World Health Organization (WHO) classification (2016), as a RCC with a morphology and/or growth pattern that does not readily fit into any of the established subtypes of RCC[4,5]. URCCs account for 4-6% of RCC cases with a broad age range of 21 to 91 years, male to female ratio being 1:16. High grade and stage at the time of presentation render an unfavourable and aggressive behavior of URCCs[7]. Histological heterogeneity may explain a wide spectrum of radiological findings ranging from solid with a cystic component, hyperintense on T1- and isointense on T2-weighted images, show heterogenous enhancement and contain hemorrhage[8]. Grossly, URCCs are usually large, variegated in appearance and ferociously involve the renal parenchyma. The histomorphological features do not correspond to any recognized entities. The tumors with either composites of recognized types, pure sarcomatoid morphology, mucin production, rare mixtures of epithelial and stromal elements, and/or unrecognizable cell types[9] are classified under this category of neoplasms. The cellular morphology may be low-grade or high-grade. Lymphovascular invasion and necrosis are commonly seen.

Although limited by scarce data, the unclassified RCC is associated with adverse clinicopathological features such as larger tumors, increased risk of adrenal involvement and involvement of adjacent organs, and increased risk of metastatic involvement of the regional/nonregional lymph nodes and the bones compared with clear cell RCC[10]. Karakiewicz et al. also documented the association of unclassified disease with Fuhrman grade III or IV, nodal and distant metastasis, and higher mortality than was seen in clear cell tumors[11]. Thus various independent prognostic pathological markers include tumor size, nuclear grade, pathological stage, tumor coagulative necrosis, vascular invasion and recurrence after surgery. However, Crispen et al found no statistically significant differences in cancer free survival between URCC and other RCC subtypes[12]. Radical nephrectomy with adjuvant chemotherapy such as immunotherapy may be the mode of treatment. No specific targeted therapy is yet established because of limited cases and genomic instability.

Unclassified RCC is a **rare neoplasm** with heterogeneous differentiation. The co-existence of varied histological features in same tumor must alert the pathologist to consider URCC as a differential diagnosis owing to its association with **high grade, high stage and poor survival. Extensive sampling** should be performed to identify the heterogeneity of morphological patterns. An aggressive progression and dismal prognosis warrants a review of the initial radiological investigations by treating surgeons with special emphasis on metastasis.