A Rare Form of Insulin Resistance with Pseudoacromegaly

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Insulin resistance occurs in a variety of common endocrine disorders including obesity, type 2 diabetes, polycystic ovarian syndrome, and metabolic syndrome. Additionally, rare syndromes exist that result in extreme insulin resistance. These conditions help contribute to our knowledge of the mechanisms of insulin signaling and resistance.

Background

Case

12-year-old girl referred to pediatric endocrine clinic for evaluation of new onset diabetes. She was found to have an elevated random glucose in her pediatrician’s office 3 months before. Subsequently she was started on metformin and lisinopril prior to the referral.

• Birth weight: 6lb 7oz (2920 g)
• Obese since kindergarten age
• Extreme weight gain in the last 2 years
• Nearly continuous growth spurt
• Now having difficulty fitting into her clothes
• Premenarchal

Laboratory Assessments

HbA1c 6.6% (49 mmol/mol)
Cholesterol 141 mg/dL (3.64 mmol/L)
Triglycerides 189 mg/dL (2.13 mmol/L)
HDL 29 mg/dL (0.75 mmol/L)
LDL 74 mg/dL (1.91 mmol/L)
AST 31 u/mL (0.52 ukat/L)
ALT 72 u/mL (1.20 ukat/L)

Glucose 186 mg/dL (10.32 mmol/L)
IGF-1 331 ng/mL (43.6 nmol/L)
IGFBP-3 5.3 µg/mL (16038.2 nmol/L)
IGF-1 Receptor

Discussion

• We are reporting on an extremely rare form of severe insulin resistance.
  • Most insulin resistance syndromes are associated with short stature.
  • This syndrome is unique as it is associated with overgrowth.
  • Likely represents downstream defect in insulin signaling:
    • Impaired metabolic effects.
    • Enhanced mitogenic effects.
  • Genetic and functional studies may reveal a causative gene(s).
  • Potential to teach us about novel aspects of insulin signaling.

Author Disclosures

The authors have no relevant financial disclosures or conflicts of interest.

References


Radiology

Bone Age x-ray was consistent with 14 yr. This provides a predicted adult height of 72 inches (1.83 m, +3.23 SDS).

Family History

The patient’s growth chart. Height is noted in blue, well above the 97th percentile. Weight is shown in red, and is extremely elevated. Weight is out of proportion to height.

Diabetes

Stephen Stone

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