

Case-Based Diagnosis Training

Patient:

Gender: *Female*

Age: *15 days*

Clinical history and working diagnosis on the referral:

A 15 day old girl presented with raised inflammatory markers (white cell count $19.6 \times 10^9/L$, CRP 20mg/L) and raised bilirubin (28umol/L). She had blood cultures and cerebrospinal fluid (CSF) tap to assess for meningitis, and both were found to be negative for organism growth. On clinical examination, she was found to have an incidental palpable lump in the right groin, and was referred for ultrasound to assess for the cause. Ultrasound (US) demonstrated a non-reducible 1.8 x 0.9cm right inguinal hernia (picture 1) with a 0.6cm neck. The contents showed soft tissue containing anechoic follicles (picture 2, picture 3), and the neck of the hernia showed marked vascularity (picture 4). Appearances were consistent with the right ovary within a right inguinal hernia sac.

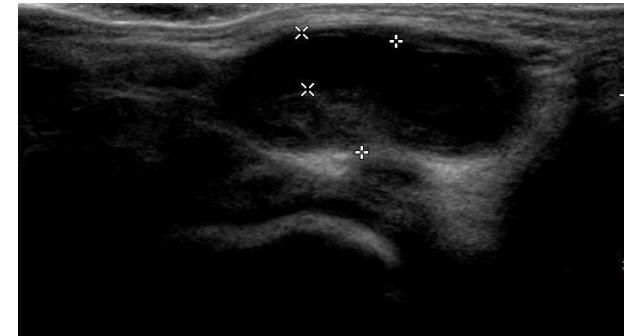
Normal variant:

The ovaries descend during embryonic development from the upper lumbar region at week 6 and relocate to the pelvis. Gonadal descent can arrest at any stage of this process, resulting in an aberrantly placed but otherwise normal ovary. This case demonstrates a right ovary which has descended further than normal into a patent canal of Nuck, a structure which is homologous to the processus vaginalis in males. The canal of Nuck extends anteriorly from the round ligament of the uterus through the inguinal canal and into the labia majora. This sac normally obliterates by 1 year of age, but if remains patent forms the Nuck diverticulum into which the ovary (and other pelvic structures) can descend within the female inguinal canal, resulting in indirect inguinal herniation of the ovary (picture 6) and other pelvic structures.

Submitted by:

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Please add pictures (radiograph, ultrasound, CT or MR images) and schematic drawing of the developmental process *if applicable* by clicking on the symbols within the boxes below:



Picture 1: US right groin demonstrating anechoic sac containing hyperechoic soft tissue



Picture 2: US right groin demonstrating an indirect inguinal hernia containing multifollicular right ovary

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Additional information

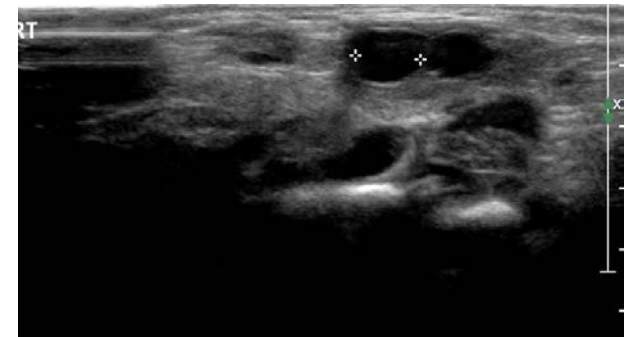
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Underlying step in embryological development:

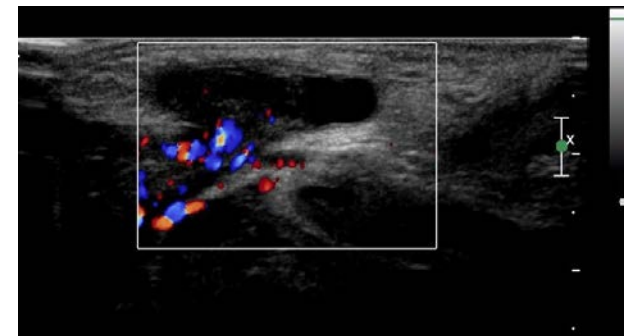
At week 6-8 of embryological development (picture 7), the gonads are undifferentiated. In the absence of testosterone, the female mesonephros atrophies and the upper gubernaculum becomes the ovarian suspensory ligament, connecting the upper pole of the ovary to the upper rear body wall. The lower gubernaculum attaches to the inferior pole of the ovary to become the ovarian ligament, and further down becomes the round ligament of uterus, which extends into the inguinal canal terminating in the labia majora (picture 8). Continuation of ovarian descent in combination with a patent canal of Nuck allows for herniation of the pelvic organs, including the ovary, uterus and fallopian tubes, through the deep inguinal ring into the inguinal canal and labia majora (picture 6).

Potential differential diagnostic entities:

- Inguinal hernia
- Hydrocoele of canal of Nuck
- Endometriosis (adults)
- Inguinal lymphadenopathy



Picture 3: Anechoic <5mm foci within the hernial sac, consistent with ovarian follicles

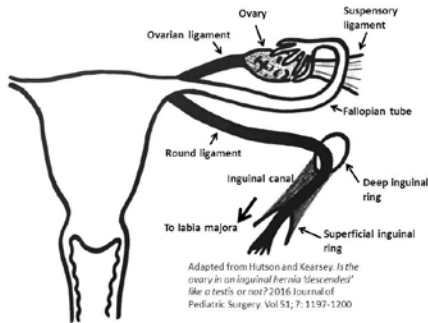


Picture 4: Doppler USS with ovarian vessels seen at the neck of the hernia

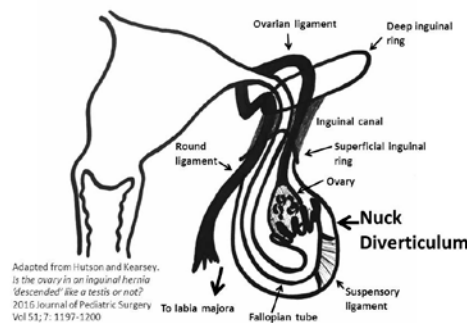
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Additional pictures

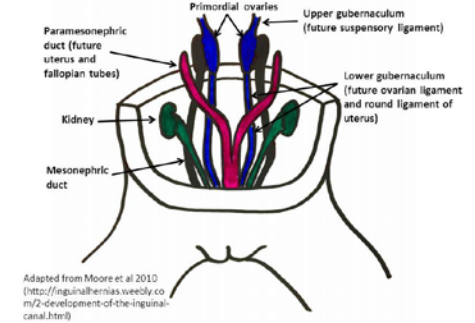
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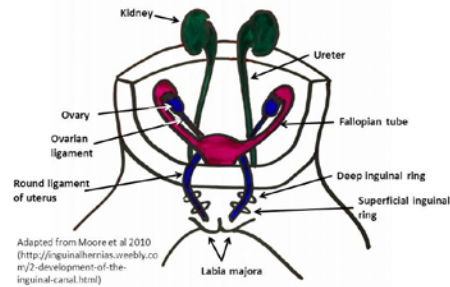
Picture 5: Normal female reproductive anatomy and inguinal canal



Picture 6: Persistent canal of Nuck containing left ovary and fallopian tube



Picture 7: Gonadal position week 7 of embryological development



Picture 8: By 7 months, the ovaries are in normal anatomical position within the pelvis.

References

- Ho Choi K and Baek HJ. *Incarcerated ovarian herniation of the canal of Nuck in a female infant: Ultrasonographic findings and review of literature.* 2016 Aug; 9: 38–40.
- Hutson JM and Kearsley I. *Is the ovary in an inguinal hernia 'descended' like a testis or not?* 2016 Journal of Pediatric Surgery, Vol 51; 7: 1197-1200
- Ibrahim H, Khaled M, Gates T et al. *Irreducible Indirect Inguinal Hernia in Premature Infant with Ectopic Uterus and Bilateral Adnexa.* J La State Med Soc. 2016 Nov-Dec;168(6):194-195.
- Ming YC, Luo CC, Chao HC et al. *Inguinal hernia containing uterus and uterine adnexa in female infants: report of two cases.* Pediatr Neonatol. 2011 Apr;52(2):103-5
- Yao L, Mou Y and Wang H-X. *Sonographic diagnosis of an ovary-containing inguinal hernia with the formation of a corpus luteum in an adult female.* Ultrasound in Obstetrics and Gynaecology. 2009; 34: 358–360
- Adapted from *Human embryology: organogenesis 21.3 Differentiation of the gonads.* Last accessed 10/01/18 <http://www.embryology.ch/anglais/ugenital/diffmorpho05.html>
- Adapted from *Inguinal hernias: Development of the inguinal canal.* Last accessed 10/01/18 <http://inguinalhernias.weebly.com/2-development-of-the-inguinal-canal.html>