QUIZ 1 (2020-21)

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Answers

Q1-3. Multilocular cystic renal cell neoplasm of low malignant potential; Q4-5. Placental site nodule

1 (3)

2 (1)

3 (4)

4 (2)

5 (4)

6 (3)

1. The following is characteristic of this entity
    a. Vascular invasion (Not seen)
    b. Prominent eosinophilic nucleoli with perinucleolar halo (seen in Hereditary leiomyomatosis associated RCC)
    c. Clusters of clear cells with fibrous septa (Characteristic finding)
    d. Expansile nodules of neoplastic clear cells (not a feature. If present, the diagnosis would be Clear cell RCC)
2. Which of the following is associated with this entity?
    a. 3p deletion (Multilocular cystic renal neoplasm of low malignant potential and clear cell RCC)
    b. Germline mutation of the fumarate hydratase gene (Hereditary leiomyomatosis associated RCC)
    c. Xp11 translocation (Translocation associated RCC)
    d. Mutation in the folliculin gene (Birt Hogge syndrome-associated with Chromophobe RCC)

4. The condition shown in the image is associated with
    a. Squamous dysplasia (SCC)
    b. Intrauterine pregnancy or abortion
    c. M spike in serum and urine protein electrophoresis and immunoelectrophoresis (amyloidosis)
    d. Loss of e-cadherin expression, with associated mass lesion of breast (Lobular carcinoma of breast)

5. Which statement applies to this condition?

    a. The serum hCG levels would be markedly elevated (no, seen in choriocarcinoma)

b. Associated with cerebral cortical tubers, skin lesions, cardiac rhabdomyomas, lymphangioleiomyomatosis, and clear cell renal cell carcinoma (tuberous sclerosis, associated with PECOMAs of uterus)
c. HPV 16 and 17 association (SCC)
d. Present as yellow white nodules, single or multiple, 1-14 mm in size, in endometrium, cervix or myometrium (placental site nodule/plaque)

Multilocular cystic renal neoplasm of low malignant potential

- Accounts for less than 1% of all renal tumors
- Affects middle aged adults
- Male to female ratio of 1.2:1 to 2.1:1
- 90% discovered incidentally during imaging for other reasons
- Macroscopically – tumor consists exclusively of variably sized cysts separated by thin septa and filled with clear, serous or gelatinous fluid
    - Solid mural masses are incompatible with this diagnosis
- Microscopically
    - Cysts are lined by single layer of tumor cells, with abundant clear cytoplasm, and small nuclei without nucleoli (usually ISUP grade 1 or 2 nuclei)
    - Important diagnostic feature: presence of clusters of clear cells with fibrous septa, without expansile growth
    - Necrosis, vascular invasion and sarcomatous transformation are incompatible with this diagnosis
- IHC
    - PAX8 and CA9, similar staining pattern like clear cell RCC
- Genetic profile
    - *VHL* mutations in 25% of these tumors
    - Chromosome 3p deletions in 74% of these tumors
- Prognosis: excellent

Placental site nodule

- Incidental finding on curettage, biopsy, or hysterectomy
    - During evaluation for uterine bleeding, infertility, or hysterectomy for other diagnoses
- Serum HCG not elevated
- 50% located in cervix, 50% in endometrium or superficial myometrium
- Benign, no treatment necessary
- MACROSCOPICALLY
    - Described on hysteroscopy as small, white-red or yellow-white nodule with hemorrhage and necrosis, rarely as polypoid
    - Rarely identified grossly as 1- to 14- mm tan, firm nodules or plaques, may be single or multiple
- MICROSCOPICALLY

    - Round to oval with lobulated margins, central hyalinized core
    - Rare decidualized stromal cells and chronic inflammation at periphery
    - Trophoblast cells dispersed as single cells, clusters, or cords with eosinophilic, hyalinized extracellular matrix
    - Minimal pleomorphism
    - Minimal mitotic activity, Ki-67 < 5%
- IHC
    - hPL: Focal staining
    - PLAP: Diffuse staining
    - Ki-67: Trophoblast nuclear staining < 5%
    - p-63: Positive nuclear staining
    - Pan-CK: marks trophoblastic cells