INTRODUCTION

CASE HISTORY

A 45 years old Labourer father of 5 children from Mandi Bahauddin was admitted through out-patient department in February, 2010. Presented with painful swelling in lower abdomen for last 15 days. Swelling rapidly increased in size. It started in the RIF then involved the hypogastrium and became painful. Pain was constant dull ache, relieved somewhat after micturition. There was H/O Anorexia & Significant weight loss over 6 months. There was no H/O Bowel complaints or urinary complaints.

There was no past history of prolonged illness requiring hospitalization or investigations.

In Personal history. He was Smoker for last 20 years, smoked 1 pack per day. He was married with 5 children (4 sons 1 daughter). Youngest is 3 years old. He belonged to Lower Middle class. Family history was Insignificant.

On examination, he was Middle aged man sitting comfortably, well oriented with Pulse of 82/min, BP: 110/70 mm of Hg, Temp: 98fh & R/R: 18/min.

There was no pallor, Jaundice, Clubbing, Cyanosis or Pedal oedema. JVP was normal. No Lymph nodes or Thyroid swelling were palpable.

Systemic examination was Unremarkable

Oro-dental hygiene was good.

Abdomen

Umbilicus was central & inverted. There was visible bulge in Right Iliac Fossa. A palpable mass was present in hypogastrium and RIF measuring 10 x 9 cm on ultrasound abdomen and pelvis. Large fibrous mass in right side of pelvis 10x9 cm pushing urinary bladder up. It was tender, hard, partly mobile, upper limit was reachable but lower limit was not. Liver and spleen were not palpable. On digital rectal examination, a hard extra-rectal mass was palpable anteriorly, about 7 cm from anal verge. Scrotum was not developed. Testes were not palpable in scrotum or groin bilaterally. His phallus was normally developed with external urinary meatus at the tip. There was normal distribution of the pubic hair and other secondary sexual characters were present. Respiratory and Cardiovascular system were unremarkable.

PROVISIONAL DIAGNOSIS of Bilateral undescended testes with Lower Abdominal Mass (? Testicular Tumor) was made.

INVESTIGATIONS

FBC: Hb 13.4 g/dl, TLC 9100 /cumm, Platelets 230000 /cumm ESR: 29, Neutrophils 77%, Lymphocytes 12%, Monocytes 6%, LFTs: Bilirubin : 0.8 mg/dl, ALT: 19,
PT: 15/13, APTT: 31/30, BSR: 135 mg/dl, 
RFTs: Urea: 33 mg/dl. 
Alpha Feto Protein: 
  4.19 ng/ml (Normal value upto 8.4) 
B-HCG: 15.6 mIU/ml (Normal value <5), 
S. LDH: 925 U/L (Normal value < 480) 

**Semen Analysis** 
Azoospermia 
Ultrasound abdomen and pelvis: Large fibrous mass in right side of pelvis 10x9 cm pushing urinary bladder up.

**MRI pelvis** 
20 cm sized mass in the pelvis extending into the lower abdomen with well defined borders. Uterine cavity rudimentary vagina can be appreciated.

**Conclusion** 
Tumour arising from right undescended testis. Lymph nodes behind the uterus forming a mass, pushing the urinary bladder forward.

Diagnosis of testicular tumour with bilateral cryptorchidism was made.
OPERATIVE FINDINGS

A Large right testicular intra abdominal mass. Uterus with fallopian tubes and blind ended vagina was present. Left intra abdominal testis was normal looking. There was a Large lymph node mass in the hypogastrium behind the uterus, extending to blood vessels and right ureter. No ascites or liver mets were present.

OPERATIVE PROCEDURE

Right testicular tumour mass was resected. Uterus with fallopian tubes, left testis and blind ended vagina were resected en-block with lymph nodes except a rim
of tissue which was engulfing iliac blood vessels.

**Post-Operative Course**

Post-Operative Course was uneventful. Patient was referred to NORI Hospital for further treatment (Chemo /Radiotherapy).

The Blood Group of the patient and all his children was done which was “O” positive.

DNA testing and karyotyping was not done because of psychosocial issues.

**HISTOPATHOLOGY**

2 specimens were sent for histopathology. Specimen 1 was tumour & specimen 2 was of left testis and uterus. Specimen 1: was reported as classical seminoma. Specimen 2: Left testis showing atrophy. Nodes showing metastatic seminoma with a triangular cavity in front of this mass.

**DISCUSSION**

KEY ISSUES: were

1) Presence of Uterus with fallopian tubes and blind ended vagina.
2) Seminoma Testis in Right Undescended Testis.
3) Fertility issues in Patients with Bilateral Cryptorchidism.

Sexual differentiation is on the basis of Psychological Sex and Organic sex

Organic sex depends on Chromosomal Sex, Gonadal sex & Phenotypic sex

**Male**

XY, Gonad-Testes, Mullerian regression factor. Testosterone is responsible for development of Wolffian duct, while DHT for development of External genitalia

**Female**

XX, female because of presence of Ovary. Absence of Testes, No testosterone to develop Wolffian duct. Absent Mullerian regression factor results in persistence of Mullerian duct structure.

**Intersex**

Disparity between chromosomal, gonadal and phenotypic sex.

Summary in our case, Phenotypic male with well developed phallus and other secondary sexual character. But gonads were bilaterally impalpable. Intra- abdominal testes. Lt testis normal & Classical seminoma in right testis (Gonadal sex-male).
Presence of female internal organs. What is the possibility?

In Male Pseudohermaphrodite, Gonad could both be testes/ovary. Chromosomal and gonadal sex-male but external genitalia-female. Impaired synthesis, secretion, conversion or action of testosterone. Impaired synthesis, secretion, or failure to respond to mullerian regression factor. Female genital duct in otherwise normal male. This is one possibility.

**Female Pseudohermaphrodite**
Chromosomal and gonadal sex-female but external genitalia-male. Classical example congenial adrenal hyperplasia. Our case does not fit into this category.

**Defect in chromosomal / gonadal sex**
Mixed gonadal dysgenesis is common cause of intersex. Phenotype female or male.

Testes are located intra-abdominally. Uterus, vagina and at least one fallopian tube invariably present. High incidence of tumour development in gonad. Karyotype 45x/46xy but always chromatin negative. This is another possibility.

**Cryptorchidism**
Cryptorchidism is the most common genital problem encountered in pediatrics. Cryptorchidism literally means hidden or obscure testis and generally refers to an undescended or maldescended testis. Overall, 3% of full-term male newborns have cryptorchidism, decreasing to 1% in male infants aged 6 months to 1 year. The prevalence of cryptorchidism is 30% in premature male neonates. Factors that Predispose to cryptorchidism include prematurity, low birth weight, small size for gestational age, twinning, and maternal exposure to estrogen during the first trimester. A recent study found that almost 23% of index patients with undescended testes had a positive family history of cryptorchidism, as opposed to 7.5% in control families.


Paternity rates among formerly cryptorchid and control men are shown in Table.

Giltay et al. describe an unusual case of true hermaphroditism—that is, the presence of both ovaries and testes in a single individual. The boy’s somatic tissues are likewise a mixture of karyotypically normal male and normal female cells.

Zayed F et al reported A male phenotype (XY) hermaphrodite treated for seminoma, fathered a healthy child by IVF–ICSI technique. It is generally known that almost all hermaphrodites are infertile, however, German et al. observed spermatogenesis in an hermaphrodite. Furthermore, in abstract, both Inatomi et al. and Manba et al. have reported the delivery of infants fathered by a true hermaphrodite under natural conditions in Japan.

A 38 years old patient was seen in 2001 by urologist for infertility and impalpable in the right side of the pelvis. He had laparotomy, and right sided orchidectomy of a right testicular seminoma which was excised in addition to what appeared as a uterus and tubes in 2001. This patient getting treated and cured from seminoma, the couple had in vitro fertilization (IVF) and intracytoplasmic sperm injection (ICSI) using frozen testicular sperm and had a healthy baby.

A successful pregnancy outcome using frozen testicular sperm from a chimeric infertile male with a 46, XX/46, XY karyotype was reported in 2005 by Sugawara et al. A successful second delivery outcome using refrozen thawed testicular sperm from an infertile male true hermaphrodite with a 46, XX/46,
XY karyotype: was reported Sugawara et al in 2012.

Regarding incidence of Testicular Tumors in Patients with Cryptorchidism, Campbell-Walsh Urology, 9th edition states, “It is a well established fact that children born with undescended testes are at increased risk for malignancy. The recurrence rate is approximately 40 times greater.” In Adult and Pediatric Urology it is stated that “The combined risk for all cryptorchid males, irrespective of the location of the testes, has been calculated at 20 to 46 times greater than for patients with normally located testes.” In Pediatric Urology, WB Saunders Co, Philadelphia (2001) chapt 46. it is stated that “Individuals born with an undescended testis have approximately a 40-fold incidence of testicular malignancy over those born with scrotal testes.” Gehring et al reported a similar pattern with a 46%, 21% and 32% rate of seminoma, embryonal tumors and teratocarcinoma, respectively, in testicles that underwent malignant degeneration following orchiopexy. These values compare with an 89% predominance of seminoma in patients who had uncorrected cryptorchidism and later had testicular cancer.

Batata et al reported that when comparing pathological types between cases with and without orchiopexy or hormonal treatment, a predominance of seminoma was observed in uncorrected cases (30 of 42 or 71.4%). However, in corrected cases the distribution of tumor type was remarkably similar to that in the series by Johnson et al, including seminoma in 29% of cases, embryonal tumors in 33% and teratocarcinoma in 35%.

Conclusions (Learning Points)

- Never forget to examine genitalia during abdominal examination.
- Bilateral cryptorchidism does not mean infertility.
- Even hermaphrodites can become parents.
- Seminoma is the most common tumor in undescended testis.

REFERENCES


10. Campbell-Walsh Urology, 9th edition states, “Children born with undescended testes are at increased risk for malignancy”.

11. Adult and Pediatric Urology “The combined risk for all cryptorchid males, irrespective of the location of the testes, has been calculated at 20 to 46 times greater than for patients with normally located testes.”


