, which agrees with other reports.[11] Most cysts remain asymptomatic for many years, although there is a possibility of them becoming inflamed, rupturing and draining. Limited success has been reported with the use of both oral retinoids and liquid nitrogen cryotherapy for treating suppurative lesions.[12] Fig. 3 shows the histological features and Fig. 4 shows the clinical appearance of SM nodules.

The differential diagnosis of a radiolucent nodule on mammography includes galactocele, lipoma, fat necrosis, epidermal cyst, severe nodulocystic acne, lipomatosis and xanthomatosis. An infected SM cyst can present as a dense lesion mimicking a fibroadenoma or well-circumscribed carcinoma on mammography.[5,13,14] A well-circumscribed hypoechoic or isoechoic nodule on sonography is nonspecific by itself. Sonographically, a nodule with a fluid-fluid or fluid-debris level

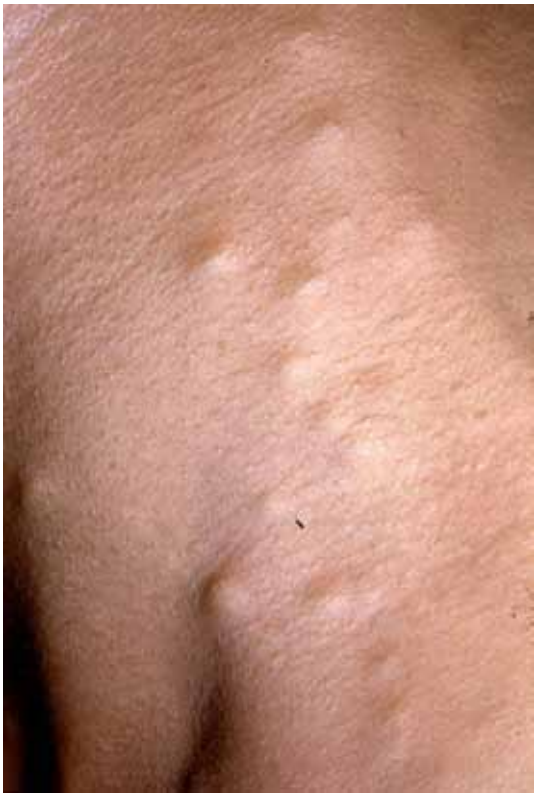

Fig. 4 Photograph shows the typical clinical appearance of steatocystoma multiplex superficial nodules (of another patient) distributed over the forearm.

can represent a galactocele, haematoma, abscess or intracystic papillary carcinoma.[15]

The presence of multiple superficial radiolucent nodules on mammography that correspond to well-circumscribed nodules, which can be hypoechoic, isoechoic or with fluid-debris level in the mid/deep dermis on sonography (with distribution over the anterior chest wall, axillae and proximal extremities), as well as an asymptomatic history since early adulthood, allows a physician to make a clinical diagnosis of SM, thus avoiding an unnecessary invasive workup.

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