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Klinefelter’s syndrome with hypogonadotropic hypogonadism

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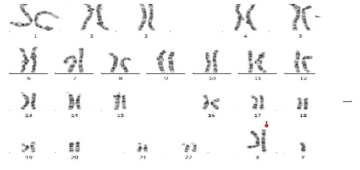
Klinefelter’s syndrome is usually characterised by the signs of androgen deficiency and low serum testosterone coupled with elevated luteinising hormone and follicle-stimulating hormone levels, consistent with hypergonadotropic hypogonadism. Low levels of gonadotropins in karyotypically proven cases are not expected, they are extremely rare occurrences. We report a case of a patient who was diagnosed to Klinefelter’s syndrome (47 XXY) with low gonadotropin levels. Case A 26-year old male was referred to our hospital for the evaluation of obesity. He had a history of surgery at the age of 18 with Rathke's cleft cyst and since then has been taking testosterone intermittently for hypogonadism. And He underwent surgery with gynecomastia at the age of 20. Serum LH, FSH and testosterone levels were low, indicating hypogonadotropic hypogonadism. But, he was eunuchoidal appearance with small and firm testes. So, chromosomal analysis was performed with suspicion of Klinefelter’s syndrome. The result showed that a 47, XXY karyotype which confirmed Klinefelter’s syndrome. We report a case of Klinefelter’s syndrome with delayed diagnosis due to low gonadotropin level caused by Rathke’s cleft cyst.

Peripheral blood karyotyping
- 47 XXY pattern



2010.11.19 sella MRI -Rathkecleft cyst