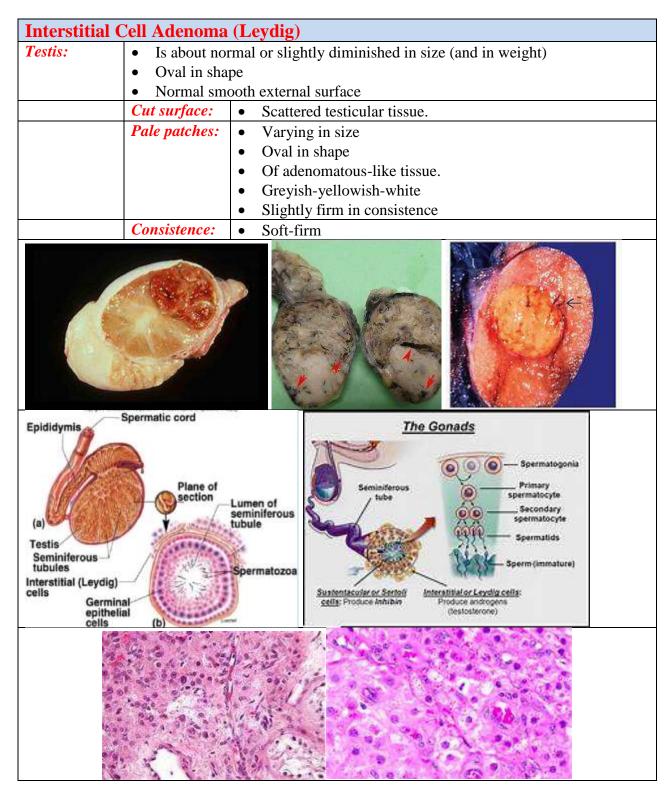
409 XX. Diseases of the Male Genital System



N.B.:

- Histologically, the seminiferous tubules were less mature than would be expected from the age of the patient and, they were made up of closely-packed tubules containing a relatively larger number of Sertoli cells compared with the spermatogons.
- Leydig cells were hyperplasic, prominent and revealed some atypical forms.
- This testis belonged to a person (16 years old) who had well-developed breasts and areolae, feminine external genitalia but with vulval hypoplasia, a testicular feeling of the left gonad (found at the left inguinal region near the external ring) and a testicle-like gonad in the right inguinal canal.
- This person, presumed to be (and behaving as) a girl (with high-pitched voice and a feminine distribution of axillary and pubic hair), complained of primary amenorrhea i.e. has never menstruated.
- This case is an example of the testicular feminizing syndrome" which is a rare form of **male pseudo hermaphroditism** characterized by the foetus starting development as male then shows varying degrees of feminization i.e. a genetic male who resembles a female.
- The general features:
- Well-developed breasts, large areola and nipple, female habitus, no menstruation, absence of uterus, short vagina, large clitoris and bilateral immature inguinal or intra-abdominal testes.

| Undescended Testis (cryptorchism) | |
|-----------------------------------|--------------------------|
| Testis: | Small in size |
| | Keeps its shape |
| | Is atrophied |
| | Shows increased fibrosis |

N.B.

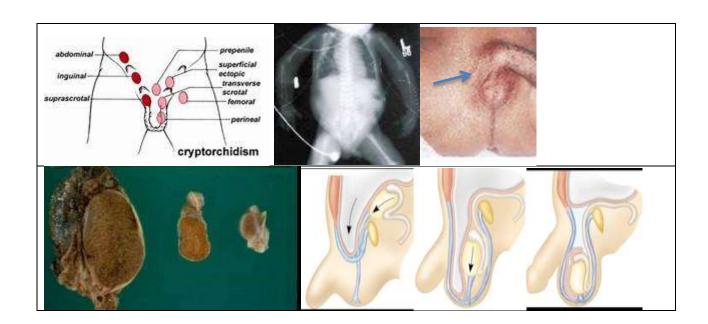
- Common sites of undescended testes
 - 1. Near the kidney.
 - 2. Internal abdominal ring.
 - 3. Inguinal canal (common).
 - 4. External abdominal ring.

Common causes:

- 1. Adhesions around the testicle.
- 2. Narrow external ring.
- 3. Deficient gubernaculum.
- 4. Hormonal deficiency.
- 5. Disturbance in anterior pituitary.

Effects:

- 1. Torsion.
- 2. Traumatic orchitis.
- 3. Atrophy and fibrosis \rightarrow sterility.
- 4. More liability to malignant change.



Noonan Syndrome

- Autosomal dominant disorder (variable penetrance)
- Normal karyotype
- Phenotype is like that of Turners syndrome
 - Low set ears
 - Right sided congenital heart defects (left-sided in Turners)
 - Epicanthic folds
 - Short stature
 - Webbed neck
 - Cryptorchism (50% of males)
 - Primary hypogonadism
- Can affect either sex



