

HAIR-AN SYNDROME RESISTANT TO BOTH MEDICAL AND SURGICAL TREATMENT. A CASE REPORT

Medikal ve cerrahi tedaviye dirençli bir hair-an sendromu: olgu sunumu

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ÖZET

HAIR-AN sendromu; hiperandrojenizm, insulin rezistansı ve akantozis nigrikans ile karakterize bir endokrinopatidir. Bu sendromun primer patofizyolojik bozuklukları hiperandrojenizm ve insulin rezistansıdır, akantozis nigrikans ise bu patolojilere eşlik eden cilt değişiklikleridir. Bu makalede 10 yıldır polikistik over sendromu ile beraber olan ve medikal ve cerrahi tedaviye dirençli olan bir HAIR-AN sendromu sunulmuştur. Her türlü tedaviye rağmen gerilemeyen hiperandrojenizm olgusunda; ilk olarak overyan wedge rezeksiyon uygulanmış olup şiddetli hiperandrojenizmin devam etmesi üzerine medikal endokrinoloji ile ortak karar alınarak bilateral salpingoofektomi yapılmış ve gelecekteki doğurganlığı korumak için çıkarılan over dokusu dondurulmuştur. Ovaryan kriyo endikasyonları içerisinde literatürde ilk tanımlanan vaka olması açısından bu olgu önem arz etmektedir.

Anahtar Kelimeler: polikistik over sendromu, hiperandrojenizm, hirsutizm, insülin rezistansı

ABSTRACT

HAIR-AN syndrome is an endocrinopathy which has characteristics of hyperandrogenism, insulin resistance and acanthosis nigricans. In this syndrome the primary pathophysiological disorder is hyperandrogenism and insulin resistance, acanthosis nigricans is a skin modification to these pathologies. In this article a 10 year old medical and surgical treatment resistant HAIR-AN syndrome with polycystic over disease is represented. Despite to all medical treatment methods no regression was seen because of this ovarian wedge's resection is performed first. After this operation, severe hyperandrogeny continued as a result of this associated decision taken with medical endocrinology and salpingo-oophorectomy is performed. Her ovarian tissues are cryopreserved to conserve her fertility for future. In the indications of ovarian cryopreservation this case is first in the literature which makes this case an important case.

Key words: polycystic ovary syndrome, hyperandrogenism, insulin resistance, hirsutism

INTRODUCTION

Hair –An syndrome : An endocrinopathy characterised by hyperandrogenism, insulin resistance and acanthosis nigricans. The primary pathophysiologic disorders of this syndrome are insulin resistance and hyperandrogenism. Acanthosis nigricans is a skin disorder that is found together with these pathologies. In this article a case of a Hair-An, polycystic Ovarian Syndrome patient who has been resistant to both medical and surgical treatment for the past 10 years is presented.

Case Report.

A 29 year old patient with a menarche age of 14, hairy chin at the age of 18 presented with acne, frontal baldness (fig 1), temporal alopecia, oily skin, clitoral hypertrophy (Fig 2) and colour changes in the inguinal and axilla regions and was diagnosed with Polycystic Ovarian Syndrome.

Figure 1



Figure 2



Oral contraceptive treatment was started and the patient was followed up with OC treatment as regular cycles were observed. Metformin was added to the treatment

protocol after 5 years as the patient developed insulin resistance and irregular menstrual cycles. Laboratory tests done due to the amenoreik state of the patient

irrespective of medical treatment for the past one year showed FSH 3.45 IU/L, LH 6.68 IU/L, total testosterone 221.9 ng/dl, Free testosterone 6.8 pg/mL, prolactin 12.9 ng/ml, DHEA-SO₄ 411.5 µg/dl, 17-hydroxyprogesteron 2.57 ng/ml and estradiol as 20 pg/ml. Tumor markers fell into the normal ranges and suprarenal Computed tomography was normal. Due to higher levels of 17-OH progesterone a differential diagnosis of adrenal hyperplasia was made but a normal ACTH stimulated test was reported. Karyotype analysis was normal and both ovaries were seen to be larger than normal in the pelvic ultrasound. Diagnosis of HAIR-AN syndrome was settled on and further investigations to rule out ovarian stromal pathology and microenvironmental androgenicity were made. Laparoscopic Bilateral Ovarian Wedge resection was performed and theca cell hyperplasia was reported as the pathology.

Follow up after surgery observed non decreasing androgen levels and the patient was presented to an endocrinology council for consultation who suggested bilateral ooforectomy. Bilateral ooforectomy was performed. To protect the fertility of the patients the removed ovaries were frozen in our clinic using the slow freezing method.

DISCUSSION

In this article a case of an HAIR-AN syndrome resistant to all types of antiandrogenic treatments showing higher levels of androgens and frontal alopecia even after Ovarian Wedge Resection is presented. Polycystic Ovarian Syndrome is the most commonly seen endocrinopathy in the reproductive ages.(1) Polycystic Ovarian syndrome is a hyperandrogenic state characterised by polycystic ovarian morphology and chronic oligoanovulation (2,3). Polycystic Ovarian Syndrome is commonly found with insulin resistance and compensatory hyperinsulinemia which are major factors in androgen production.(4) Patients are at a higher risk

of infertility, dysfunctional Uterine bleeding and metabolic diseases such as Diabetes Mellitus, Dislipidemia and Hypertension.(5) In the presence of Hyperandrogenism, insulin resistance and acanthosis nigricans we see Hair-An Syndrome. There has been a moderate increase in the number of women with anovulatory Plasma testosterone levels and hirsutism. PCOS women with Free and unbound testosterone levels are almost twice that of the normal populace.(6) Treatment with a low dose oral contraceptives has been found to be effective in the treatment of Acne and hirsutism. Suppressing free testosterone levels with oral contraceptives is clear.(7,8) By creating a microenvironment in the presence of high Leutinizing Hormone and testosterone levels during a laparoscopic ovarian Drilling procedure (destroying theca cells) a fall in testosterone levels was shown. (9,10) In the presence of high androgen levels in treatments that are resistant to medical treatment surgical diathermy is used as an alternative treatment. Anovulation dependent hyperandrogenism should be considered when levels of testosterone are below 200ng/L and a differential diagnosis of an androgen producing tumor and diagnostic laparoscopy should come to mind in cases where testosterone levels are above 200ng/L. Eventhough laparoscopy and ovarian biopsy is not an indicated during the work up of diagnosing hirsutism differential diagnosis of a tumor or preparation of a microenvironment by ovarian biopsy or drilling could be thought of. However in this article in spite of performing a bilateral ovarian wedge resection a resistant HAIR-AN case with anovulation non regressing clinical symptoms and a high degree frontal baldness is presented. In cases like this further studies on androgen and insulin receptor polymorphism could be useful. However since menopause has been induced in the patient surgically ovarian tissue has been frozen to protect the

fertility of the patient in the near future. Among the indications for ovarian cryo this case presented is the first in the

medical literature and should be considered with such importance.

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