RENAL MEDULLARY CARCINOMA: A SYSTEMATIC REVIEW

C. Saldana1, G. Logonadane1, J. Caldesaro2, M. Chabut Houdo1, H. Bousion1, I. El Sayed1, A. DeLaTalle1, Y. Alory2, C. Tournigand1

1Medical Oncology, CHU Henri Mondor, Créteil, FRANCE
2Pathology, CHU Henri Mondor, Créteil, FRANCE
3Urology, CHU Henri Mondor, Créteil, FRANCE

Aim: Renal medullary carcinoma (RMC) is a rare, highly aggressive renal neoplasm. In absence of prospective trials, a systematic review of RMC publications was performed in order to characterize clinical features and treatment.

Methods: We found 68 articles on RMC in PubMed from 1987 to January 2014 (key words "Carcinoma, renal cell" OR "kidney neoplasm" AND "medullary").

Results: 123/135 cases were analysed (12 missing data). Median age was 21.9y (10-69). Male/female ratio was 2:1. Afro-Americans represented 83% of pts, Caucasians, Arabs / Brazilians were 8.7% and 2.6% respectively. Cases related to sickle cell trait accounted 89% while homozygous form and hemoglobinopathy free status were 8% and 3% respectively. Common initial symptoms were hematuria (49%), flank pain (55%) and weight loss (14%). Time from symptoms to diagnosis had a mean of 4.7 m. At diagnosis, only 2% had localized tumor while 98% were metastatic: secondary lymph node involvement (62.5%), lung (48%), liver (33%), bone (32%), pleura (16%) and brain (16%). Right kidney was involved in 74% of cases. Survival data was available in 82 pts (66%). Ten pts (12%) were alive at the moment of publication. Median overall survival was 9 m (0-92). Patients with visceral metastasis (96%) had the worst mean survival (6.3m) while pts with isolated-lymph-node involvement (12%) showed better outcome (OS = 14m). No responses were detected with immunotherapy (OS = 3m). Best outcomes were displayed with chemotherapy combination regimens based on platinum derivatives, gemcitabine, paclitaxel (n = 9) or based on MVAC (n = 14), with median survivals of 10 and 15 months respectively. High-dose chemotherapy followed by autologous stem cell transplantation seems useful but not sufficiently explored (n = 1, OS = 22m). Anecdotal cases of targeted therapy included: Bortezomib (n = 2, OS= 27 m and 6 m), Thalidomide (n = 2, OS = 52 m and 2 m), Sunitinib (n = 10, OS = 2 m), Imatinib (n = 1, OS = 1m). Loss of immunoexpression and gene inactivation of SMARCBl/INI were the only molecular alterations constantly found (n = 28/28).

Conclusions: RMC, a rare subtype of renal cell carcinoma with poor outcome, mainly affects a particular population of young afro-Americans with hemoglobinopathy. Better biological understanding and molecular characterization of RMC will facilitate improving the treatment and survival of these patients.

Disclosure: All authors have declared no conflicts of interest.