Chromophobe kidney cancer accounts for approximately 5% of cases of renal cell carcinoma (RCC). While the genetics of clear cell RCC has been a major focus of research, little is known about the biology of chromophobe tumors. There is ample preclinical rationale for the use of targeted therapy in clear cell tumors, and agents targeting the VHL/HIF pathway are now widely used in clinical practice. However, there is limited experience with targeted agents in non-clear cell tumors. Recently, a few case reports have emerged which report the use of mTOR inhibitors in chromophobe tumors. Here, we report our experience with targeted therapy in a patient with advanced chromophobe RCC who had a durable partial response to temsirolimus. We also include a literature review summarizing the published experience with targeted therapeutic approaches in chromophobe RCC. Additionally, the preclinical rationale for the use of mTOR inhibitors in this population based on our characterization of the hereditary form of chromophobe kidney cancer, Birt-Hogg-Dube syndrome, is discussed.

Key Word:
mTOR inhibitors, Chromophobe Kidney Cancer