A Case of Secondary Polycythemia

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Abstract : According to the WHO, polycythemia is characterized by either an elevated hematocrit level (above 49% in males and 48% in women) or an elevated haemoglobin level (above 16.5 g/dl in men and 16.0 g/dl in women). There are two types of polycythemia: primary and secondary.A 36-year-old male patient complained of giddiness for two years on and off. He had been taking regular medication for dyslipidemia and hypertension for a year. He had been an alcoholic and smoker for 15 years before giving up smoking last year. Atorvastatin 10 mg plus 75 mg of aspirin, known as ecospirin-AV, was used to begin the patient's treatment. Phlebotomy was performed on the patient. Smoking appeared to be a key factor in this case, and it was believed to be the primary cause of secondary polycythemia.The therapy options for erythrocytosis include phlebotomy and low-dose aspirin.

Index Terms : Secondary polycythemia, phlebotomy, Hemoglobin, Erythropoietin, Polycythemia Vera (PV)

Introduction

According to the World Health Organization (WHO), polycythemia is characterized by either an elevated hematocrit level (above 49% in men and 48% in women) or an increased haemoglobin level (above 16.5 g/dl in men and 16.0 g/dl in women)[1]. The Greek words poly (many) and cythemia (blood cells) are the origin of the word polycythemia[2].

There are thought to be 22 cases of polycythemia vera per 100,000 people. The prevalence shows a male preponderance across all racial and ethnic groups, with a male-to-female ratio of about two to one. The typical age of PV presentation is 60 years, and patients are rarely seen before the age of 40 [9]. There is a dearth of information on the epidemiology of secondary polycythemia[2] .

There are two categories of polycythemia:Primary and Secondary.The raised proliferation of erythroid primogenitor cells as a result of a cellular abnormality which causes primary polycythemia. Erythropoietin levels are suppressed in these patients. It refers in two main categories. Vera polycythemia Due to a mutation known as the JAK mutation, this neoplastic condition is caused by an increase in erythroid progenitor cells and

an increase in erythropoietin sensitivity. and Pure erythrocytosis: Patients who have pure erythrocytosis are those who just have an isolated increased RBC mass without any additional precipitating factors[2].

Because of chronic elevations in erythropoietin levels caused by hypoxia, secondary polycythemia (SP) is a condition where the body has abnormally high volumes of red blood cells[3]. As a result, the blood becomes thicker, increasing the risk of a stroke. It is a uncommon condition where the primary function of the red blood cells is to carry oxygen from the lungs to every cell in the body. RBC production occurs continually in the bone marrow.Within a few weeks, the body will start to manufacture more red blood cells if one relocates to a higher altitude where oxygen is more scarce [4].

There will be elevated EPO levels and red blood cell counts in secondary polycythemia. Alcohol abuse, Hypertension ,Obesity and smoking are risk factors for secondary polycythemia (erythrocytosis)[4]. Breathing issues, chest discomfort, abdominal pain, weariness, weakness, muscle soreness, headache, tinnitus, blurred vision, a burning or "pins and needles" sensation in the hands, arms, legs, or feet, and mental sluggishness are all signs of secondary polycythemia.[4].

Low-dose aspirin (40 mg to 100 mg once or twice day) is the mainstay of treatment for secondary polycythemia[10].It works as a blood thinner and reduces the risk of stroke (thrombosis) brought on by erythrocytosis, an excessive red blood cell synthesis. Phlebotomy and venesection are two blood withdrawal methods that both remove up to a pint of blood to reduce the concentration of red blood cells in the blood [4]

Case report :

A 36 years old male weighing 76kg presented to the general medicine department with complaints of on and off giddiness for 2 years. 1 month ago he was admitted for complaints of rash and discoloration of skin for which he was treated with sapat lotion (salicylic acid).

His past medical history revealed that he is a known case of dyslipidemia and hypertension for 1 year for which he is taking amlodipine 5mg once daily. He was a smoker and alcoholic for 15 years and discontinued smoking 1 year ago. He had a history of falling to either side on sudden stops while walking and increased giddiness on standing.

The patient was examined, and his blood pressure was found to be normal at 148/90 mmHg on day 1 and 150/90 mmHg on day 2. The patient was conscious, oriented, and lethargic. The respiratory systems and cardiovascular systems were normal.. An abdominal exam revealed that it was soft, non-tender, and free of organomegaly. Haemoglobin levels were 20.2g/dl (13-18g/dl), hematocrit was 60% (40-54%), red blood cells count was 5.54 $x10^6$/microliter (4.50-6.50 $x10^6$/microliter), platelet count was 258 $x10^3$/microlitre (150 - 450

x10^3/microlitre), Total cholesterol was 229 mg/dl (<200mg/dl), triglycerides was 141mg/dl (<150 mg/dl), High-density lipoprotein (HDL) was 33mg/dl (>60 mg/dl), Low-density lipoprotein (LDL) was 167mg/dl (<100mg/dl) and Very low-density lipoprotein (VLDL) was 28mg/dl (<30mg/dl).

Blood tests for kidney and liver function, random blood sugar, and serum electrolytes were all within acceptable limits. An picture of normocytic normochromic blood was confirmed by peripheral smear. A pelvic and abdominal ultrasound revealed mild hepatomegaly and a fatty liver. On many occasions, the oxygen saturation levels (SpO2) varied between 92% and 94%.Left ventricular hypertrophy with a 55% ejection fraction was visible on a 2D echo. The remaining organs were healthy.

High Hemoglobin and Hematocrit prompted further workup and consideration of the possibility of PV. To exclude PV, revised WHO 2016 diagnostic criteria for myeloid neoplasms were employed. A bone marrow biopsy was performed, and the results revealed erythroid and megakaryocytic hyperplasia with normal megakaryocyte shape.

EPO levels in the serum was14 IU/ml (4 to 24 IU/ml). The family history of JAK2 was lacking and the cytogenetic investigation for the JAK2V617 mutation was negative. This led to the elimination of PV as a possibility. Smoking appeared to be a significant factor in this case, and it was believed to be the primary cause of secondary polycythemia.

Atorvastatin 10 mg plus 75 mg of low-dose aspirin was used to begin the patient's treatment. The patient had phlebotomy and went more than a year without experiencing any symptoms.

Discussion :

The diagnosis of polycythemia, which is based on a composite evaluation of clinical and laboratory features, is currently made using the updated WHO 2016 criteria[5].Above the age of 60, PV is more prevalent [6].In the current case, the patient is a 36-year male diagnosed with secondary polycythemia. The patient is a smoker with a 1-year history of Hypertension. In a laboratory investigation, a greater haemoglobin level was discovered, satisfying the first important PV criteria. A bone marrow biopsy that indicated erythroid and megakaryocytic hyperplasia with normal megakaryocyte morphology supported the second essential criteria. Although it was negative in the current case, the JAK2V617 mutation is the third crucial element in the diagnosis of PV. Since the serum EPO level was normal, the minor criteria for the diagnosis of PV were not met.

This eliminated the possibility of PV and left the decision of whether to diagnose secondary polycythemia or relative polycythemia up to the individual. Smoking history and high blood pressure favour secondary polycythemia.

The patient responded favorably to phlebotomy..Phlebotomy produced a personal advantage in the mature in instances with severe chronic pulmonary disease and subsequent polycythemia, according to Dayton and colleagues[7].In contrast to the former, secondary polycythemia is characterized by an increase in RBC mass brought on by elevated serum EPO levels, but Mahe et al.[8] suggestion that this is not always the case.Investigation for the purpose begins with repeat and confirmation of the raised hemoglobin and size of an EPO degree to indicate whether to pursue primary or secondary causes after which in addition investigations as suitable[8].The management alternatives for erythrocytosis encompass low-dose aspirin and phlebotomy.However, there is no proof that phlebotomy should be used often to treat secondary polycythemia[8]. But as per our hospital guidelines, we proceeded with phlebotomy for our patient.

Conclusion :

In order to distinguish between primary and secondary polycythemia, a thorough history, comprehensive physical, general examination, and laboratory investigations are necessary. Phlebotomy can benefit patients with secondary polycythemia and enhance their overall well-being.

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