INTRODUCTION

Primary amenorrhea is the absence of menstruation by 16 years of age in the presence of secondary sexual characteristics or by 14 years if the secondary sexual characteristics are not developed.\(^1\)\(^2\) The incidence varies from 0.48 to 1.2\%.\(^3\) Two local studies have quoted the incidence as 0.005 to 0.06.\(^3\)\(^4\)

For the onset of normal menstruation, intact hypothalamic pituitary ovarian and uterine axis and 46XX chromosomes is necessary. Patent genital tract outflow is also important. The various causes of primary amenorrhea range from abnormal karyotyping to absent uterus. This is an unusual case of an anatomically normal uterus and outflow tract with bilateral andometriomas presenting as primary amenorrhea.

CASE REPORT

Mrs. MN, 23 years old, was married 4 years ago. She came with primary amenorrhea for which she never consulted any gynaecologist. She was also complaining of cyclical pain in lower abdomen which usually subsided by rest and analgesics. She also had cyclical mastalgia. Her marital relations were satisfactory. Her sisters had normal menarche. On examination, her height was 5'2". Breasts were well developed. Secondary sexual characteristics were like normal female. Abdominal examination was normal. On pelvic examination vagina was well formed and of normal length. Cervix was felt. Uterus was normal size. Bilateral adnexal masses were felt.

Her ultrasound report showed normal size uterus with 4 mm thick endometrium. There was bilateral adnexal masses, right one was 8 x 8 cm and left 4 x 4 cm. Her serum FSH, LH and Prolactin levels were within normal range. Karyotyping was 46XX, EUA before laparoscopy showed normal external genitalia.

On laparoscopy, her uterus was normal size, both tubes were normal looking and there were bilateral endometriotic cysts, sizes were same as mentioned in ultrasound report. No other endometriotic spots or adhesion were seen in the pelvis. Laparotomy was proceeded and cysts were drained, with stripping of capsule and reconstruction of both ovaries. Histo-pathology of tissue confirmed endometriosis.

Her postoperative period was uneventful. After 6 weeks she was given trial of combined oral contraceptive pills, (75 µg gestodene and 20 µg ethinylestradiol) for a 21-day cycle. She did not show any response to it. She was counseled about her problem and advised about the proper follow-up.

On her next visit another cycle of same oral pill was repeated. It was planned to do hysteroscopic guided biopsy of endometrium on next visit if she did not respond to second cycle of pill. Unfortunately she was lost to follow-up and did not come back.

DISCUSSION

Primary amenorrhea has wide range of causes, which need systematic approach for diagnosis. This problem poses imbalance in sexual relationship and fertility on one hand and psychological impact on the person and family on the other hand. Some of the problems can be overcome easily like imperforate hymen while some times a problem can come which can not be managed.

In this case these were many problems to be considered. Patient was genotypically female. There were bilateral endometriotic cysts. The patient was menstruating in the ovaries but not through the...
endometrium. This shows the totipotential nature of epithelium of peritoneal cavity which was responding to the ovarian hormones. Timmreck, did not mention this type of abnormality in 266 cases. A larger study by Petal et al. comprising about 295 cases of primary amenorrhea also did not show such type of abnormality. A case report by Sheikh and Sirichand showed primary amenorrhea with granulose cell ovarian tumour, patient responded well to the removal of tumour.

This could be a case of endometrial hypoplasia as the endometrial thickness was 4 mm. Patient was given a trial of oral contraceptive pills and watched for withdrawal bleeding, but she did not respond. Another cycle of oral pills was also repeated. Baker et al. reported similar case where everything was normal and endometrial biopsy revealed absence of endometrium. A similar type of case has been reported by Tavasuli et al.

It is a common observation that many of the obstructed pathologies of genital tract are associated with pelvic endometriosis with primary amenorrhea.

Many of the questions arise in this unique case like, what will be the next test to confirm the case, endometrial sampling, hysteroscopic guided, endometrial biopsy was planned but patient lost her follow-up. If the endometrium is unresponsive how it will be made responsive? What about the future fertility, will she have recurrent endometriosis. If she could not menstruate and conceive, will she need radical surgery? This case is still a dilemma as far as management is concerned.

REFERENCES