**INTRODUCTION**

Lipodystrophia centrifugalis abdominialis infantilis (LCAI), a rare but benign subcutaneous fatty tissue disorder, is first observed and reported by Imamura *et al.* in 1971. Clinical presentation include (1) a large depressed lesion resulting from the loss of subcutaneous fatty tissue and involving mostly abdominal region, (2) centrifugal enlargement of the depressed lesion,
slightly erythematous, swollen border with some scaling in the surrounding area, onset before the age of 3 years, and no significant systemic disorder or other skin abnormalities is associated. We hereby describe a young
Taiwanese girl with this rare condition. She is believed to be the first such case reported in Taiwan.

**CASE REPORT**

A 7-year-old Taiwanese girl was brought to the dermatology clinic at Chang-Gung Memorial Hospital in March 2002 for a closer examination of a depressed asymptomatic skin lesion measuring 10 x 20 cm² on her right inner thigh (Fig. 1A). According to her parents, she was the product of a full-term, uncomplicated pregnancy and delivery, and had achieved all her milestones at the expected age. There was no history of similar skin lesions in the family. Her parents stated that a 2 x 2 cm² erythematous patch existed on the girl's right inguinal area since birth, and was once treated as napkin dermatitis in infancy with no improvement. When the girl was 2-years-old, the depressed skin area began to slowly enlarge centrifugally and extended to her central lower abdomen, right pubis and right labia majus. This lesion had remained stationary since. By examination, there is some wrinkling of the depressed skin and the fine underlying bluish vessels are visible through skin. The lesion is well-defined by its slightly erythematous change. In addition, a 5 x 6 cm² soft, slightly bulging but asymptomatic mass locating in the lower portion of the depressed region was palpated (Fig. 1B). Regional lymph node was not detected.

Laboratory studies of complete blood count, differential ratio and electrophoresis were normal. Magnetic resonance image (MRI) study of the protruding mass proved absence of any solitary tumor. A biopsy specimen was taken from the depressed area of the thigh and examined histologically. Histopathology showed relatively unremarkable dermis (Fig. 2A), but mild inflammatory infiltrate of lymphocytes and histiocytes in the subcutaneous fatty tissue (Fig. 2B).

**DISCUSSION**

Imamura *et al.* first proposed the term lipodystrophia centrifugalis abdominalis infantilis (LCAI) for a rare but benign subcutaneous fatty tissue disorder in 1971. Its main characteristics are a well-defined skin depression with a scaling and slightly erythematous border, onset before age of 3 years, and occurring mostly on lower abdominal and inguinal area. In 1972, Morishima added another characteristic feature of regional lymph node swellings. Histological study of the depressed region often reveals a decrease of the subcutaneous fat along with an inflammatory infiltration in the lower dermis and subcutis, consisting mostly of lymphocytes and histiocytes. Our patient’s histopathology showing only mild inflammatory infiltrate of lymphocytes and histiocytes in the subcutaneous fatty tissue may mostly be explained by the fact that the lesion has remained stable for the past 5 years since age 2. Similar histological findings have been cited in cases reported by Makino and Higuchi. More than 100 cases have been documented since Imamura *et al.* first gave this ailment the name lipodystrophia centrifugalis abdominalis infantilis, which occurred predominantly in Eastern Asians. In addition to the 86 cases diagnosed in Japan, Lee *et al.* reported 8 Korean cases, Giam *et al.* and Tay *et al.* both reported 1 Singaporean-Chinese case, and Zachary *et al.* and Müller *et al.* each reported a Caucasian case. One previous Caucasian case reported by Cairns was dismissed by Imamura due to its insufficient criteria for the diagnosis. The case reported by Cairns displayed keloid-like appearance around the depression with symptoms of itching. Most of all, this particular patient revealed pathological changes that resembled more closely to scleroderma than LCAI. Lee *et al.* has, on the other hand, attempted to revise LCAI to lipodystrophia centrifugalis abdominalis juvenilis due to the fact of finding this dermatosis in young children aged 2 to 9 years old and not confined to infants. Many authors have contemplated on the association of LCAI with Kawasaki disease, underlying immunological and even collagen disorders but since each has been proven to be a single incidence so far, and no formal rela-
tion has been established.

Up till now, there is no effective therapy for LCAI, and application of topical steroids have not prevented the extension of depressed lesions. The progression of the depressed lesion often ceases at the onset of puberty, with subsequent improvement and partial recovery spontaneously.

Differential diagnosis of this entity should include macular atrophy, atrophoderma idiopathica progressiva, panatrophy of Gowers, Rothmann-Makai syndrome, lupus erythematosus profundus, adiponecrosis subcutanea neonatorum, lipodystrophia progressiva as well as morphea, progressive lipodystrophy, and insulin lipoatrophy. Age of onset and pathological features are important factors for differentiating LCAI from the above entities. In cases of atrophoderma idiopathica progressiva and pantrophy of Gowers, the age of onset is mostly beyond the teen years. Rothmann-Makai syndrome may be differentiated by its granulomatous formations by histiocytes and widespread fibrosis in later stages. Lupus erythematosus profundus, which also has lymphocytic infiltration in the subcutaneous fatty tissue, is different from LCAI by characteristics of hydropic degeneration of basal layer, follicular plugging and fibrinoid degeneration of dermal collagen in pathological finding. Likewise, histological exam of adiponecrosis subcutanea neonatorum often reveal necrosis of fatty cells with marked inflammatory foreign body reaction, as well as elongated needle-shaped clefs in rosettes. Impression of Lyme borreliosis also needs to be considered, but history of tick bite and classic erythema migrans with itchy irritation has not been reported in our case. Correct diagnosis for LCAI should not be difficult in patients with an Asian background presenting with the five unique features as described previously. In addition to being the first reported LCAI in Taiwan, our patient poses as a very interesting case due to the finding of a protruding soft mass located in the region of decreased subcutaneous fat. MRI study showed no presence of tumor mass. The bulging skin is believed to have resulted from loose underlying subcutaneous tissue. This patient will be followed for the observation of the progression of her lesion. Knowing that LCAI, though rare, occurs mostly in Eastern Asians, it is essential that dermatologists in Asian countries have a full understanding of the clinical picture and course of this skin disorder. This way, physicians in this specialty may be able to make differential diagnosis and offer proper consultation for the parents of these young patients. In the future, we hope to collect more LCAI cases and thereby determine the incidences and characteristics of this rare disorder in the Taiwanese population.

REFERENCE
10. Müller S, Beissert S, Metze D, et al.: Lipodystrophia centrifugalis abdominalis infantilis in a 4-year-old caucasian girl: association with partial IgA deficiency and


