He was managed with standard treatment of DKA and improved.

But interestingly, he had localized swelling of the thumb and the index finger from birth [Figure 1a and b]. He had undergone corrective surgeries at the age of 3, 8, and 13 years. Despite these surgeries, he again had progressive enlargement of the involved fingers. There was no family history of similar illness, and he did not have any neurocutaneous markers. Radiograph of hand showed soft tissue overgrowth on the radial aspect of right hand along with degenerative bony changes [Figure 2]. Arterial Doppler of the involved hand was normal. MRI hand was done and findings were consistent with macrodystrophia lipomatosa. There was excessive soft tissue, predominantly fat, bone, and nerve on the radial aspect of the right hand [Figure 3a and b]. It was also associated with degenerative changes at the first metacarpophalangeal joint. Hence, final diagnosis of "Macrodystrophia lipomatosa" was kept. No association between diabetes and this condition could be found.

Macrodystrophia lipomatosa is a congenital, non-hereditary condition characterized by localized tissue overgrowth involving the upper and lower extremities.[1-3] It usually involves only one limb and peculiarly occurs over the distribution of plantar nerve in lower limb and median nerve in upper limb. Lower limb involvement is more common.

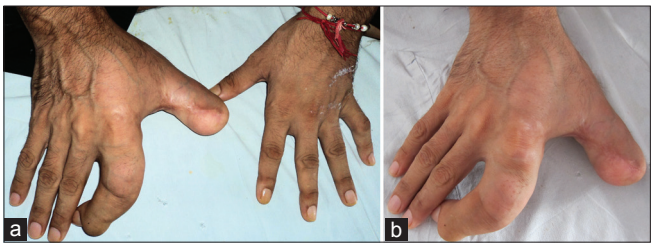

Figure 1: (a) Localized enlargement of radial aspect of right hand. (b) Close up of the localised enlargement

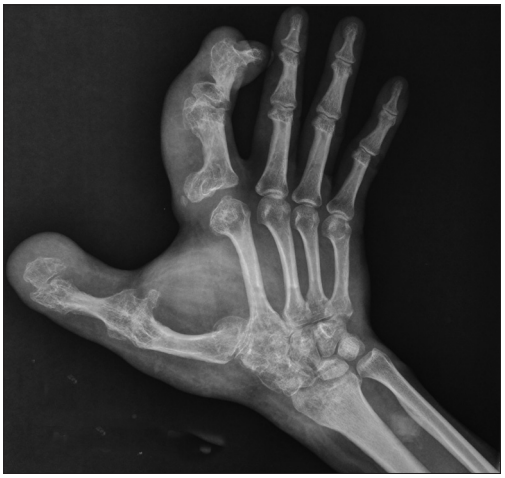

Figure 2: X-ray right hand

Localized acromegaly

Sir,

This 26-year-old male presented with diabetic ketoacidosis (DKA) in our emergency department. He had history of diabetes at the age of 24 years and had been on irregular treatment with insulin and oral antihyperglycemic drugs.

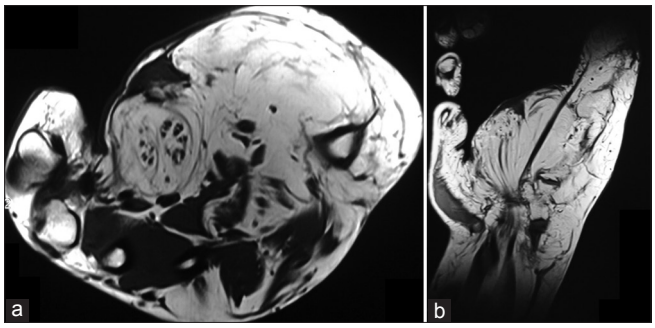

Figure 3: (a) MRI hand (cross-sectional view). (b) MRI hand (coronal)

This rare disorder was first described by Feriz in 1925 in patient with lower limb involvement.[4] Exact etiology is not known but various hypotheses have been put forward. These include altered somatic growth during limb bud development and disturbed fetal circulation.

Though previously the diagnosis was established by histopathology, now the diagnosis can be confirmed by MRI. MRI shows unencapsulated excess fibrofatty tissue as it has high signal on T1-weighted images, intermediate on T2W, and low signal on fat suppressed sequences.[4-6]

Differential diagnoses of localized acromegaly are arteriovenous malformation, neurofibromatosis type 1, hemangiomatosis, lymphangiomatosis, and Proteus syndrome. All these conditions can be differentiated from macrodystrophia lipomatosa on the basis of MRI findings except for Proteus syndrome. Macrodystrophia lipomatosa is sometimes considered localized form of Proteus syndrome which includes multiple malformations and is associated with AKT3 activating mutations.

Gaurav Palikhe, Sridhar Subbiah, Rama Walia, Anil Bhansali

Department of Endocrinology, PGIMER, Chandigarh, India

Corresponding author: Dr. Anil Bhansali, Professor, HOD, Department of Endocrinology, PGIMER, Chandigarh-160012, India.
E-mail: anilbhansali_endocrine@rediffmail.com

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Quick Response Code:	Website: www.ijem.in
	DOI: 10.4103/2230-8210.100702