Sir,

We describe here a case of steatocystoma multiplex presenting as acral subcutaneous nodules. This case is unique due to: (i) acral distribution and (ii) presentation as subcutaneous nodules. As far as we know, there have been only 2 reports of steatocystoma multiplex (SM) occurring in a predominantly acral distribution.

CASE REPORT

A 47-year-old Japanese housewife was referred to our hospital with asymptomatic subcutaneous nodules on her forearms. She first noticed the subcutaneous nodules 10 years previously, and they had gradually increased in both size and number. They were not associated with any pain or tenderness. Her family history and past clinical history were unremarkable. Physical examination revealed multiple, mobile, well-defined, elastic-hard, 5–10 mm subcutaneous nodules on the flexor surface of the forearms; 8 on the right and 3 on the left (Fig. 1). There were no other lesions except for the forearms and no nail changes. Clinically, the lesions were initially thought to be multiple lipomas and an excisional biopsy was performed to confirm the suspected diagnosis. Upon biopsy, a well-circumscribed ovoid cyst was isolated in the superficial subcutaneous fat layer. The cyst contained yellow creamy material. Biopsy specimen showed a well-encapsulated subcutaneous cyst with infolded walls lined by squamous epithelium without a granular layer. Sebaceous gland lobules are found within the cyst wall (Fig. 2). From these findings, the diagnosis of SM was finally made.

DISCUSSION

This case of SM is unique due to (i) its acral distribution and (ii) presentation as subcutaneous nodules. SM can appear anywhere on the body but is more common in areas where the pilosebaceous apparatus is well developed, such as the trunk, neck, axilla, inguinal region, scalp and the proximal extremities. Acral SM, which involves the extremities more prominent than the trunk, is rare and has been described in only 2 reports (1, 2). Chu (1) reported a 25-year-old man with a 20-year history of asymptomatic nodules on the arms and chest, which showed findings consistent with SM upon histopathological analysis. The patient had no family history and no nail changes like in pachonychia congenita (PC). Rollins et al. (2) reported a 32-year-old Filipino woman with an 8-year history of multiple cystic nodules on the distal upper and lower extremities. The patient’s family history was insignificant, and she had no changes in the nails. With regard to the depth of the lesion, steatocystoma is thought to result from a hamartomatous malformation of the pilosebaceous duct junction and is usually located in the mid-dermis (3). In our case, the lesions were palpated as subcutaneous nodules mimicking multiple lipomas, which is not a well-described presentation in textbook SM references. Dermatologists should be aware of that SM may present as acral subcutaneous nodules.

Covello et al. (4) reported that keratin 17 mutations commonly underlie both PC type-2 and SM, however,
they could not find a correlation between genotype and phenotype. Furthermore, they could not detect any keratin 17 mutations in sporadic cases of SM (4). These observations suggest a multifactorial basis, including both genetic and environmental factors, for this disease. The reason why our case exhibited an acral distribution and presentation as subcutaneous nodules is not understood, but a combination of genetic factors including keratin 17 abnormalities, other keratin defects and/or environmental factors, may be involved in the unique clinical appearance.

REFERENCES