J. of Obst. & Gyn. of India Vol. 51, No. 3: May/June 2001 Pg 177-178

Intrauterine Adrenogenital Syndrome

Madhu Nagpal, Surat Kaur, Davinder Pal

Govt. Medical College, Amritsar.

A 22 years old girl was brought to the O.P.D. by her parents next day after her marriage with complaint of abnormal external genitalia & inability to do intercourse as complained by her husband. Her menarche was six years back with regular menstrual history. Her menstrual cycle was 3-5/30. Breast development was normal but hair growth was excessive and coarse especially in pubic and axillary region.

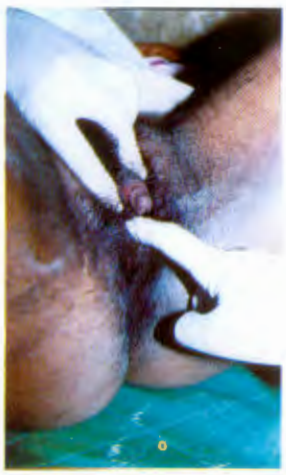

Fig. 1 : OPD presentation showing erected phallus with narrow opening and midline fusion line below it

On local examination, phallus was hypertrophied to 2.5cm x 2cm in dimensions. Examination easily aroused marked erection of phallus to 4cmx2.5cm in dimensions with distinctly palpable median raphe on its inferior aspect. Labia were unidentifiable & labial folds of the two sides were fused to form a membrane covering the introitus leaving a small opening below the phallus. This opening represented common outlet for the menstrual blood and urine. Finger tip could be inserted in the opening. (Fig 1). Family history revealed presence of similar enlarged phallus in one of the patient's sisters but she had normal sexual life and was having three children.

Patient was fully investigated. Hb=11.2gm%, urine C/E NAD. FBS=92.mg%, Bld. Urea=32mg%, S.Cr=1.3mg%, U/S abdomen revealed normal uterus, ovaries & vagina and urinary system was also normal. Buccal smear was positive for Barr Bodies. Urinary 17 Ketosteroids = 15.5mg/24hrs. (normal range 9-24). Pregnanetriol levels could not be determined due to non-availability of the test. Surgery was planned to give the patient feminine genitalia matching her female functional performance.

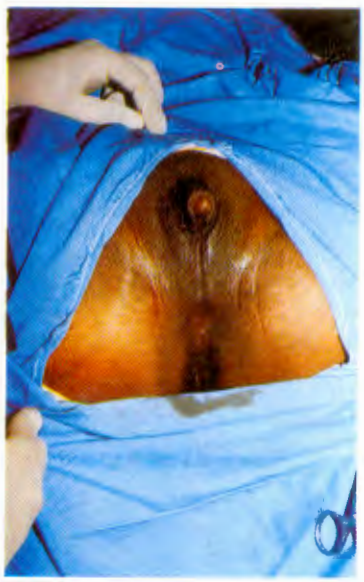

Fig. 2 : Showing external genitalia as appeared prior to surgery

Fig. 3 : Showing formation of vaginal introitus, urethral meatus with catheter in situ and dissection of phallus

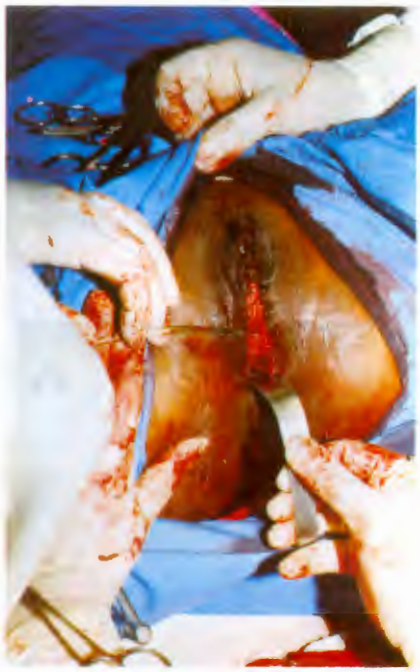

Fig. 4 : Showing vaginal passage, urethral opening and restructured clitoris after surgery

Under spinal anaesthesia membrane representing fused labial folds was incised in the midline exposing vaginal and urethral openings. Hymenal ring became visible. Labia reconstructed by forming a new mucocutaneous junction. P/V showed normal uterus and adnexa though vagina accommodated two fingers tightly. Enlarged phallus was excised. Clitoral vessels ligated at the base. Overlying skin repaired and restructuring done to mimic clitoral folds (Fig 2,3 & 4). Patient was discharged with proper instructions for consummation of marriage after proper healing.

Intrauterine adrenogenital syndrome is a familial disease transmitted by a recessive gene. Due to excessive production of androgens in female foetus, sex differentiation of external genitalia i.e. vulva and introitus are affected. Internal sex organs are normal. The above case fitted well into this syndrome presentation and is thus being reported.