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CASE SERIES OF PRIMARY AMENORRHEA-A HAPPY ENDING **KEY WORDS**:

Obstetrics & Gynaecology

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Introduction: Menstruation is a very important parameter in the development of feminine nature of female sex. Attainment of menarche brings in a lot of confidence in a girl. There are various reasons behind in not attaining menarche. The incidence of primary amenorrhea being less than 1%, it accounts for significant physical and psychological trauma to the girl and the family. With the advancement in the investigation and treatment modalities it is possible to restore the menstrual functions and thereby preventing complications. Though restoration of reproductive capacity is again a question and now with the advancement in various ART techniques, this is also possible now. **Aim:** This study brings in various presentations of primary amenorrhea that are encountered over a period of 6 months at Tagore Medical College and Hospital, Chennai. **Material Methods:** Primary amenorrhea can be due to diverse cause and needed detailed evaluation. We report two such cases of 15 year and 17 year old individuals with primary amenorrhea. This is a prospective study done over a period of 6 months among primary amenorrheic patients. They were investigated and managed and were followed for the treatment response.

INTRODCUTION:

ABSTRACT

Primary amenorrhea is defined as not attaining menarche by 13 years of age with secondary sexual characteristics or beyond 15 years irrespective of secondary sexual characteristics. Menarche starts with thelarche, pubarche, growth spurt and finally resulting in menarche. Majority of cases of primary amenorrhea are due to anatomical defects, elevated follicle stimulating hormone levels, hyper prolactinemia, hypothalamic and pituitary causes.

Gonadal dysgenesis includes Turner syndrome which makes up to 43% of primary amenorrhea cases. Turner syndrome is due to defective X chromosome which is missing partially or completely. Its main manifestation is growth disorder, reproductive abnormalities, cardiovascular and autoimmune diseases. Timely diagnosis is difficult and with the availability of non-invasive prenatal testing ,this can be identified at an early stage. The incidence of primary amenorrhea being less than 1%, it accounts for significant physical and psychological trauma to the girl and the family. With the advancement in the investigation and treatment modalities it is possible to restore the menstrual functions and thereby preventing complications.

Case Report 1:

A 15-year-old girl was brought by her mother to the pediatrics department-Tagore Medical College and Hospital with the complaints of not attaining appropriate height, evaluated with X-ray since her height was found to be less for her age. Bone age was found to be 12 years. Further she was referred to OBG department-TMCH for further evaluation. General top to toe examination done and found that she had webbing of neck, short stature, high arched palate, low hair line, widely placed nipples and fungal infection of the nails. Her mother also had a concern for the girl not attained menarche since the other siblings in the family and the mother herself attained menarche between 12-14 years of age. Provisional diagnosis of Turners syndrome was made. USG was made and found that she had streak ovaries and infantile uterus. Further hormonal profile was done and she was advised Karyotyping. ECHO showed mild Aortic stenosis, trivial aortic regurgitation with ejection fraction of 68% and with normal cardiac chambers. The provisional diagnosis that we made was confirmed with karyotyping report-45X0. Hence diagnosed as Turners syndrome. Patient was given withdrawal bleeding and had her first episode of bleeding. Further the patient was planned for Growth hormone supplementation.

Case Report 2:

An 18-year-old girl brought by her mother to TMCH OBG OPD with the complaint of **not attained menarche**, with **cyclical abdominal pain** and **white discharge** on and off, with

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headache and no vision complaints at present. Patient had prior history of sudden bilateral vision loss and MRI was done and was diagnosed with pituitary adenoma. Visual acuity at that time was no perception of light. For which she underwent right frontal craniotomy with sub frontal approach with excision of the Space occupying lesion. HPE consistent with pituitary apoplexy. Immediate post-op patient vison improved and she was put on Tablet Eltroxin 50mcg.Patient had 3 siblings and no one in the family has similar complaint. General examination was done and found that she is tall with high arched palate with normal head size and shape, acanthosis nigricans with breast tanner stage 1 and no axillary and pubic hair, limb length discrepancy since she had history of fall and fracture with shortening of left lower limb. Further blood investigations done and found that she had low estradiol, low growth hormone and low progesterone levels with hyperprolactinemia of value more than 200ng/ml and TSH of 10.6 m IU/ml. Further she was evaluated with ultrasound where she was found to have infantile uterus with both the ovaries under developed. She was then advised to do MRI-recurrent macroadenoma of pituitary gland with chronic tumor bleed and further with MRI contrast it was found that she had Sella heterogenous enhancing mass lesion with suprasellar enhancing solid component perilesional collection with blooming signal s/o pituitary macro adenoma with tumour chronic bleed. As per neurosurgeon's opinion patient was put on Tab. Cabergoline 2.5mg once weekly and to review after a month for tumour excision. The patient underwent tumour excision after one month and further she was followed up at TMCH regularly and she came up with her first menstrual cycle.

DISCUSSION:

Age at menarche has been constantly declined over a period of time and it is 13 years in India now. Awareness must be created regarding the problems associated with not attaining menarche. According to various studies it has been noted that gonadal dysgenesis as the most common cause for primary amenorrhea. Nowadays the number of cases of Turners syndrome is rising because of the early and easy approach to the clinician and the anxiety of the mothers as the girl has not attained menarche and the required growth potential for her age. Cases with organic lesion in the brain leading to primary amenorrhea has also been noticed over a period of years. The most common being pituitary adenomas as in our case she had recurrent macroadenoma. Early diagnosis and prompt treatment has restored their menstrual capacity.

CONCLUSION:

This case series emphasise the knowledge towards clinical and genetic aspects that are needed for detailed evaluation of patients with primary amenorrhea. Thus, a proper

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management of the case and its associated condition to prevent later complications. Therefore early treatment can bring about normal menstrual cycle and has a great impact on physical and psychosocial enhancement for the family and the society.

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