



**Lipodystrophia centrifugalis abdominalis infantilis
accompanied by idiopathic encephalopathy**

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6 Lipodystrophia centrifugalis abdominalis infantilis accompanied
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24 methylprednisolone pulse therapy
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6 Lipodystrophia centrifugalis abdominalis infantilis (LCAI) is a rare
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8 localized lipoatrophic disorder [1]. It is predominantly reported in
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10 Japanese, although it was recently reported in Caucasian [2, 3]. In LCAI,
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12 the subcutaneous fat of the abdomen and upper groin is usually affected.
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14 The etiology of the disease is unknown. We report a case of LCAI
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16 accompanied by idiopathic encephalopathy.
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21 The present patient was a 9-year-old Japanese boy. He had no past
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23 medical history. According to his mother, a dusky-reddish patch was first
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25 noted on his right abdomen when he was 4 years old, which spread
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27 centrifugally. Inguinal lymph node swelling appeared at the age of 5 years,
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29 with increasing depression of the area on his right abdomen. The patient
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31 was first treated with a topical steroid cream, but the depressed area
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33 enlarged.
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38 The patient experienced acute encephalopathy at the age of 9 years. He
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40 presented at another hospital because of status epilepticus that followed
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42 high fever and disturbance of consciousness. Magnetic resonance imaging
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44 of the brain was performed, and T2-weighted and fluid-attenuated inversion
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46 recovery imaging showed abnormal high-intensity areas predominantly in
47
48 the left cerebral hemisphere. Electroencephalography showed generalized
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50 slow waves predominantly in areas of the left hemisphere. Results of
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52 polymerase chain reaction for infectious pathogens were negative for the
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54 DNA of herpes simplex virus (HSV), human herpes virus (HHV)-6, and
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6 HHV-7 in the cerebrospinal fluid (CSF). CSF analysis revealed the
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8 following: protein, 14 mg/dL (within the normal range); glucose, 100
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10 mg/dL; and cell density, 8 cells/mm³ (mainly lymphocytes). Blood culture
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12 was sterile. Paired serum serology by immunoglobulin M antibody capture
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14 enzyme-linked immunosorbent assay was negative for HSV, measles virus
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16 (Morbillivirus), and rubella virus. The cause of the encephalopathy was not
17
18 identified. The patient received two courses of methylprednisolone pulse
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20 therapy and oral phenobarbital just after admission to the hospital. For
21
22 supportive therapy, oral prednisolone was administered at an initial dose of
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24 1 mg/kg per day for 2 weeks. After 1 month, T2-weighted imaging showed
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26 no high-intensity areas. After the immunosuppressive intervention, the
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28 abdominal depressed area stopped spreading. The patient reported no
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30 seizures at the 6-month follow up at our outpatient clinic. On physical
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32 examination, the patient presented with a depressed area with a
33
34 translucent whitish skin surface on his right abdomen (Fig. 1A). No
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36 other significant skin changes were detected. There was no family
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38 history of a similar disorder. Histological examination of a biopsy
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40 specimen taken from the margin of the depressed skin area after the
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42 spreading had stopped showed little inflammation (Fig. 1B).
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49 The pathogenesis of LCAI remains unknown. It is hypothesized that
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51 immunological dysfunctions are associated with the development of
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53 LCAI, because inflammatory cell infiltration, including lymphocytes
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55 and histiocytes, is often seen in the degenerating fatty tissue. Although
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6 there were two cases of co-existing LCAI and epilepsy in the Japanese
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8 literature, there are no previous reports of idiopathic encephalopathy
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10 preceded by LCAI [4].
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14 The treatment of LCAI is not established. A variety of treatments have
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16 been used, including systemic therapies (such as corticosteroids,
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18 chloroquine, penicillin, vitamin E, and ibuprofen) and topical therapies
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20 (such as corticosteroids) [5]. None of these treatments prevents the
21
22 enlargement of depressed lesions, although oral and topical steroids
23
24 occasionally reduce inflammation in the surrounding area. In the present
25
26 case, enlargement of the depressed area and spread of inflammation ceased
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28 after immunosuppressive therapy, although we cannot exclude the
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30 possibility that disease progression stopped spontaneously after the
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32 treatment.
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38 In conclusion, to our knowledge, this is the first report of LCAI
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40 accompanied by idiopathic encephalopathy. In our patient, LCAI was
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42 successfully treated by methylprednisolone pulse therapy. The present case
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44 strongly suggests that certain immunological dysfunctions are associated
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46 with LCAI.
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Figure Legend

Figure 1. Clinical features and pathological findings of the patient

(A) A well-defined depressed lesion in the lower abdomen (shown by arrows). (B) In the skin biopsy taken from the lesion margin after it had stopped spreading, almost no inflammation is observed in the adipose tissue. Bar: 1mm

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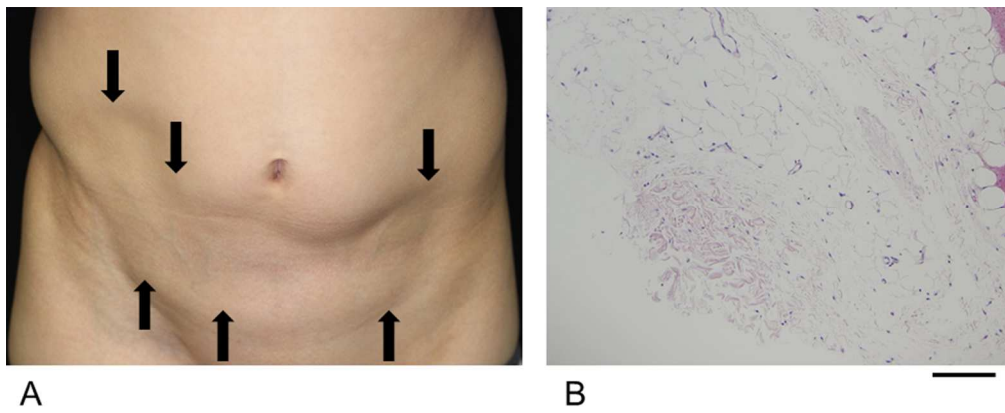


Figure 1. Clinical features and pathological findings of the patient
(A) A well-defined depressed lesion in the lower abdomen (shown by arrows). (B) In the skin biopsy taken from the lesion margin after it had stopped spreading, almost no inflammation is observed in the adipose tissue. Bar: 1mm

180x71mm (300 x 300 DPI)

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