A 16-year-old girl presented with a swelling in the medial aspect of her knee of 1-month duration. During clinical examination, a large soft-tissue mass was identified over the medial aspect of her left knee.

Computed tomogram (CT) showed a well-defined hypodense mass in the medial aspect of her right distal thigh, measuring 7 cm × 5.1 cm in maximum axial dimensions, displacing the vastus intermedius and extending inferiorly into the patellofemoral compartment of the knee joint. She underwent a biopsy, followed by complete excision.

Gross appearance of the resected specimen and microscopic images from the resected tumor have been displayed [Figures 1 and 2].

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Questions

Q1. What is the diagnosis?
Q2. What other name is assigned to this tumor after the name of the pathologist who described it for the first time?
Q3. Which specific immunohistochemical marker for this tumor is displayed in figure 3?
Q4: This tumor shares a morphological continuum with which other tumor(s).

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There are no conflicts of interest.

Answer of Catechism (Quiz 5)

(1) Diagnosis: clear cell carcinoma of ovary
(2) Likely specific immunohistochemical marker: Napsin A
(3) Other sensitive and specific marker: Hepatocyte nuclear factor 1 beta and IMP3
(4) Which clinical history should be checked: Endometriosis.

DISCUSSION

Microscopic examination of sections revealed a tumor composed of solid-cystic components, including cells with vacuolated cytoplasm and prominent nuclei and nucleoli, arranged in solid and papillary growth patterns, including 'hobnail-like' arrangement of tumor cells around hyalinized stroma (Figure 1).

By immunohistochemistry, tumor cells were diffusely positive for cytokeratin (CK)7 and Napsin A (granular cytoplasmic staining) [Figure 2], focally positive for glypican 3, while negative for WT1 and ER. In addition, tumor cells showed focal immunostaining for WT1 (Wild type).

Clear cell carcinoma of the ovary is one of the relatively uncommon subtypes of an epithelial ovarian cancer (EOC). In view of its overlapping histopathological features with other ovarian tumors, such as a yolk sac tumor and various other EOCs, it can be misdiagnosed, as occurred in this case, which was diagnosed as a high grade serous adenocarcinoma (HGSC), elsewhere.[1,2]

Careful assessment of histopathological features, such as vacuolated cells with prominent nuclei arranged in a ‘hobnail-like’ arrangement around hyalinised papillary cores, as noted in the present case, are useful morphological clues. Furthermore the diagnosis can be confirmed by positive expression of certain sensitive and specific immunohistochemical markers, such as Napsin A, Hepatocyte nuclear factor-1beta (HNF-1beta) and IMP3 and negative expression of WT1.[2,4] A clinical history of co-existing endometriosis should be checked in such cases. An exact diagnosis has significant treatment implications, as CCC ovary is relatively chemo refractory, in contrast to a HGSC.[5] After completing two cycles of neoadjuvant chemotherapy, there was no significant clinical response in the present case.
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There was an overwhelming response from the readers to Catechism 5. Some of the other answers besides the correct answer given by various readers are mentioned below-

1. Mesothelioma of ovary
2. CK 5/6
3. Calretinin, WT-1, D2-40
4. History of asbestos exposure, previous H/O pleural effusion, previous H/O ascites

The winners of Quiz 5 are-

(a) Dr. Gowripriya G, Consultant Histopathologist, Dr Rela Institute and Medical Centre, Chennai
(b) Dr. Surbhi Satvik Bansal, Assistant Professor, Cancer hospital and research institute, Gwalior (MP)

Congratulations to the Winners! Dr. Ranjan Agrawal, Editor-in-Chief