Case Report
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Case presentation & review of literature for a rare case of adolescent premature ovarian insufficiency

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Introduction

Premature ovarian insufficiency (POI) in a well adolescent is an extremely rare event and its occurrence raises important questions about causation, which may signal other systemic concerns. Its occurrence may have a significant impact on a woman's future health, fertility and motherhood. Beyond the psychological devastation and the symptomatic havoc that dominate the "now" lie long-term health implications of relatively swift and sometime dramatic loss of ovarian function seen with it.

The background age-specific incidence of idiopathic POI in early to mid-adolescence is so rare as to be also unknown, with the annual incidence reported as 10/100 000 in girls younger than 20 years.[1] Whether spontaneous or induced, POI renders a woman deficient in ovarian endocrine and reproductive function and hold myriad implications, both short and long term. In literature there is limited research and very few cases reported with adolescent POI. In view of its inimitability we hereby present a very interesting and unique case of POI in Miss A, an adolescent girl.

Case presentation

Miss A attended gynaecology outpatients with secondary amenorrhoea and episodes of hot flushes and sweats on and off for 9 month duration. It as preceded by one episode of prolonged heavy periods for which was treated with provera by general practitioner for one month following which amenorrhoea developed. She attained menarche at the age of 11 years and had regular menstrual cycles thereafter. She has not had any medical problems in the past and is not known to be on regular medications. There is no contributory surgical or family history. She has two elder sisters absolutely well and fine.

Examination showed well-developed secondary sexual characteristics with a normal BMI. Systemic examination was unremarkable. A battery of investigations conducted on her did not reveal any reason for her complaints. Raised levels of follicle stimulating hormone and low serum estradiol levels done 6 weeks apart-confirmed diagnosis of POI. Subsequently she was referred to tertiary care centre where they further investigated and started her on oral combined oral contraceptive pill.

Discussion & Review of literature

POI is defined as depletion or dysfunction of ovarian follicles with cessation of menses before age 40 years, and it has previously been referred to as premature menopause or primary ovarian failure or premature ovarian failure. POI is a pathologic condition that should not be considered a hastening of natural menopause. Its incidence is idiopathic in 74-90 percent of cases but can be familial or sporadic in about 4-33 percent. Some of the known causes are genetic aberrations, autoimmune ovarian damage, iatrogenic following surgery, radiotherapy or chemotherapy, environmental factors like viruses and toxins and metabolic causes like galactosaemia, 17 OH deficiency.[2]

Clinically most cases POI manifest after attainment of normal pubertal development and after the establishment of regular menses, however 10% can present with primary amenorrhoea. Menstrual irregularities may also be accompanied with constant or intermittent hypoestrogenic symptoms that define the vasomotor instability as happened with Miss A.[3,4] Occurrence of POI generally has been related to either follicle dysfunction or follicle depletion. Follicle dysfunction indicates that follicles remain in the ovary, but a pathologic process prevents their normal function (as a result of an FSH-receptor mutation). Follicle depletion indicates that no primordial follicles remain in the ovary.[5] Miss A had a normal pelvic scan indicating of follicle dysfunction to be the likely mechanism. In few case studies its occurrence in adolescents has also been linked to HPV vaccination; however there has been no clear mechanism stated and may require more research in that area,[6,7] interestingly Miss A did have HPV vaccine as a part of national immunisation.

There is no consensus on criteria to identify primary ovarian insufficiency in adolescents, and delay in diagnosis is common. To diagnose it there needs to be an evidence of persisting hypergonadotropic hypogonadism, established through assessment of circulating levels of the follicle-stimulating hormone and serum estradiol six weeks apart. Gonadotropin and estradiol values may be altered by concomitant use of hormonal preparations and thus should only be obtained in patients who are not taking hormonal medications**.** Before that other causes that need to be ruled out are pregnancy, thyroid abnormality, adrenal insufficiency, parathyroid disease, genetic aetiology, diabetes. Assessment of hypothalamo-pituitary ovarian axis, skeletal health and likelihood of resumption of ovarian function need to be considered.[4] A multidisciplinary approach is paramount with professionals from various disciplines and specialties participating to address the myriad requisites of the affected population (psychological support, fertility management, evaluation and management of coexisting endocrinopathies, prevention and management of chronic health sequelae). Individualized needs of the afflicted family may additionally merit attention and should not be discounted.[8]

For adolescents with primary ovarian insufficiency, the objective of treatment is to replace the hormones that the ovary would be producing before the age of menopause, making the treatment distinctly different from hormonal therapy for menopause that focuses on the treatment of menopausal symptoms. Regardless of the etiology, patients with primary ovarian insufficiency are oestrogen deficient. Thus, young women with primary ovarian insufficiency may need higher doses of oestrogen than menopausal women to ensure adequate replacement and optimal bone health. There have been various different views on dosage, combination and route of administration. Transdermal, oral or occasionally transvaginal estradiol can be considered to mimic a physiologic dose range and to achieve symptomatic relief. Although data from randomized, controlled trials are lacking, most experts agree that physiologic oestrogen and progestin replacement is reasonable in the case of young women with primary ovarian insufficiency and should be continued until they reach the age when menopause usually occurs.[9] Occasional resumption of ovarian activity can occur and pregnancy can happen in about 5-10% of these women.[10]

A long term follow up of bone density in women in POI concluded the stability of bone mass throughout a period of 8 years in women taking combined HRT however experiencing early osteopenia or osteoporosis, explaining the need for drug therapy.[11] There are no published data to support specific recommendations for dual-energy X-ray absorptiometry scanning in adolescents with oestrogen deficiency. Some experts suggest monitoring bone density annually in adolescents with oestrogen deficiency during early to mid puberty to document peak bone accrual and then every 2 years in late adolescence.

Data in the adolescent population are lacking and there are no standard screening regimens for cardiovascular disease in this population, vigilant monitoring is warranted, and practitioners should help patients optimize cardiovascular health. Hence achieving a heightened level of awareness of symptomatology, timely diagnosis, counselling and intervention may alleviate few of the consequences and long term sequelae. A greater understanding of female reproductive biology and the physiologic effect of primary ovarian insufficiency enable health care providers to offer counselling for these young women. Once primary ovarian insufficiency is diagnosed, patients should be evaluated at least annually. Physicians should address the special needs of this population and counsel family members and patients on the risk of co morbidities associated with primary ovarian insufficiency and the condition's potential for genetic inheritance. Referral to accurate medical information is encouraged. Achieving a heightened level of awareness of symptomatology, timely diagnosis, counselling and intervention may alleviate few of the consequences and long term sequelae. Large-scale genomic sequencing has recently identified new mechanisms of POI which will change care as genomic medicine is now being integrated into standard care.[12]

Acknowledgments

None.

Conflicts of interest

None.

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