

Polycythemia Vera in a Patient with Breast Cancer

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Background

A 65 years old female non-smoker who presented with left breast mass, general weakness and hepatosplenomegaly. On examination there was facial plethora and left breast swelling since two years ago which increased gradually in size in last month. Her blood pressure was 130/90, pulse rate 90 per minute. Cardiac examination revealed normal heart sound and no extra-sound, murmurs or gallops, peripheral plus were normal in all four extremities. Her chest was normal on auscultation and there were no signs of clubbing or cyanosis, abdominal examination soft with palpable liver and spleen.

Investigations on Admission

Haemoglobin 19.8 gm/dl, RBC 7.65 million, HCT 60.1%, MCV 78.5 L/l, MCH 25.9 L/pg, MCHC 32.9 g/dl, leukocytes 6.200 mm³ neutrophils 52%, monocytes 7%, lymphocytes 41%, platelet

498.000 mm³. Hepatic enzymes and LDH were normal. Creatinine was 0.7; she had never received a red blood cell transfusion. Her Chest CT scan showed ill-defined irregular mass lesion with areas of necrosis in the left breast measuring 27 × 23 cm and enlarged left axillary lymph nodes 1 cm, otherwise normal. Echocardiography is normal and abdominal sonography showed moderate hepatosplenomegaly with gallbladder hyper-echoic deposit, measured about 6.11 mm at the posterior wall could be stone.

She underwent modified-radical mastectomy on 18/2/2016, Biopsy report: Invasive ductal carcinoma, grade III with eight out of ten lymph nodes were positive for malignancy staged T1N1Mx with triple negative receptors. She started chemotherapy cyclophosphamide 800 mg, Epirubicin 160 mg and 5-Fluorouracil 800 mg every 3 weeks and completed 6 courses followed by radiotherapy (Table 1). Gradual and dramatic normalization of complete blood count during chemotherapy and during the 30 months after chemotherapy.

FEC Chemotherapy	Hemoglobin/ gm/dl	Hematocrit/%	WBC/mm ³	RBC/10 ¹² /L	Platelets/mm ³	Abdomen Sonogram
Before chemotherapy	19.8	60.1	6200	7.65	498000	Hepatosplenomegaly
After 3 courses	13.2	39.1	4100	4.68	510000	Normal
After 6 courses	12.1	38.4	4300	4.6	643000	Normal
After 6 months	13	38.7	6700	4.8	586000	Normal
After 1 year	13.3	38.5	4100	4.9	510000	Normal
After 2 years	15.2	46	11000	6	343000	Normal

Table 1: Follow up before, during and after chemotherapy.

Polycythemia Vera (PV), also known as erythrocytosis or polycythemia (rubra) vera is being first recognized in 1951 by William Dameshek as “myeloproliferative neoplasia (MPN)” [1]. The classic Philadelphia-negative MPNs, as they are now referred to include primary myelofibrosis (PMF), polycythemia vera (PV) and essential thrombocythemia (ET), each is characterized by mutually exclusive janus kinase 2 (JAK2), calreticulin (CALR) and myeloproliferative leukemia oncogene virus (MPL) mutations. JAK2 mutation is the most frequently occurring gene mutation, occurring in approximately 98% of PV cases, 50% to 60% of ET cases and 55% to 65% of PMF cases [2].

The mechanisms underlying disease progression and symptom development are not very well understood. PV characterized by the over production of red blood cells as a result of acquired mutations in an early blood-forming cell. Since these early blood-forming cells have the capability to form not only red cells, but also white cells and platelets, any combination of these cell lines may be affected. This

condition develops slowly and may remain undetected for many years. Polycythemia vera affects slightly more men than women. The disorder is estimated to affect approximately 2 people per 100,000 in the general population. It occurs most often in individuals more than 60 year old, but can affect individuals under 20.

Typical symptoms include fatigue, pruritus, loss of appetite, night sweats, splenomegaly, abdominal pain, bone pain, weight loss, microvascular complications and anemia [3]. In MPN and especially polycythemia vera which is severely understudied and there is no literature exploring pharmacological approaches of FEC chemotherapy (Epirubicin, Cyclophosphamid and 5-Fluorouracil) to help improve symptom burden and QoL in these patients, it is hoped that this new observations. To assure comparability between the two diseases and the effects of epirubicin, cyclophosphamide and 5-Fluorouracil, we suggest conduct further investigations and observations.

References

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