Isolated Growth Hormone Deficiency-A Case Report

Dr.P.Sekar, Dr.P.Karthikeyan, Dr.M.Sowjan, Dr.Mahesh Inbasekaran

Department of Pediatrics, SRM Medical College Hospital and Research Centre, Kattankulathur, Chennai.

Abstract- Growth hormone is an anabolic hormone released in pulsatile manner in the circulation. It is one of the uncommon causes of short stature in children and is largely idiopathic. The case report is about a 12 Year old boy who presented with short stature. He was diagnosed to have Isolated Growth hormone deficiency. Growth hormone was started after diagnosis was made.

Index Terms- Growth hormone, pituitary

I. INTRODUCTION

Growth hormone has a well-defined role in promoting childhood growth and in maintaining normal adult body composition. Growth Hormone deficiency can be isolated or associated with pituitary disturbances¹. Epidemiological studies suggest that idiopathic isolated growth hormone deficiency occurs more frequently than multiple pituitary hormone deficiency in children whereas in adult onset GH deficiency is frequently due to pituitary adenomas, surgeries and irradiation². Synthetic human growth hormone (somatropin) shows improvement in children with isolated cases of growth hormone deficiency if started early provided that it should be administered under strict supervision.

II. CASE REPORT

A 12 year old male presented to our institute with history of not gaining height. There was no history of perinatal insult, constitutional delay or history suggestive of any systemic causes of short stature. Birth weight and length were adequate and according to race. Dentition was however delayed. On examination the boy was alert with stable vitals and no abnormalities detected in systemic examination. There were no dysmorphic features. The anthropometric measurements showed weight 28 kgs and height 125 cms. His height for age was below 3rd percentile (CDC) and weight for age was also below 3rd percentiles (CDC). However his weight for height was between 90th and 95th percentile (CDC). The mid parental height was 169.5cms. Head circumference was 52 cm. The ratio of upper and lower segment and arm span suggested being proportionate short stature. Penile length was 2cm and there were no secondary sexual characters. Investigations showed no abnormalities in routine blood, stool and urine examinations. The blood glucose, renal function test, liver function test and thyroid function tests were all normal. No abnormalities were present in the chest X-ray. However, his bone age was delayed (Bone age: 6 years) (fig-1). Clonidine stimulation test (Growth hormone assay) and insulin tolerance test was done which was consistent with Growth hormone deficiency. Insulin like growth factor was below normal (29.47 ng/ml). ACTH, FSH, LH and prolactin levels where within normal limits MRI Brain showed a long narrow truncated stalk and an ectopic posterior pituitary (fig-2). Growth hormone (nordilet) 3 Units was started subcutaneously, once daily, 6 days in a week in the patient along with zinc. Growth hormone was continued till final height was reached. A reasonable approach to stop Growth hormone therapy is to assume that near final height is reached in boys by the age 16. Regular follow-up was advised so that frequent blood glucose levels and thyroid hormonal assay could be monitored. There was a significant increase in his height on follow up after 7 months and the height velocity was 6 cms following growth hormone therapy (fig 3).

Fig 1 showing: bone age of 6 years

Fig 2 showing: a truncated stalk (long arrow) and an ectopic bright spot(short arrow).
III. DISCUSSION

Natural growth hormone is released in a pulsatile manner from the adenohypophysis spontaneously and in response to various physiologic stimuli. Its production is stimulated by growth hormone-releasing factor and inhibited by somatostatin, which are both produced by the hypothalamus. Growth hormone binds to receptors on hepatic tissue and other cells, stimulating the production of insulin-like growth factor-I either locally or at the site of growing bone. Growth hormone binds to a specific growth hormone–binding protein (GHBP) and circulates. This GHBP is the extracellular portion of the growth hormone receptor. IGF-I binds to one of several IGF-binding proteins (IGFBPs) and circulates almost entirely (>99%) in the bound state. IGFBP-3 accounts for most of the IGF-I binding and this binding protein directly depends on growth hormone. Growth hormone deficiency is an uncommon cause of short stature. In one study done in England and Wales, the approximate incidence of growth hormone deficiency was only 1 in every 30,000 births, about half of the patients had idiopathic deficiency and half had deficiency secondary to intracranial disease. These patients with low serum growth hormone levels associated with maternal deprivation were excluded. Several cases apart from being idiopathic causes for a growth hormone deficiency could be due to CNS tumors including (craniopharyngiomas) and malformations of perinatal trauma, lack of oxygen at birth. Leukemias, CNS trauma, CNS radiation, abnormalities in the hormone receptors and very rarely it may be due to a genetic defects, which in some Instances may also be heredity. Children with GHD usually present with short stature and a low growth velocity for age and pubertal stage. Alternative causes of poor growth needs to be considered and excluded. Age at presentation can vary from the first few months of life to adolescence. Typically the GH-deficient child has increased subcutaneous fat especially around the trunk. The face is immature with a prominent forehead and depressed midfacial development; this is related to the lack of GH effect on endochondral growth at the base of the skull, occiput, and the sphenoid bone. Dentition is delayed. In males the phallus may be small, and the average age of pubertal onset is delayed in both boys and girls. Radiograph shows bone age lower than the chronological age. Growth hormone levels and binding protein levels (IGF-I and IGFBP-3) will show whether the growth problem is caused by a problem with the pituitary gland. MRI of the head can show the hypothalamus and pituitary glands. Treatment with growth hormone will usually result in marked acceleration of linear growth. This is most pronounced in the first two years of therapy. In one study of more than 12,000 children, growth hormone replacement therapy was started at an average age of 9.2 ± 4.1 years and produced an increase in growth velocity from 4.4 cm per year to 10.0 cm per year. The younger the patient at the initiation of treatment and the more severe the growth deficiency, the better the response to early therapy.

REFERENCES


AUTHORS

First author - Dr.P.Sekar, Professor of pediatrics.
Second Author – Dr.P.Karthikeyan, Assistant professor of pediatrics. Karthik.dch@gmail.com
Third Author – Dr.sowjan.M, Post graduate in pediatrics, Sowjan86@gmail.com
Fourth Author – Dr.Mahesh Inbasekaran, Assistant professor of radiology, drmahesh2211@gmail.com