

Acromegaly in a patient with Klinefelter Syndrome

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Introduction

Klinefelter syndrome is known as the set of symptoms that result from two or more X chromosomes in males. There is no known association of this syndrome with GH hypersecretion. The most remarkable symptom is gigantism and it can also be observed in androgen deficient states as such as the Klinefelter syndrome and some more genetic syndromes such as the Sotos syndrome, the Marfan syndrome, the homocystinuria, and the fragile X-syndrome. Acromegaly and gigantism are associated with increased level of GH and serum insulin like growth factor-1. It is correlated with significant morbidity and mortality if left untreated. Herein we presented a case with both Klinefelter and acromegaly

Case

➤ A 40-year-old male with previously known hypergonadotropic hypogonadism due to Klinefelter syndrome (47, XXY) was referred to our clinics with the symptoms of increased ring size, arthralgia, excessive sweating and headache. Physical examination revealed multiple skin tags, mild coarsening of the facial features, soft fleshy hands and interdental separation. He was 186 cm in height and 82 kg in weight.

➤ In the hormone panel, basal gonadotropins were elevated with low plasma testosterone, spot growth hormone (GH) was 4.22 µg/ L serum and insulin like growth factor-1 (IGF1) level was 611 µg/ L which was above the age matched reference range (105-280 µg/L). We performed OGTT with 75 mg oral glucose and the nadir GH was 1.0 µg/L. MRI scan of the pituitary revealed an adenoma 12 mm in size. Adenoma was resected with endoscopy guided transsphenoidal approach and the histopathology was consistent with adenoma stained positive with GH. His IGF-1 was normalized and spot GH was < 1 µg/ L three months after the operation

➤ **Conclusion:** Acromegaloidism was reported in Klinefelter syndrome which was recognized as a condition which resembles acromegaly by its clinical manifestations without excess secretion of GH and somatomedins. Ours was the first Klinefelter case in the literature diagnosed with acromegaly that means clinical tests are indicated in the presence of suspicious findings.

