A 74-year-old female patient consulted our hospital for muscle weakness of the upper and lower limbs. The patient had a 10-year history of hyperCKemia ranging from 316 to 1164 U/L (upper limit of normal: 200 U/L) and a 5-year history of dysphagia and difficulty in climbing up stairs. In addition, there was a 2-month history of difficulty in washing her hair owing to finger weakness prior to consultation. There was no family history of muscle diseases. Neurological examination demonstrated muscle atrophy of the bilateral upper and lower limbs, which showed muscle weakness corresponding to approximately grade 4 of the Medical Research Council scale on manual muscle testing. A needle electromyogram revealed myopathic changes. Muscle biopsy demonstrated endomysial inflammation, invasion of nonnecrotic fibers, and rimmed vacuoles, establishing a diagnosis of inclusion body myositis (IBM). The patient’s symptoms gradually progressed. On examination at age 75, the grip strength of the right and left hands was 1.5 and 3 kg, respectively (the normative grip strength of females aged 70 years or more is 15 kg or more). When the patient was asked to make a fist with the right hand at full strength, she was unable to flex the distal interphalangeal (DIP) joints except for the second digit (Fig. 1A). She could fully flex the proximal interphalangeal (PIP) and metacarpophalangeal (MP) joints of the second to fifth digits. T1-weighted MRI of the right forearm showed a decrease of the forearm muscle volume and hyperintensity suggestive of fatty replacement in the flexor digitorum profundus (FDP) muscle (Fig. 1B). The hyperintensity was not apparent in other muscles including the flexor digitorum superficialis (FDS) (Fig. 1B).

Preferential involvement of the FDP muscle has been reported in patients with IBM [1–3], and results in finger flexion impairment pronounced at the DIP joints [1]. So far,
however, the inability to disguise the nails has not been given a lot of attention in other papers. The characteristic fist of our patient is considered indicative of IBM, and could support a diagnosis of IBM, although this sign may emerge in the middle or late stage of the disease.

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References