Case Report: A Rare Case of MPN-PV Treated With a High Dose of Jakafi

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Introduction

- **Myelofibrosis (MF) and polycythemia vera (PV) are Philadelphia chromosome (BCR-ABL1)-negative subtypes of chronic myeloproliferative neoplasms (1). Patients diagnosed with PV experience a broad range of symptoms associated with a reduced quality of life, and have an increased risk of death compared to age-matched subjects without PV.**

Case presentation

- **We report a case of 59 years old female with complaints of generalized weakness, fatigue and abdominal discomfort, diagnosed on September 2010 with Myeloproliferative neoplasm (MPN)-polycythemia Vera (PV).**
- **The routine blood tests revealed a Hct 57%, Plt 457G/L, WBC 11G/L. A PCR-BCR-ABL came back negative whereas the JAK2 was positive. On general examination she had a Splenomegaly of 18 cm, with no palpable lymph nodes.**
- **Patient started Phlebotomies every 2 to 3 months. Her lab results were well controlled till April 2013, where more phlebotomies were needed each month with an increase of the platelet's counts > 1000000 G/L, Hct uncontrolled (range?), and WBC reaching 18 G/L.**
- **On May 2013, Hydroxyurea was initiated with dose adjustment according to the count level along with Phlebotomies. Hydroxyurea was interrupted few times because of skin, GI toxicities and Leukopenia's.**
- **on June 2017, the patient presented for follow-up with severe myalgias, fatigue and abdominal discomfort. On examination, She had a Splenomegaly 20 cm.**
- **July 2017: initiation of Jakavi (Ruxolitinb) at 5 mg twice per day whereas the recommended dose is 10 mg twice per day.**
- **After 6 months of introducing the Jakavi, clinical and biological improvement was seen with a Hct 51%, WBC 11.8, Plt 433 G/L with a partial response.**
- **Jakavi was increased to15 mg twice per day because the 10 mg was not available. Blood tests on October 2018 showed Hct 39%, platelets 267 and spleen measurement of 9.4 cm.**
- **Two years later on October 2020, the patient was still on Jakavi 15 mg twice daily with a complete remission, normalization of blood test and spleen measurement**

Discussion

- **This case report describes a rare case of Polycythemia Vera .**
- **No reported cases in MPN-PV treated in 2 different doses with a clinical and biological improvement .**
- **Ruxolitinib, a JAK1/JAK2 inhibitor (2011) (2) and fedratinib, a JAK2/FLT3 inhibitor (2019) (3) are approved in the USA for MF patients with splenomegaly and/or constitutional symptoms regardless of the presence of mutated *JAK2***

Conclusions

- **Our case of a 59 year-old female with MPN-PV following 2 different doses of treatments is quite unique and warrants to be reported.**
- **Ruxolitinib demonstrates a noteworthy evolution in treatment options by offering patients the hope of living longer and experiencing significant improvement in quality of life.**
- **Routine patient monitoring and optimized management of patients who present special challenges will help improve patient outcomes**
- **There is a need to conduct more observational and experimental studies to understand the mechanisms of association between MPN-PV and the chemotherapy .**
- **Monitor for signs of disease progression, including hydroxyurea resistance or intolerance and changes in symptom severity based on routine assessments with a validated objective tool such as the MPN-SAF.**

References

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