



General Characteristics and Outcome of Renal Extramedullary Hematopoiesis in Patients with Myeloproliferative Neoplasms-A literature review

Shehab Mohamed^{1,2}, Mohamed Alkahateeb³, Ahmed Abdalhadi ³, Abdulqadir, Nashwan ³, Khaldoun Obeidat ³, Yahaa Imam ³ , Mohamed Yassin ²

¹ MD Anderson , Houston, Texas, USA; ² NCCCR, Doha, Qatar; ³ HMC, QATAR

CONTEXT

- Kidneys are a rare but serious site for Extramedullary hematopoiesis (EMH) with myeloproliferative neoplasms (MPNs)

OBJECTIVE

- Review cases of EMH affecting the kidneys, focusing on demographics, clinical manifestations, treatment and outcomes.

DESIGN AND SETTING

- We reviewed the literature for studies, case series and case reports that reported on renal EMH with a primary or background diagnosis of MPNs from 1990-2020

RESULTS

- Forty-three patients (79 % males) were retrieved.
- Mean age of the cohort was 65 years (range 49-87years).
- Most of the cohort was white Caucasian 91 %.
- The type of MPNs was mostly MF, seen in 35 patients (81.4%), while PV was seen in 5 patients (11.6%), ET and CML were seen in 2(4.7%) and 1 (2.3%) patient, respectively.

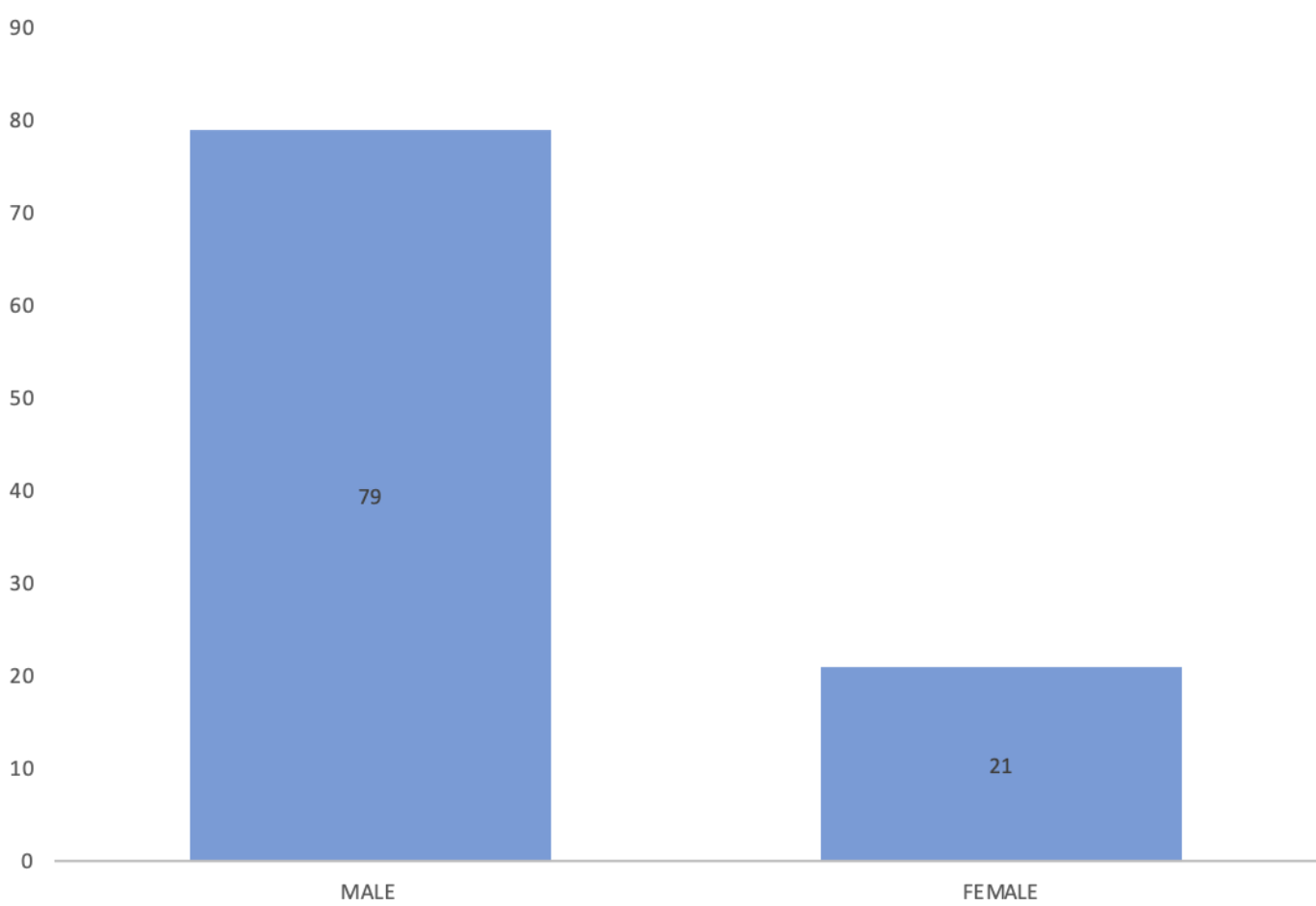


Fig. 1 Gender Distribution

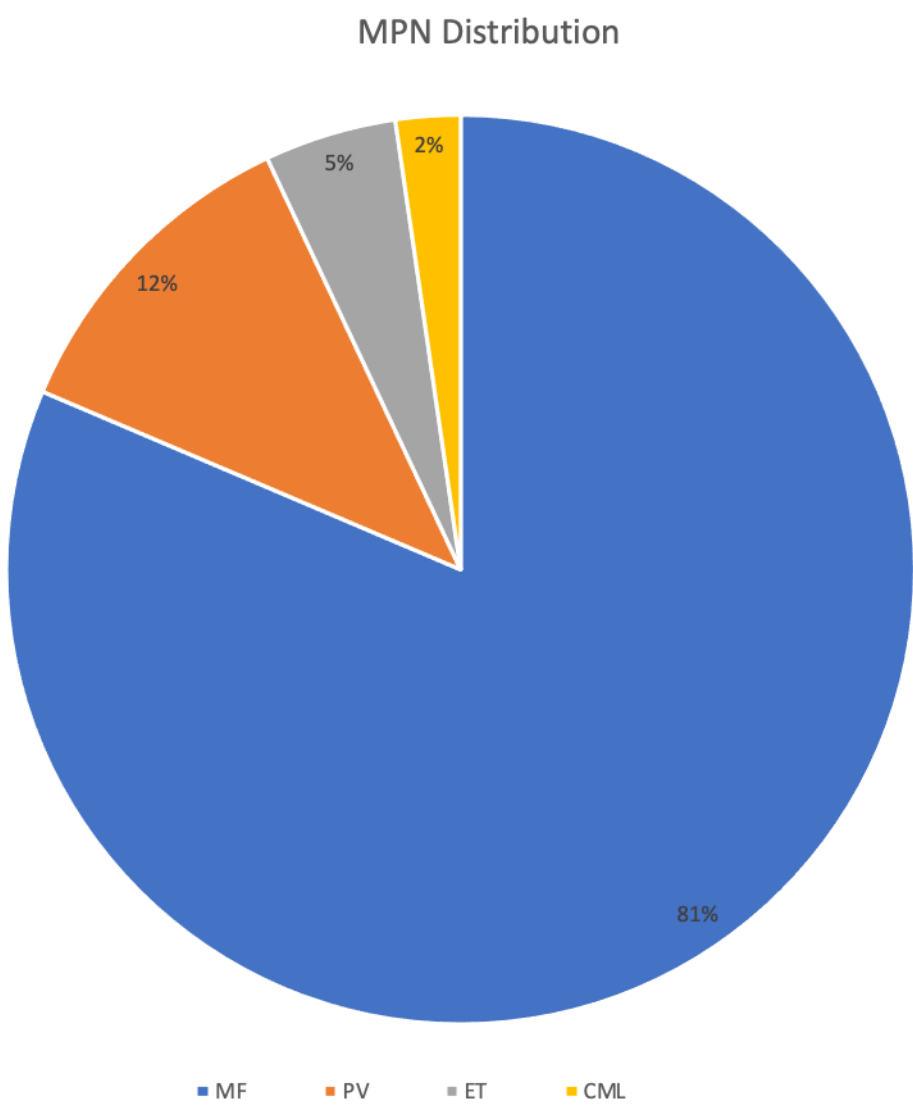


Fig. 2 MPN Distribution Percentage

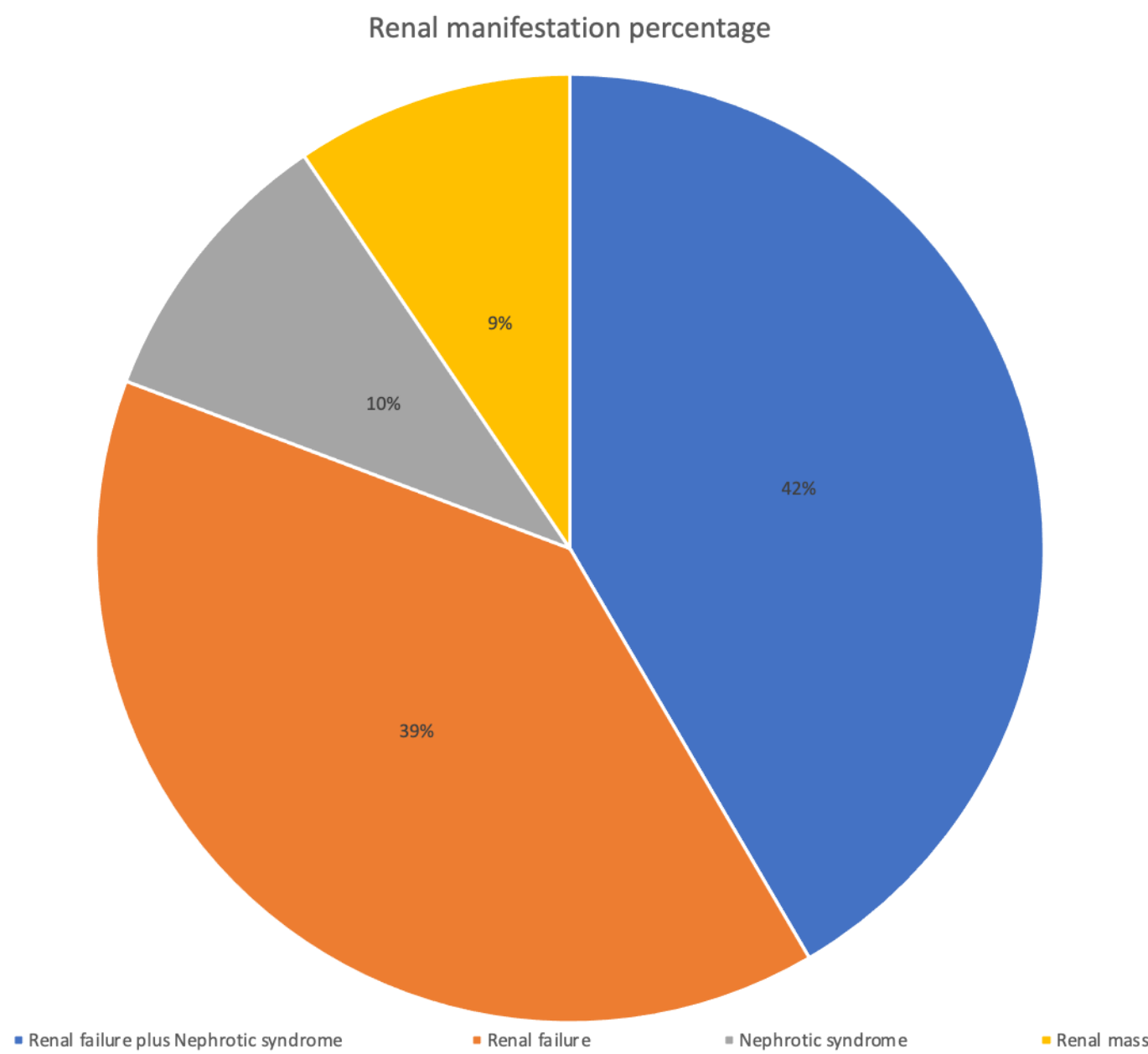


Fig. 3 Renal Manifestation Distribution

RESULTS (CONTINUED)

- The most common renal manifestation was renal failure and nephrotic syndrome in 39.5 % of the cases, followed by renal failure alone 37.2 % and nephrotic syndrome only in 9.3 % and interestingly a renal mass was found 9% .
- Average duration to developing EMH from time of diagnosis was 7.8 years (0- 20 years). Four patients developed EMH post-transplant, 3 of them allogenic while one autologous.
- The JAK mutation was reported in 7 patients and CALR mutation in one patient.

RESULTS (CONTINUED)

- Patient were treated with different medication including diuretics, anti-hypertensive (mainly ACE inhibitors) and steroids. However, for MPNs 25 % received HU, 14 % received Thalidomide or Thalidomide analogue while 16 % received JAK inhibitors; Ruxolitinib. The CML case was treated with Imatinib.
- The average duration of Follow up was 19 months with 42.5% death rate by the end of follow up.
- Kidney function improved in 31 % of patients, with Ruxolitinib administration correlating with improved kidney function and survival (6 out of 7 patients).

CONCLUSIONS

- Unexplained Increase in renal function or proteinuria or kidney mass in a setting of MPNs, should raise the suspicion of this diagnosis.
- There is currently no consensus or guidelines on management of such cases.
- Ruxolitinib with the limitations of the small numbers treated may offer some benefit, however further studies and exploration of this drug and others in well-constructed prospective studies is needed.

REFERENCES

1. Said SM et al. Myeloproliferative neoplasms cause glomerulopathy. *Kidney Int.* 2011; 80: 753–759.
2. Caijo Y et al. [Glomerular disease associated with myelofibrosis (author's transl)]. *Sem Hop.* 1979; 55: 489–493.
3. Perazella MA et al. Nephrotic syndrome associated with agnogenic myeloid metaplasia. *Am J Nephrol.* 1994; 14: 223–225.
4. Holt SG et al. Extramedullary haematopoeisis in the renal parenchyma as a cause of acute renal failure in myelofibrosis. *Nephrol Dial Transplant.* 1995; 10: 1438–1440.
5. Schnuelle P et al. Idiopathic myelofibrosis with extramedullary hematopoiesis in the kidneys. *Clin Nephrol.* 1999; 52: 256–262.
6. Au WY et al. Focal segmental glomerulosclerosis and mesangial sclerosis associated with myeloproliferative disorders. *Am J Kidney Dis.* 1999; 34: 889–893.
7. Woodward N et al. Renal myelofibrosis: an unusual cause of renal impairment. *Nephrol Dial Transplant.* 2000; 15: 257–258.
8. Pamuk ON et al. Nephrotic syndrome associated with agnogenic myeloid metaplasia. *Leuk Lymphoma.* 2002; 43: 661–663.
9. Sukov WR et al. Sclerosing extramedullary hematopoietic tumor: emphasis on diagnosis by renal biopsy. *Ann Diagn Pathol.* 2009; 13: 127–131.
10. Kaygusuz I et al. Focal segmental glomerulosclerosis associated with idiopathic myelofibrosis. *Ren Fail.* 2010; 32: 273–276.
11. Rajasekaran A et al. Primary myelofibrosis associated glomerulopathy: significant improvement after therapy with ruxolitinib. *BMC Nephrol.* 2015; 16: 121.
12. Koch CA et al. Nonhepatosplenic extramedullary hematopoiesis: associated diseases, pathology, clinical course, and treatment. *Mayo Clin Proc.* 2003; 78: 1223–
13. Christensen AS et al. Chronic kidney disease in patients with the Philadelphia-negative chronic myeloproliferative neoplasms. *Leuk Res.* 2014; 38: 490–495.