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CASE REPORT

A case of Lawrence Syndrome "acquired generalized lipodystrohy" with type 1 diabetes mellitus in 7 years boy of Bahawalpur

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SUMMARY

Lipodystrophy syndromes are rare disorders characterized by either generalized or partial lack of adipose tissue. They are congenital or acquired. These syndromes are associated with various metabolic and harmonal disorders leading to severe comorbidities including hypertriglyceridemia, diabetes mellitus, acanthosis nigricans, xanthomas, polycystic ovarian syndrome (POS) and non-alcoholic fatty liver disease.
Keywords: Lawrence syndrome, lipodystrophy, diabetes mellitus

INTRODUCTION

Lipodystrophy syndromes consist of rare disorders characterized by either partial or generalized lack of adipose tissue, called partial or generalized lipodystrophy without nutritional deprivation[1]. Lipodystrophy is either congenital or acquired[2]. These syndromes are associated with various metabolic and harmonal disorders which lead to severe comorbidities including hypertriglyceridemia, diabetes mellitus, acanthosis nigricans, xanthomas, polycystic ovarian syndrome (POS) and nonalcohlic fatty liver disease[1,3]. The comorbidities are related to extent of fat loss, gender, age etc . Main causes of mortality in these cases are cardiomyopathy, cardiac failure, renal and liver failure, myocardial infarction, sepsis etc[4,5,6]. The syndromes are associated with autoimmune disorders like hemolytic anemia, thyroiditis, common viable immune-deficiency etc[7].

Majority of complications of these syndromes occur owing to deficiency of adipose (fat) tissue which causes ectopic fat/lipid deposition in muscles, liver or other organs of body leading to insulin resistance which causes hypertriglyceridemia, diabetes mellitus, hepatic fatty disease etc[6]. These cases are resistance to standard treatment for glycemic control.

CASE REPORT

We present a case of 7 years old boy, resident of Bahawalpur, who presented with hyperphagia, uncontrolled blood sugar level and generalized loss of body fat. He had severe insulin resistance, type 1 diabetes mellitus, raised cholesterol levels, hypertriglyceridemia, deranged liver & renal function tests. On physical examination, he had generalized loss of body fat and hepatosplenomegaly.

His hemoglobin was 12.8mg/dL, total leucocyte count was $7.2X10^3$, platelet count was $214X10^3$, serum bilirubin was 09mg/dL, serum ALT (alkaline transaminase) was 241U/L, serum alkaline phosphatase was raised (669 U/L), serum urea was raised (66mg/dL), serum creatinine was raised (1.6mg/dL), serum albumin was reduced i.e., 2.3G/dL. His cholesterol levels and triglyceride levels were markedly raised i.e., 785mg/dL and 3167mg/dL respectively. His blood sugar random (BSR) was 409mg/dL and glycosylated hemoglobin was also raised >9%. His C peptide and anti-tissue transglutaminase (Anti TTG) were normal.

Table 1: Laboratory findings

S/No	Laboratory Test	Normal Range	Patient result
1	Hemoglobin	12 – 18 mg/dL	12.8 mg/dL
2	Total Leucocyte Count	4 – 11 X 10^3/ mm^3	7.2 X 10^3/ mm^3
3	Platelets Count	150 400 X 10^3 / mm^3	214 X 10^3/ mm^3
4	Serum Bilirubin	1 – 17.1 mg/dL	09 mg/dL
5	Serum Alanine Transaminase	<40 U/L	241 U/L
6	Serum Alkaline phosphatase	<279 G/dL	669 G/dL
7	Serum Urea	10-50 mg/dL	66 mg/dL
8	Serum Creatinine	0.6-1.1 mg/dL	1.6 mg/dL
9	Serum Albumin	3.8-4.4 G/dL	2.3 G/dL
10	Serum Sodium (Na)	136 -146 mmol/L	135mmol/L
11	Serum Potassium (K)	3.5 -5.1 mmol/L	4.8mmol/L
12	Serum Cholestrol	<200mg/dL	785 mg/dL
13	Serum triglycerides (TG)	<150mg/dL	3167 mg/dL
14	Serum HDL	>35 mg/dL	24 mg/dL
15	Blood Sugar Random	<140mg/dL	409mg/dL
16	Hb1Ac	Below 6.0 %	> 9%
17	C peptide	0.9- 7.1 ng/ml	0.11 ng/ml
18	Anti Tissue Transglutaminase levels	Positive = >18 U/ml	11.41 U/ml

Figure 1: Child with Acquired Generalized Lipodystrophy

He was admitted and put on regular insulin according to sliding scale and oral metformin, but failed to control his raised blood glucose levels. His echocardiography was also done, which was normal. Ultrasound abdomen revealed enlarged liver and spleen. Patient is on regular follow up. Despite regular follow-up and efforts for glycemic control, his blood sugar and serum triglycerides levels are high.

DISCUSSION

Lipodystrophy syndromes are usually categorized into four major types: Congenital generalized lipodystrophy (CGL), familial partial lipodystrophy (FPLD), acquired generalized lipodystrophy (AGL)

and acquired partial lipodystrophy (APL)[1]. There are other subtypes mentioned in literature including progeroid disease, autoinflammatory disorders, associated with HIV(Human immunodeficiency virus), lipodystrophy caused by injectable drugs etc[6]. We have presented the first case of acquired generalized lipodystrophy in Pakistan and we will limit our discussion to this particulat type of lipodystrophy i.e., Acquired Generalized Lipodystrophy also known as Lawrence Syndrome. It is a rare disorder with less than 100 reported cases[7]. AGL is more frequent in females. Female to male ratio being 3:1. It appears commonly in childhood before adolescence but may appear later in life. There is progressive generalized loss of body fat including soles and palms[1,6]. AGL is frequently associated with various autoimmune diseases.

Our case of Lawrence syndrome was associated with type 1 diabetes mellitus with features of generalized lipodystrophy, raised serum triglycerides levels, hepatosplenomegaly, raised liver, renal enzymes. Similar case was reported by Kumar R et al[8]. Their case of AGL also had associated type 1 Diabetes Mellitus just like ours. Two similar cases were reported by Perk et al, where AGL cases also had type 1 Diabetes Mellitus[9]. Halpern B et al reported a case of AGL who had associated CVI (common viable immunodeficiency) which is an autoimmune disorder[7]. In CVI (common viable immunodeficiency) body doesn't produce antibodies and patient presents with generalized loss of fat and recurrent infections.

CONCLUSION

Lawrence syndrome (Acquired Generalized lipodystrophy) is a rare disorder. Clinicians should be aware of such syndromes for early detection and timely care and management.

Conflict of interest: Nil

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