Sarcomatoid and Rhabdoid Renal Cell Carcinoma Pathology

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Definition

Definition of sarcomatoid renal cell carcinoma

Sarcomatoid renal cell carcinoma (SRCC) is currently defined in the 2004 World Health Organization (WHO) classification of renal tumors as any histologic type of renal cell carcinoma (RCC) containing foci of high-grade malignant spindle cells.[1]

Many studies have defined a tumor as SRCC if even a small amount of sarcomatoid differentiation is present,[2, 3, 4, 5] whereas other studies have excluded tumors with a sarcomatoid component of less than 20% of the tumor volume[6] or less than one microscopic low-power (40x) field in size.[7]

However, some evidence exists of increased risk associated with sarcomatoid components comprising 5-10% of total tumor volume,[5, 7] indicating that even small amounts of sarcomatoid differentiation may be clinically relevant and should be included in the pathology report.

Focal spindling due to noncohesion of tumor cells is not considered to represent sarcomatoid differentiation.[7] Tumors with pure sarcomatoid morphology due to overgrowth of the RCC components are classified as SRCC if evidence of epithelial differentiation can be demonstrated through immunohistochemical or ultrastructural analysis.

In cases in which the histologic subtype of the RCC component is not recognizable, the tumor is designated as unclassified RCC with sarcomatoid differentiation.[1]

Definition of rhabdoid renal cell carcinoma

Adult rhabdoid renal cell carcinoma (RRCC) is currently recognized in the literature as any histologic type of RCC containing foci of high-grade malignant cells with rhabdoid morphology, characterized by large eccentric vesicular nuclei, prominent nucleoli, globular eosinophilic paranuclear inclusion bodies, and abundant eosinophilic cytoplasm.[8, 9, 10, 11]

The proportion of rhabdoid components in published series of RRCC ranges from 1-90% of the total tumor volume[8, 10, 11] ; however, occasional tumors composed entirely of rhabdoid cells have been described in which no RCC component is detected despite extensive sampling.[9, 12]

RR CC was named for its morphologic resemblance to pediatric malignant rhabdoid tumor (MRT) of the kidney, which is a highly aggressive tumor characterized by cells that resemble rhabdomyoblasts and by genetic alterations involving chromosome 22, particularly the *hSNF5/INI1* gene on 22q11.2.[13, 14] MRT usually arises in children younger than 3 years, and the occasional cases previously reported as adult MRT are now considered more likely to be RRCC in which rhabdoid components have overgrown the original histologic tumor type.[8]

Go to Renal Cell Carcinoma and Clear Cell Renal Cell Carcinoma for more complete information on these topics.

Contributor Information and Disclosures

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