

Klinefelter Syndrome with Morbid Obesity Before Bariatric Surgery: A Case Report

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Received 2016 November 24; Revised 2017 February 10; Accepted 2017 February 18.

Abstract

Introduction: Klinefelter syndrome is a disorder of chromosomes in which common karyotype is (47XXY). Hypogonadism, gynecomastia and azoospermia could be detected in this syndrome. Decreased basal metabolic rate and interest in activities, loss of muscle mass, weight gain, the deficiency of sex hormone and mood changes cause obesity which cause morbid obese in this case.

Case Presentation: A 34-year-old morbid obese (BMI = 60.40) male was come to the Laparoscopy research center of Iran University of Medical science at 26 January 2016. He was diagnosed as Klinefelter syndrome by genetic testing (47XXY karyotype). He reported suffering from knee cellulitis, headache, low back pain and varices. His nutrition habits was fast eating, Sweet eating and Snack eating. The beginning of his obesity was in his childhood. The best treatment for his obesity and its side-effects is altering in his life style, low calorie diet. Finally, if these methodes fail to lose weight, bariatric surgery is suggested.

Conclusions: In morbid obese patients appropriate diet, change in calorie intake and life style should be considered as a first line of treatment and finally, surgery may be an option to treat obesity. These two methodes can often help reduce the risk of other diseases (e.g., diabetes, heart disease, and sleep apnea) that are associated with severe obesity.

Keywords: Klinefelter Syndrome, Morbid Obesity, Bariatric Surgery, Case Report

1. Introduction

Klinefelter syndrome (KS) is a disorder in which male physical and cognitive development changes and signs differ individually. (47 XXY) is the most karyotype in this syndrome (1). KS is the most common sex chromosomal disorder. Mostly, the extra copy of X chromosome cause this genetic syndrome (2). KS usually remains unrecognized and only of 25% KS patients are diagnosed (3).

Possible symptoms are as follows: testicular dysfunction, azoospermia, decreased libido, gynecomastia, increased plasma gonadotropins in males, increased risk of the metabolic syndrome, and change in body composition, with accumulation of body fat and decreased muscle mass (1, 4). Risk factors for metabolic syndrome identify in childhood and include obesity (5). This disease presented itself in childhood by delayed development of speech, learning disabilities and they have infertility, reduced body hair, gynecomastia, and tall stature later in their life (6).

Phenotypes of this disorder are: Small testicles, disability in sperm producing, gynecomastia, decrease of serum testosterone, increases of luteinizing hormone (LH) and follicle stimulating hormone (FSH) (5).

Testosterone replacement therapy may be used as medical treatment for those who have low level of this hormone and consequences of hypogonadism (7). Failure to produce sex hormones can cause metabolic syndrome, obesity, and diabetes mellitus (5). Testosterone treatment is recommended to reduce abdominal fat and increase lean mass and muscle strength (7).

Here we report a case of Klinefelter syndrome with morbid obesity before bariatric surgery.

2. Case Presentation

2.1. Patient Information

A 34-year-old morbid obese (BMI = 60.40, Weight: 225 kg, Height: 193 cm) male came to the Laparoscopy research center of Iran University of Medical science at 26 January 2016. He reported suffering from knee cellulitis, headache, low back pain and varices. His nutrition habits was Fast eating, Sweet eating and Snack eating. The beginning of his obesity was in his childhood. He is non-smoker and nondrinker and has no addiction to drugs. He take ciprofloxacin and clindamycin medicine.

2.2. Diagnostic Assessment

He diagnosed as Klinefelter syndrome by genetic testing (47XXY karyotype). Hormone testing shows increases in luteinizing hormone (LH = 14, normal rate: 2 - 9.5 mL) and follicle stimulating hormone (FSH = 21 normal rate: 3 - 8.7 mL) and decreased in serum testosterone (= 7, normal rate: 9 - 30 ng/dL), other lab tests are mentioned in table 1. Fatty liver grade 1 was diagnosed by Sonography. Doppler Sonography shows significant back flow at saphenofemoral junction, great saphenous vein, insufficiency in medial leg perforators veins, medial legs superficial varicose veins and gaiter venous ulcers. There is no skin pigmentation and no sign of DVT (deep vein thrombosis).

Table 1. Laboratory Results Two Weeks After the First Visit

Results	Rate
Hb: 14.8	Normal rate: 13.5 - 17.5 g/dL
Vit D3: 20.2	Normal rate: 20 - 30 ng/mL
HDL: 35	Normal rate: 40 mg/dL <
Ferritin: 56	Normal rate: 15 - 200 ng/mL
FBS: 103	Normal rate: 70 - 105 g/dL
HbA1c: 5.6	Normal rate: 4 - 6 %
BUN: 9	Normal rate: 8 - 23 mg/dL
Zn: 80	Normal rate: 70 - 102 μ mol /L
SGOT: 36	Normal rate: 5 - 40 U/L
SGPT: 36	Normal rate: 7 - 56 U/L
Alk.ph: 68	Normal rate: 45 - 115 U/L
Uric Acid: 10.9	Normal rate: 3 - 7 mg/dL
PTT: 36	Normal rate: 25 - 30 s
PTH: 78.38	Normal rate: 10 - 70 pg/mL
INR: 1.2	Normal rate: 0.8 - 1.2
U/C: abnormal + protein	
TSH: 6	Normal rate: 0.4 - 5 μ units/mL
Choles: 181	Normal rate: 120 - 199 mg/dL
LDL: 131	Normal rate: 130 mg/dL >
TG: 91	Normal rate: 150 mg/dL >
Albumin: 4.31	Normal rate: 3.5 - 5 g/dL
Vit B12: 619	Normal rate: 110 - 1500 pg/mL

2.3. Physical Examination

Physical examination shows small genital (hypogonadotropic hypogonadism).

2.4. Interventions

As common before surgery low calorie diet was suggested, Surgery usually is considered as a treatment option for patients with a BMI of 40 kg/m² or more who fails to lose weight by exercise or diet program.

Testosterone therapy and psychiatrist visits were done for him. The millon clinical multiaxial inventory - third edition (MCMI-III) test has shown that he has obsessive characteristic and anxiety due to his morbid obesity. Moderate exercise and changes in lifestyle were recommended to him. Morbid obesity is serious and potentially life-threatening conditions. A healthy lifestyle that includes a healthy diet and mild exercise is important for him. In the first visit we recommended him to change his wrong habits and two weeks later we suggested him to use a diet by administered calories (1800 calories), besides change in lifestyle and healthy diet is recommended for him. A consulting session about long-term side effects of surgery, such as possible need for reoperation, gallbladder disease, and malabsorption was conducted.

2.5. Follow up and Outcomes

Weight loss was followed and after 4 months his BMI decreased from 225 kg/m² to 194.87 kg/m². Weight lost during 6 weeks was significant (change score was 30.2 kg/m²). Our future perspective is bariatric surgery. Bariatric surgery is an option to treat this morbid obese patient. It can often help reduce the risk of other additional diseases such as diabetes, heart disease, and sleep apnea that are concomitant with severe obesity (e.g.).

3. Discussion

Klinefelter syndrome is the most common sex chromosome anomaly and is associated with weight abnormalities. The most patients with Klinefelter syndrome have (47XXY) karyotype (3). The number of X chromosomes in Klinefelter syndrome patients affects phenotypes and severity of symptoms (5). Morbid obesity is not common in these patients, and it is likely related to the X chromosome structural changes (3). Failure to produce sexual hormones, low body energy and decreased interest in activity and other symptoms of this disorder can cause obesity. This rare case is presented here to emphasize both the significance of genetic model of morbid obesity and the need for assessment of comorbidities in such patients.

In this case recommended treatments for obesity are change in eating habits, lifestyle and doing exercise. Then after the final approach was bariatric surgery. This case report demonstrates appropriate treatment for this patient and the important role of sexual hormones and

chromosomal abnormalities on weight changes, besides changing in eating habits is also crucial in their treatment and weight loss too.

Recent studies report KS is associated with abdominal obesity and increased risk of metabolic syndrome. Although patients with KS have abdominal adiposity, their BMI is normal and decrease in muscle mass, increase in body fat and change in body composition are common in these patients (4, 8). Testosterone treatment causes decrease in fat mass. Increased body fat mass in KS is seen in childhood and adolescence (5).

Information is little about the impact of KS on weight and it is not clear that this morbid obesity is related to chromosomal abnormalities, hypogonadism or other hormonal conditions. The aim of reporting present case is to examine whether KS is associated with morbid obesity and how a suitable diet can help this person.

This is a controversial issue that morbid obesity is caused by KS itself or affected by hypogonadism (7).

However, the association between KS and morbid obesity is vague; this article is the first description of KS that is associated with morbid obesity. Also an important role of dietitians who work and consult with morbid obese patients, to investigate the possible association between morbid obesity and KS is raised.

Acknowledgments

Special thanks to the person who participated in this study and Laparoscopy research center of Iran University

of Medical Science. Patient's name is not mentioned in this article and consent form was obtained from him.

References

1. Capasso F, Panetta F, Ierardo G, Parisella V, Polimeni A. Klinefelter syndrome: Case report. *Oral Implantol (Rome)*. 2009;2(3):27-36. [PubMed: 23285366].
2. Zoller B, Ji J, Sundquist J, Sundquist K. High Risk of Venous Thromboembolism in Klinefelter Syndrome. *J Am Heart Assoc*. 2016;5(5) doi: 10.1161/JAHA.116.003567. [PubMed: 27208002].
3. Cho BW, Kwon SE, Kim SK, Lee T, Han JY, Lee JE. Early onset of puberty in an obese boy with Klinefelter syndrome. *Ann Pediatr Endocrinol Metab*. 2016;21(1):39-42. doi: 10.6065/apem.2016.21.1.39. [PubMed: 27104178].
4. Gravholt CH, Jensen AS, Host C, Bojesen A. Body composition, metabolic syndrome and type 2 diabetes in Klinefelter syndrome. *Acta Paediatr*. 2011;100(6):871-7. doi: 10.1111/j.1651-2227.2011.02233.x. [PubMed: 21342256].
5. Bardsley MZ, Falkner B, Kowal K, Ross JL. Insulin resistance and metabolic syndrome in prepubertal boys with Klinefelter syndrome. *Acta Paediatr*. 2011;100(6):866-70. doi: 10.1111/j.1651-2227.2011.02161.x. [PubMed: 21251059].
6. Pradhan D, Kaman L, Dhillon J, Mohanty SK. Mediastinal mixed germ cell tumor in an infertile male with Klinefelter syndrome: A case report and literature review. *J Cancer Res Ther*. 2015;11(4):1034. doi: 10.4103/0973-1482.150697. [PubMed: 26881632].
7. Chang S, Skakkebaek A, Gravholt CH. Klinefelter Syndrome and medical treatment: hypogonadism and beyond. *Hormones (Athens)*. 2015;14(4):531-48. doi: 10.14310/horm.2002.1622. [PubMed: 26732150].
8. Han SJ, Kim KS, Kim W, Kim JH, Lee YH, Nam JS, et al. Obesity and Hyperglycemia in Korean Men with Klinefelter Syndrome: The Korean Endocrine Society Registry. *Endocrinol Metab (Seoul)*. 2016;31(4):598-603. doi: 10.3803/EnM.2016.31.4.598. [PubMed: 28029029].