Premature Ovarian Failure in a 16 year old girl

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Citation

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Abstract

Ovarian failure is normal during menopause but pathologic before 40 years of age. The authors report a case of ovarian failure in a 16 year old girl, most likely due to Savage Syndrome, who started hormonal replacement for normal development of sexual characteres and for osteoporosis prevention.

INTRODUCTION

Ovarian failure is a normal occurrence during menopause, and this event is determined by genetic inheritance. Ovarian failure occurs before 40 years of age in 1% to 5% of women and is considered pathologic – premature ovarian failure. If ovarian failure occurs before puberty, the patient`s breasts will not develop and gonadal agenesis results.

Despite the array of causes of ovarian failure, in most cases the etiology cannot be determined. The pathogenesis of spontaneous ovarian failure in most cases is unknown. Two mechanisms are presumed to play a role—follicle depletion and follicle dysfunction. If follicle deplection is suspected, the causes can be low inicial follicle number (pure gonal dysgenesis, thymic aplasia/hipoplasia or idiophatic) or accelerated follicle atresia (Turner Syndrome, autoimmunity, viral infections and others). In the case of follicle disfunction, we have to consider steroidogenic enzyme defects, autoimmunity, signal defects, specific gene defects and idiophatic (resistant ovary syndrome or Savage Syndrome).

We present a case of premature ovarian failure in a 16 year-old girl and discuss the possible etiology, treatment and follow-up in these cases.

CASE REPORT

A 16 years old patient was sent by her doctor to our appointment because of primary amenorrhea. Her pubarche was at 14 years of age but she had no axillary hair, and the breast development was Tanner stage 2. She had no signs of hirsutism or galactorrhea. She had not started sexual intercourse. There was no history of infections, autoimmune disorders, radiation or chemotherapy. The gynecological examination revealed genital atrophy, with a apparentely normal vagina. Her pelvic ultrasound revealed a small tubular uterus and the ovaries had a volume of 2.35 cc and 2.44 cc.

The thyroid function was normal, as well as the prolact, cortisol and SO4-DHEA (dehydroepiandrosterone sulfate). She had high levels of FSH (follicle-stimulating hormone) and LH (luteinizing hormone) and low estrogen and progestin levels.

The karyotype was determined – 46,XX

Figure 1

Picture 1 – The girl was Tanner Stage 2

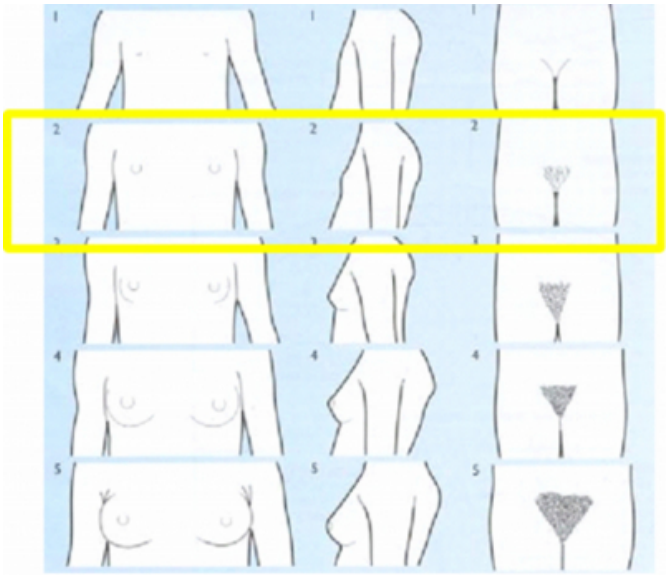
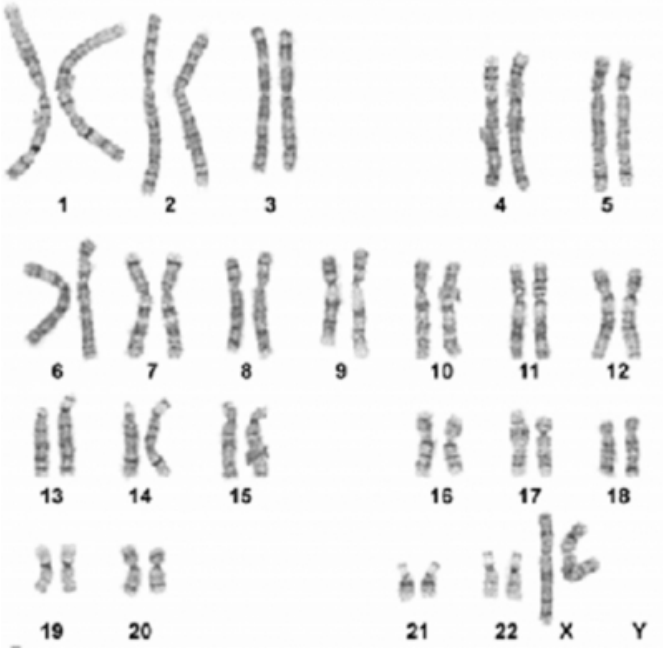


Figure 2

Picture 2 – Karyotype was 46 XX – normal



DISCUSSION AND CONCLUSION

This 16 year-old girl had a normal karyotype, a partial development of secondary sexual characteristics and hypergonadotropic hypogonadism. This case is compatible with premature ovarian failure, probably a gonadotropin resistance, also referred to as Savage Syndrome, likely due to FSH receptor dysfunction.

Biopsy is not advisable because diagnosing resistant ovarian failure will not affect management. We initiated in this case, as indicated in individuals with primary amenorrhea associated with all forms of gonadal failure and hypergonadotropic hypogonadism, cyclic estrogen and progestin therapy to initiate, mature and maintain secondary sexual characteristics, with good results. Prevention of osteoporosis is an additional benefit of estrogen therapy.

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