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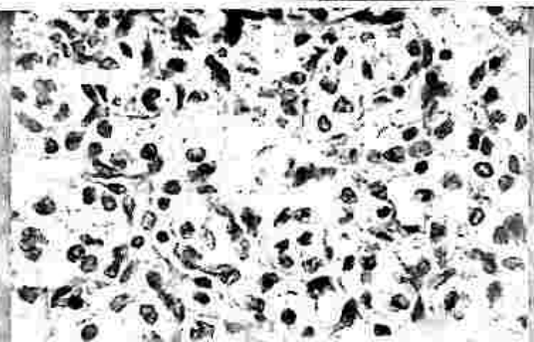
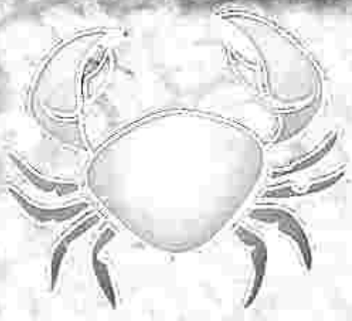
CASE REPORT





EXPERIENCE WITH INTERSEX SURGERY AT BINGHAM UNIVERSITY TEACHING HOSPITAL, JOS: A CASE REPORT

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Abbreviations:
CAH: congenital adrenal hyperplasia
DSD: disorder of sex development, ICSI: intracytosplamic sperm injection
BHUTH: Bingham University Teaching Hospital

ABSTRACT

Intersex, describes variations in sex characteristics including chromosomes, gonads and genitals that do not fit typical binary notions of male or female beings.
The management of the condition can be very challenging. Reports of the surgical treatment are particularly scarce.
Objective: To report a successful surgical management of a case of intersex in a young Nigerian.
Case Report
A 15 – year- old boy presented with ambiguous genitalia and breast enlargement. History revealed that he was born with a small penis and lack of testes on one side. He gave a history of monthly menses and having to urinate while standing. He was assigned a male gender, even though he developed large breasts.
Conclusion: Biopsy results showed ovotestis of the ovary consistent with truehermaphroditism.

INTRODUCTION

Background/ Masculinizing Surgery

Intersex describes variations in sex characteristics including chromosomes, gonads and genitals that do not fit typical binary notions of male or female beings.[1] The external genitalia do not resemble that of either a boy or a girl. These disorders of sex development (DSD) are medical conditions in which the development of chromosomes, gonads, or sex anatomies vary from normal and may be incongruent with each other.[2, 3, 4, 5] Deformities are easily recognizable at birth but some may require experts to recognize them. Parents, who determine to find out why their children look more like the opposite sex that had been erroneously assigned them, discover such deformities later. It is then experts are called in for help and eventually Intersex Surgery is considered and performed to repair the ambiguous genitalia.[2,3]

The surgeons work to make the genitals look more normal while striving to reduce the likelihood of future problems.[2, 3]Timing of surgery in infancy, adolescence or adult age has been controversial, associated with issues of consent particularly in situations where parents wanted early corrective surgery for a child with DSD.[2, 3, 4, 5]There are numerous goals of surgery. This Juvenile wanted to be a boy.

Need To Make This Report.

The essence of reporting this Intersex Case is to show its reality in Nigeria; that help is available for intersex surgery at Bingham University Teaching Hospital (BHUTH), Jos, Nigeria. This article will however, not dwell on gender assignment or sex rearing.

Case Description

Demography/History.

Ambiguous genitalia occur 1 in 18, 000 to a total of about 1 in 4,500 births in the UK with congenital adrenal hyperplasia (CAH) being the most common cause[6]; hypospadias and undescended testes make additional 1 in 300.[7] As reported CAH appears more commonly with European Jewish, Hispanic, Slavic and Italian descent.[6,8] We are not aware of a National Diary of Intersex and its surgery in Nigeria.

A 15-year old who had at birth having a small penis and a partially descended right testis was referred to BHUTH for the purpose of intersex surgery on the 29 September 2015. There was external testicular anlage only on the right groin. Parents did not seek specialist/medical counsel but chose to gender child as male.

He could not understand why he was also developing female secondary sexual characteristics. It was

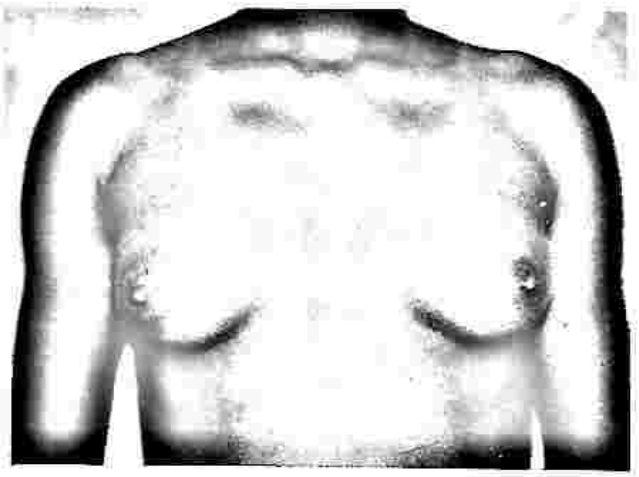

Figure 1 = Pre-Mastectomy

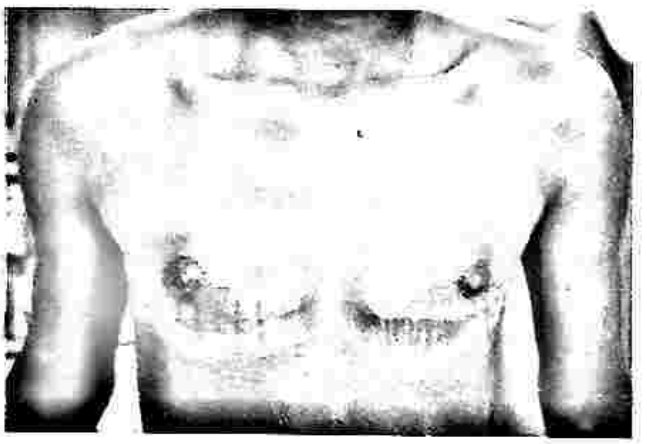

Figure1 = Post-Mastectomy

Figure 2: Absence of testis in left hypotrophic scrotum

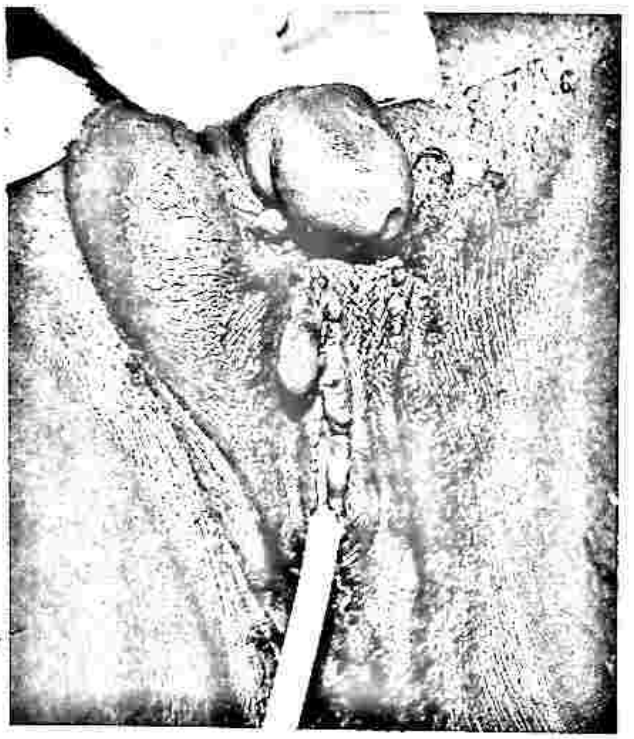

Figure 3: Showing urethral opening within the perineum as would be found in a woman. Note the absence of testis on the left.

appalling that what appeared just as minor breast enlargement had grown into full breasts bilaterally. Whereas the "penis" was gradually increasing in size there however was no proper scrotum nor palpable testis in the scrotal vestige on the right groin. There was no semblance of scrotum nor testis on the left lateral inguinal region.

Very presently, he began developing breasts, and seeing "menstrual bleeding". He bled lightly for 6 days each month since age 14. He urinated standing.

He denied being beleaguered by others; nevertheless, the community became aware since 1 year. He had some internal psychological crises and felt as in-between. He attained occasional erections. The family history did not suggest a genetically transmitted DSD such as ambiguous genitalia, infertility, primary amenorrhoea, late puberty or consanguinity.

The parents assigned him the sex of boy. He felt like a boy and wanted to be a boy. Detailed history taking revealed that urine actually came out at the perineum and not through the phallus. Juvenile and parents asserted that there had been monthly "menstrual bleeding". Parents delayed the masculinizing surgery because of indigence until they found help from Foreign Missionaries who sent patient to BHUTH for further assessment and surgery.

Table 1.Laboratory workup.

Hormone	RESULTS	Reference Range
BUN	1.8 mMol/L	Low
TESTOSTERONE	3.8 ng/ml	Adult 2.5 – 10ng/ml = low end for adolescent male.
T3	122	Normal Values
T4	2.4	
TSH	2.4nmol/L	
PROGESTERONE	0.5ng/ml	Adult 0.13 – 1.22ng/ml normal
LUTEINISING HORMONE (LH)	1.4 mIU/ml	Normal
FSH	2.4 mIU/ml	Normal
PROLACTIN	5.1 ng/ml	Normal
OESTRADIOL	80pg/ml/ml	Abnormal, raised for males
RVS (RETROVIRAL SCREENING)	Not reactive	

Physical Examination.
Examination showed a "tall boy" with the lache stage III, a micropenis with a short chordee and lack of a proper scrotum. A mass of about 3 cm was palpated in the right scrotum. He showed other features of having achieved puberty.

Results of investigation.
Imaging:
Ultrasonography of the pelvis showed a unicornuate uterus with a Fallopian tube on the left.
Laboratory:

Thyroid screening Test (TST), Prolactin (PRL), Estrogen (EST), Progesterone (PROG) were analysed. Urea and electrolyte were investigated to rule out functional limitations of the renal system; urinalysis (UA) and retroviral status (RVS) were checked.

The results of the preoperative laboratory work-up were normal for urinalysis, TSH, T3 and T4, LH, FSH, Prolactin, Progesterone and Estradiol. Testosterone of 3.8ng/ml was at the low end of normal for adolescent males. The retroviral screen was non-reactive. Chemistry values were all within normal ranges and the CO2 at 22mmol/L was normal.

Pathologist report.
Macroscopically, specimen consisted of uterus with ovary and fallopian tube measuring 12x10x4cm and weighing 50g and when cut open revealed the cervix and uterine canal. Histological sections showed a fibroblastic stroma within which few ovarian follicles as well as corpora albicans were seen. Elsewhere were sheets of seminiferous tubules lined by few Sertoli cells and the least matured germ cells: the spermatogonia. No atypia was seen.

Diagnosis
The overall picture was consistent with diagnosis of ovotestis of the left gonadal specimen.
True hermaphroditism now defined as ovotestis, was confirmed histopathologically and we proved that there was no malignancy by biopsy in line with literature.[7,8]

Intervention and Outcome

Masculinizing Surgery

An intersex surgery - masculinizing surgical procedure- was planned as the patient wanted and consented to retain his male gender.

Informed consent was obtained from the Juvenile and parents. A combined general surgical and gynaecological masculinizing procedure was done, being the first sitting. This consisted of a hysterectomy, left gonadectomy, frenulo-plasty inclusive of a biopsy of the gonads to confirm ovotestis of the right ovary and to rule out malignant degeneration of the left ovary. Perineal reconstruction, scrotum-plasty, colpocleisis, perineoplasty – closing up of the vaginal opening, and orchiopexy for the right testis and a urethroplasty were deferred.

What appeared to be a small caliber vaginal opening was found to be a perineal urethral opening and what appeared to be rudimentary labial tissue lateral to the opening was actually open urethral tissue. The actual urethral opening was on the perineum only. Not enough urethral length was found for urethral pull-through of the entire phallus.

No vagina was found except for a unicornuate uterus inserting into the urethra.

There was a small testicle Prada 4 size in the right hemi-scrotum and none on the vestigial left hemi-scrotum. We placed a Foley catheter through the perineal urethral opening for urinary drainage.

Aurethroplasty was deferred to next stage of surgery since it is bedeviled by many post-operative complications.[9] The chordee was surgically removed having paid close attention to haemostasis.

We removed the round ligaments and the ovarian vessels between vicryl® sutures. All visible gynaecological structures, the left positioned unicornuate uterus, left Fallopian tube and left ovary were cleared after proper dissection and sent for histopathology.[8]The abdominal cavity was closed in layers, with nylon to skin after ensuring complete haemostasis.

The two nipples were harvested before mastectomy of the two breasts. Exact excision and dissections along lines of careful elliptical markings completed the procedure. Sterivac® suction drainages were placed in the subcutaneous layer to drain off serous collections and enhance wound healing. The nipples were inserted bilaterally at the pre-determined positions on the anterior aspects of the chest along the mid-clavicula lines at about the fourth intercostal space just below the fourth rib.

Post-operative ward management was uneventful. Length of hospital stay was 10 days, after primary wound healing. He did not manifest a disturbance of his quality of life during his stay in the hospital.

i. State of Patient when last seen

He was last seen on date of discharge and has not reported in the out-patient clinic till date.

DISCUSSION

The essence of reporting this Intersex Case is to show that help is available at Bingham University Teaching Hospital, Jos, Nigeria. Karyotyping to determine sex chromosome is feasible in Nigeria and abdominal USS to detect presence of uterus, cervix and vagina abound in Jos, Nigeria.

Gender assignment was decided by parents without proper specialists' consultation.[12]The choice of surgery appears a good one for the boy, because, he had a phallus, secondly had been experiencing penile erections, in other words he was functioning as the boy he so much desired to remain and while on admission was apparently happy with the decision for masculinizing surgery. Moreover, we saw a boy radiating with joy post-surgery.

True hermaphroditism, now known as ovotesticular disorder of sex development (DSD), a medical term for an intersex condition in which an individual is born with ovarian and testicular tissues in different combinations was proved in this case. Many questioned if our initial diagnosis of true hermaphroditism was correct but we were proved right by the histo-pathologic report.[10, 11]For what it is worth, we are pleasantly surprised that the pathology team found spermatogoniae in the ovotestis; this would be atypical to our understanding.[13]

It is reputed that some true hermaphrodites adjusted to female phenotypes are able to gender children, but none who are made phenotypically male will be able to produce children.[13]In this present age of artificial fertilization by intracytoplasmic sperm injection (ICSI) which since 1992 has led to pregnancies in the presence of severe oligospermia, he could be fortunate. [14]This was explained fully to the boy by the surgical team before and after surgery.

CONCLUSION

Early diagnosis of intersex is essential to establish the correct sex for child rearing, to avoid wrong decisions and prevent mistakes from being made.[15]

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- DUALITY OF INTEREST - Nil

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