CASE REPORT

Report of Congenital Generalized Lipodystrophy in Children's Medical Center, Tehran, Iran

Fatemeh Farahmand[1], Fatemeh Yourdkhani[2], Gholam Hosein Fallahi[1], Gholam Reza Khatami[3]

[1] Associate Professor, Pediatric Unit of Digestive Disease Research Center, Children's Medical Center Hospital, Tehran University of Medical Sciences, Tehran, Iran
[2] Research Fellow, Pediatric Unit of Digestive Disease Research Center, Children's Medical Center Hospital, Tehran University of Medical Sciences, Tehran, Iran
[3] Professor, Pediatric Unit of Digestive Disease Research Center, Children's Medical Center Hospital, Tehran University of Medical Sciences, Tehran, Iran

ABSTRACT

Generalized lipodystrophy is a rare disorder in children characterized by absence of subcutaneous adipose tissue. We evaluated 10 patients with generalized lipodystrophy by skin and liver biopsy. Eight patients with steatohepatitis and cirrhosis were treated with Ursobile (uorsodeoxycholic acid) and vitamin E. In follow-up, serum triglyceride level was decreased.

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BACKGROUND

The lipodystrophy syndromes are rare diseases of childhood, characterized by the partial or generalized loss of adipose tissue. Partial lipodystrophy occurs more commonly in females than males and generally begins during the first decade of life.

Generalized lipodystrophy may be congenital or acquired.(1), Congenital generalized lipodystrophy was first described by Berardinelli in Brazil in 1954. It was then reviewed by Seip in 1959, and became known as Berardinelli-Seip syndrome.(2), It is a rare disorder with a prevalence of less than one per 12 million individuals.(3), The disease has an autosomal recessive inheritance and affects all ethnic groups. It is clinically characterized by loss of adipose tissue, especially subcutaneous fat, increased muscular growth, long extremities, cardiomyopathy and hepatosplenomegaly. Additional manifestations include advanced bone age, evidence of liver dysfunctions, dyslipidemia, glucose intolerance or diabetes mellitus, hyper insulinemia and cardiomegaly.(4)

The acquired types may be attributed to autoimmune disorders caused by infections.(5)

So far, two patients with lipodystrophy from Iran have been reported; one case form Mashhad (6) in year 2004, and another from Tehran.(7), Herein, we report on our experience with patients diagnosed with congenital lipodystrophy between April 2000 and July 2005 at the Children's Hospital Medical Center, Tehran, Iran.

MATERIALS AND METHODS

This is a retrospective study performed at the Children's Hospital Medical Center from April 2000 to July 2005.

Ten children aged between two months and five

Corresponding author: Children's Hospital Medical Center. Dr Gharib Ave., Keshavarz Blvd., Tehran, Iran.
Telefax: +98 21 66924545
E-mail: farahmand_gast@yahoo.co.uk