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Gender Re-assignment at 17
Medical, Ethical, Religious and Cultural Dilemma!

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To The Editor: Gender re-assignment after attainment of puberty is a challenging exercise. The problem is further compounded in places with limited diagnostic and counseling facilities. We report on a teenager who was raised as a boy until he started to have regular menstruation. The difficulties encountered in the management of this case are highlighted.

Case report

A 17 year old male student was referred to the Federal Medical Centre Yola by his school teacher and stepfather because of breast enlargement and periodic perineum bleeding associated with school absence. The Teacher confirmed the story and noticed that the student had been walking with a stoop in an effort to hide the enlarged breasts.

The patient first noticed the breasts enlargement at the age of 13 and a year later started to have a monthly vaginal bleeding for 4 – 6 days. The patient was given a female name at birth, however at the age of two years the family thought that the child had good sized "phallus". The child was taken to a local hospital where the family was told that the gender cannot be assigned until the child is older and therefore he was given a male name and kept company with the same gender. With development of female secondary sexual characteristics around the time of puberty, the parents were convinced that the child had been assigned the wrong gender and therefore they sought medical advice. On examination the patient was depressed and dressed in male attire but had female fat distribution. The growth parameters were normal with height of 163 cm and weight of 58kg. Pubertal assessment showed breast development & pubic hair tanner stage IV. Genital examination revealed a phallus of 6cm with redundant labia majora. The labia minora was almost fused in the midline, partially occluding the vaginal opening. There was also a reducible right inguinal hernia. Rectal examination indicated the presence of a uterus. Further investigations revealed normal electrolytes and total testosterone. There were no facilities to do 17 hydroxyprogestrone (17 OH-P), cortisol, ACTH or chromosomal karyotype however buccal smear was positive for Barr body indicating the presence of two X chromosome. Abdominal ultrasound scan showed normal looking uterus. Both ovaries were visualized and appeared normal with no mass in the suprarenal area. A laparoscopy confirmed the presence of female internal genitalia with normal looking ovaries. A clinical diagnosis of simple virilizing congenital adrenal hyperplasia (CAH) was made and the patient and her family were counseled for gender reassignment. She had clitorovaginoplasty and right inguinal herniorrhaphy as single stage and histopathology confirmed normal corporal tissue. Admission to the gynaecology ward was associated with dramatic psychological outburst but as time went by she gradually acclimatized to the new environment. The patient had accepted the gender reassignment after a series of counseling sessions by the local Imam, parents, teachers, nurses and doctors who individually and collectively helped her through this difficult part of her life.

Comments

This case illustrated the medical and social consequences of delaying the gender assignment of a child with ambiguous genitalia. In retrospect, this stress would have been minimized if the child was referred at an early stage to a pediatric unit with experience in dealing with this emergency. During the management of this case we were faced with two main issues: firstly making the diagnosis with limited resources and secondly dealing with the psychosocial consequences of gender reassignment at this age. As in most developing countries, no adequate psychology service was available to help in the counseling process; however the combined efforts of the local religious leader (Imam), senior members of the community, nurses and doctors were successful. The fact that the parents were convinced before seeking medical advice that the child was actually a female made our task rather easier.

The other challenge of this case was working out the diagnosis. Although making a definite diagnosis in such cases can be difficult without proper diagnostic facilities such as chromosomal karyotyping , 17 OH-P, cortisol, ACTH and urine steroid profile which were not available even at the regional teaching hospital, we utilized our clinical skills and the available facilities to reach a logical conclusion. The history of female gender assignment at birth and the development of female secondary sexual characteristic at the expected time with menarche at 14 years old indicated that we are dealing with a virilized female. This also suggests that the hypothalamic pituitary ovarian axis is intact and that the outflow tract is patent. The combination of normal female internal genitalia, Clitoromegally in a person with 2 X chromosomes, as indicated by positive barr body on buccal smear, and normal electrolytes were in keeping with simple virilizing (none salt wasting) type of CAH. The total testosterone level was within the normal female range, however neither free testosterone nor dihydrotestosterone which acts on the external genitalia were available. Although patients with Turner's mosaic 45XO/46XX or mixed gonadal dysgenesis may have presence of Barr bodies, the clinical phenotype, Clitoromegally, normal sized uterus and ovaries and regular period can not be explained by either [1,2]. The development of menarche has ruled out the

possibility of androgen insensitivity syndrome (46XY). The fact that the Clitoromegally was observed at 2 years of age and persisted until puberty suggested that maternal androgen ingestion during pregnancy was an unlikely possibility in this case.

To reassign a sex for a 17year old who had been raised with the "dominant" gender one has to be unequivocal regarding the potentials for fertility and capacity for normal sexual function [3]. Although the size of the phallus was 6cm, the capacity for fertility without a penile urethra is remote. For an adolescent, the surgical management was aimed at achieving near normal cosmetic and function. Historically, clitoral reconstruction has evolved from clitoridectomy, through clitoral recession and now clitoral reduction. The disadvantages of clitoridectomy and clitoral recession are the loss of sensation, anorgasmia and pain following clitoral engorgement at time of sexual arousal respectively [4,5] Clitoroplasty with preservation of the neuromuscular bundle the Spence-Allen technique obviates both of these risks and was the procedure performed on this patient.

In conclusion we reported on our experience in dealing with a case of medical, social and ethical dilemma particularly in places with limited facilities. Although our task would have been easier if the patient was younger and resources were available we showed that clinical skills together with combined efforts of various parties can be useful in places like ours. We suspect that some of our colleagues, in developing countries, had similar experience and hope that health care policy makers' pay more attention to this sensitive area.

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