Letters to the Editor

Sporadic Growth Hormone Insensitivity Syndrome

Growth retardation is a common clinical problem. In 1966 Laron *et al.* described a syndrome of familial dwarfism in which findings of Growth hormone (GH) deficiency were present despite elevated serum immunoreactive GH level(l).

An eleven year girl, product of consanguineous marriage, born to a Kashmiri couple after an uneventful 39 weeks pregnancy presented with growth retardation. The patient's birth weight was 2.5 kg. One other sibling was normal. The father's height was 170 cm and the mother's 156 cm. There was no family history of growth or developmental disorders. Growth failure became evident in the patient after the age of one year. There was no history of any chronic systemic illness like malabsorption, cardiac, renal, pulmonary or nervous system disorder. She came from an economically well off family and was consuming diet adequate in calories and proteins.

Clinical examination revealed a cherubic face small mandible, depressed bridge of nose and mild truncal obesity. Her height was 117 cm (expected 141.5 cm) and weight was 16 kg (expected 34.0 kg). There were no clinical stigmata of malnutrition, rickets, hypothyroidism or hypocortisol state. Pubertal changes had not yet set in and breast bud had not developed. Systemic examination was normal. Hemogram was normal. Serum biochemistry revealed normal glucose, urea, creatinine, proteins, calcium, phosphorus, sodium, potassium, alkaline phosphatase, and cholesterol. Roentgenograms of her chest and sella turcica were normal. Her bone age was 6-7 years by standards of Greulich and Pyle. Chromosomal analysis revealed 46" genotype. Pelvic ultrasound revealed a 29 mm long infantile uterus. Basal hormone profile revealed T3: 1.8 Hg/ml (normal 0-9-2.5 µg/ml); T4: 7.8 µg/dl (normal 5.5-13.5 µg/dl), TSH: 4.7 mu/ml (normal 1.0-5.0 mu/ml); LH: 8.4 mu/ml (normal 4.0-14.0 mu/ml): FSH 5.0 mu/ml (normal 4.0-12.0 mu/ml). Following dynamic tests were done:

(i Insulin tolerance test was performed with crystalline insulin 0.1 units/kg body weight: Basal GH 63.7 ng/ml; + 30 min: 56.19 ng/ml; + 60 min: 14.45 ng/ml; + 90 min: 65.84 ng/ml; + 120 min: 25.33 ng/ml.

(ii) L-dopa test was (performed with L-Dopa 10 mg/kg body weight orally: Basal GH: 16.19 ng/ml; +30 min: 37.8 ng/ml; + 60 min: 51.8 ng/ml, + 120 min: 21.37 ng/ml.

(iii) Glucose suppression test was (performed with glucose, 1.75 g/kg body weight orally: Basal GH: 14.77 ng/ml; + 30 min 1.78 ng/ml, +60 min: 10.64 ng/ml; + 90 min: 6.42 ng/ml; + 120 min: 8.32 ng/ml.

This patient was clinically suspected to have GH deficiency and her GH

stimulatory investigations revealed persistently elevated immunoreactive GH levels. After glucose load, GH was adequately suppressed. It would have been ideal to measure IGF-1 levels but due to nonavailability of this assay here, it could not be done. A similar case of GH insensitivity syndrome was reported earlier(2). GH insensitivity syndrome encompasses a range of etiologies one of which is GH receptor deficiency (GHRD). The reported patients, now exceeding 150, with GHRD are predominantly of Mediterranean origin(3). A remarkable concentration of GHRD was reported from an inbred Caucasian population in southern Ecuador predominantly affecting females(4). Elucidation of molecular mechanisms responsible for Laron syndrome has been rapid, and promises to be helpful in genetic counseling of the affected families.

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Multifocal Chloromas Preceding Acute Myelogenous Leukemia

We report a rare case of multifocal chloromas preceding acute myelogenous leukemia (AML) by a period of three months, the shortest time interval reported so far in the literature. A ten year old boy presented with low back ache since two months followed by progressive weakness of the lower limbs, hypersthesias and urinary incontinence. On clinical examination, the findings