

An Interesting Case of Mucosal Bleed - A Case of Polycythemia Vera

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1. Introduction

Polycythemia Vera (PV) is one of the Myeloproliferative Neoplasm (MPN). It is characterized as a panhyperplastic, malignant, and neoplastic marrow disorder occurring due to an abnormal clone of the hematopoietic stem cells with an increased sensitivity to the different growth factors for maturation. Its most prominent feature is an elevated absolute red blood cell mass because of uncontrolled red blood cell production. This is accompanied by an increased WBC (myeloid), and platelet (megakaryocytic) production.

2. Case Report

A 41 years old male patient, presented with complaints of one episode of oral bleed while washing

mouth a day before. He had previous history of occasional bleeding while biting an apple (blood staining around the bite mark). There was no history of trauma while brushing, tongue bite, bleeding from other body orifices, petechiae or purpura, coughing or straining, fever, numbness or weakness, breathlessness or orthopnoea. No other systemic complaints were present. Patient is a known case of T2DM for 6 years and is on regular treatment for it. There were no other known co-morbidities. The patient didn't have any addiction. There was no significant family history and past history. On examination, red inflamed gingiva was present. Vitality, tachycardia of 110/min. was present. BP was 190/110 mmHg after 30 minutes of rest. Per-abdominal examination revealed splenomegaly.



Fig.: Red Inflamed Gingiva

Lab. Reports:

Hb.: 21.3 g/dl, TC: 15,590 cells/cu.mm., Platelet: 6,17,000/cu.mm., RBC count: 7.65 million/cu.mm., RDW: 20.4%, PCV: 65.3%

Peripheral Smear: Normocytic Normochromic in appearance, WBCs appears increased in numbers, Thrombocytosis seen, No hemoparasites seen.

PT: 19.6 (Control=14.0), INR: 1.42

FBS: 132mg/dl, 171mg/dl, HBA1C: 8.1%, eAG(estimated average glucose): 211.1mg/dl

T.Cholesterol: 162mg/dl, HDL: 43mg/dl, TG: 82mg/dl, LDL: 113mg/dl, T.Cholesterol:HDL ratio: 3.8

Urea: 20.3mg/dl, S.Creatinine: 0.86mg/dl

Spot urine microalbumin: 1000.4mcg/mg of creatinine (↑)

Urine Routine: +2 proteins, 3-4 pus cells/hpf, nil RBCs

ECG: Normal Sinus Rhythm, Within normal limits

USG Abdomen: Moderate Splenomegaly (16.3 cm in size and shows uniform echotexture), Prostatomegaly grade I.

COVID-19 RT-PCR: Negative

S. EPO: 44.9mIU/ml (normal: 5.4-31mIU/ml)

JAK2V617F mutation by PCR: POSITIVE

Clinical Course:

Patient was admitted with the above mentioned complaints. Based on history, clinical examination and investigations, diagnosis of Polycythemia Vera with T2DM with S.HTn was made. Patient was started on anti-platelet, anti-hypertensives and oral hypoglycemic agents. Therapeutic Phlebotomy was done and around 350ml of blood was withdrawn.

Hematologist opinion was taken and advise followed. Repeat investigations done showed Hb.- 19 g/dl, TC- 15420/cu.mm., Platelets- 7.03 lakhs/cu.mm., RBC count- 6.68 millions/cu.mm. Hematocrit(PCV)- 59.7%, S.EPO level: 9.9 mIU/ml (normal: 4.3-29.0 mIU/ml). Patient is now on anti-platelet, anti-hypertensives and oral hypoglycemic agents and, is under follow-up for weekly phlebotomy until his PCV reaches a target of <45%. He is advised to strictly adhere to the medications and to maintain adequate hydration to minimize the risk of complications.

3. Discussion

Polycythemia Vera (PV) is an acquired clonal hematopoietic stem cell disorder with poorly understood etiology. Mutation in the tyrosine kinase JAK2 (JAK2V617F mutation) is seen in ~95% patients with PV. Its more common in males and is usually seen in people older than 40 years. Clinical features are mainly due to impaired oxygen delivery due to sludging of blood and mainly includes headache, dizziness, visual disturbances, aquagenic pruritis and an increase in incidence of thrombotic &/or bleeding complications. Physical examination may reveal splenomegaly (70%), hepatomegaly (30%), plethora and hypertension. Diagnosis is mainly based on an increase in Hemoglobin, PCV and RBC mass. Hematocrit (PCV) is a more reliable indicator of polycythemia than is Hemoglobin. Demonstration of JAK2V617F mutation confirms the diagnosis. Treatment ranges from low dose aspirin to reduce thrombotic episodes and therapeutic phlebotomy, to chemotherapy using Hydroxyurea or Busulphan, to

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using radioactive Phosphorus-32. Median survival in patients with PV has been extended to approximately 14 years overall because of new therapeutic tools.

4. Conclusion

A primary goal for the treatment of the patients with Polycythemia Vera is to reduce the risk of thrombosis, and to prevent secondary complications. Polycythemia Vera patients should maintain hematocrit <45% using therapeutic phlebotomy and if needed, using cytoreductive therapy also.

Conflicts Of Interest The authors declare no conflict of interest

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