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GAISBOCK'S SYNDROME: A CASE STUDY

Hematology

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ABSTRACT

Felix Gaisbock, MD, (1905) described a syndrome in patients with hypertension, increased hematocrit levels, plethoric appearances, and without splenomegaly. He hypothesized that this relative erythrocytosis was stress related. In this case report, a 38-year-old hypertensive male presented with constitutional symptoms along with new onset polycythemia. There was no splenomegaly. A workup did not show any evidence of polycythemia rubra vera (PRV) or some other secondary cause of his polycythemia. He was given appropriate supportive therapy for his constitutional symptoms. His hematocrit returned to normal levels over the next few days under observation. We concluded that this patient had Gaisbock's syndrome, a form of relative polycythemia that manifests when there is evidence of plasma contraction.

KEYWORDS

Gaisböck syndrome; erythrocytosis; arterial hypertension; polycythemia,

INTRODUCTION

Gaisbock's syndrome is a symptom complex linked with polycythemia that cannot be attributed to a diagnosis of polycythemia rubra vera or a secondary erythrocytosis caused by hypoxemia. It was first documented in 1905. Male sex, hypertension, smoking, diuretic medication, obesity, and emotional or physical stress are all risk factors for the development of Gaisbock's syndrome, in order of decreasing importance. Polycythemia has been linked to decreased plasma volume, as well as increased blood viscosity and peripheral vascular resistance, as well as and an increased risk of arterial and/or venous thrombosis. [1, 2, 3]

Patients with this syndrome typically have high blood pressure, blood viscosity, plasma proteins, serum cholesterol, uric acid, and salt excretion in the urine. The decrease in plasma volume associated with an increase in red cell count appeared to be linked to an increase in blood pressure. [4, 5]

We discuss the case of a 34-year-old male with poorly controlled diastolic hypertension who developed a febrile flu-like illness and new-onset polycythemia that cleared up over several days. The results of tests for the main and secondary causes of polycythemia were negative, confirming Gaisbock's condition.

Case report

Patient was a 34-year-old male who presented to the OPD with complaints of:

- Cough and cold for 2 days
- Sudden onset shortness of breath for 1 day
- Headache with an episode of vomiting for 1 day.

No history of TB, bronchial asthma, type 2 diabetes mellitus. Patient was a chronic alcoholic. Patient was a known case of hypertension and was on medication for 1 year. He had however missed his dosage for two consecutive days before presentation.

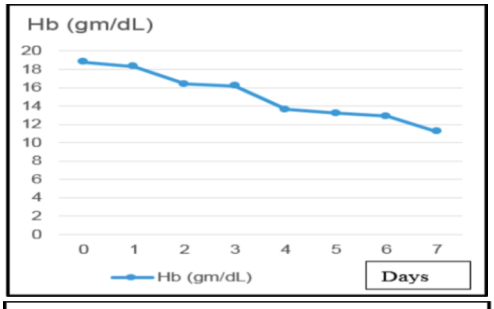
On examination:

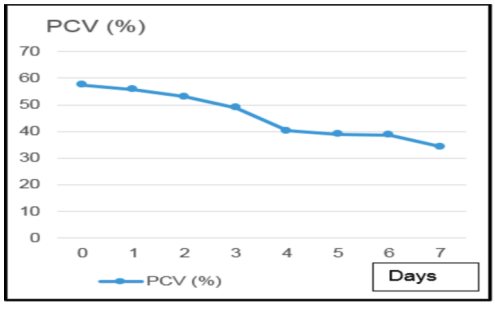
- Blood pressure: 280/200 mmHg
- Pulse rate: 150/minute
- Respiratory rate: 52/minute
- SpO2: 56% at room atmosphere
- Chest auscultation: Bilateral coarse crepitations were present
- CNS examination: drowsy and disoriented
- CVS examination: S1 and S2 were normal, no murmurs
- Per abdomen examination: soft and non-tender. No organomegaly noted.

Laboratory investigations revealed:

- Hemoglobin: 18.8 g/dL
- RBC count: 5.91 million/cumm
- Hematocrit: 57.7%
- WBC count: 21,140/cumm
- Platelet count: 1.68/cumm
- Serum uric acid levels: 6.3 mg/dL
- Serum cholesterol: 274 mg/dL
- Liver and kidney function tests: normal
- JAK-2 assay negative
- Serum erythropoietin: normal

The blood pressure was managed using nitroglycerine and diuretic infusions initially. Subsequently the patient was administered once daily amlodipine 5 mg. Serum erythropoietin levels were estimated, and the patient was tested for a Janus Kinase v617F (JAK-2) mutation, in view of his high hemoglobin and hematocrit. Erythropoietin levels were normal, and the patient's JAK-2 v617F mutation was negative. The patient was referred for therapeutic phlebotomy to the blood center. However, it was decided to observe the patient over a few days before initiating phlebotomy procedure. The patient's condition improved through blood pressure control obviating the need for therapeutic phlebotomy. The hemoglobin levels returned to normal levels (14.5 g/L). The final diagnosis was Gaisbock's syndrome associated with arterial hypertension.





DISCUSSION

Polycythemia is characterized as a higher-than-normal level of hemoglobin. This rise could be real or just visible due to a decrease in plasma volume (spurious or relative polycythemia) or in chronic inflammatory states [6]. Although the terms erythrocytosis and polycythemia are sometimes used interchangeably, some people distinguish between the two: erythrocytosis refers to increased red cell mass, whereas polycythemia refers to any increase in red cells.

Erythrocytosis refers to either a true or apparent increase in hemoglobin (Hgb)/hematocrit (Hct); distinction requires familiarity with sex-, race- and altitude-adjusted normal values, together with an appreciation of extreme normal values that exceed the 95th percentile (2 SD) of reference range and attention to clinical factors associated with plasma volume depletion (relative erythrocytosis). [10]

In 2016, the WHO classification system proposed the Hgb/Hct thresholds for diagnosis of polycythemia vera as 16.5 g/dL/49% in males and 16 g/dL/48% in females. [15]

Polycythemia in patients is frequently discovered because of an unintentional finding of high hemoglobin or hematocrit levels. At 170 g/L (17 g/dL) for men and 150 g/L (15 g/dL) for women, there is usually cause for concern that the hemoglobin level is abnormally high. Hematocrit levels of >50% in men and >45% in women are considered abnormal. Hematocrits of more than 60% in men and more than 55% in women are almost always linked to an increase in red cell mass. [12]

Even though we were unable to determine the patient's plasma volume and red cell mass, the patient's hematologic and clinical characteristics, as well as the patient's course, is consistent with Gaisbock's syndrome, in which polycythemia is attributed to a decrease in plasma volume. [1]

Gaisbock's syndrome is a rare condition marked by high hematocrit and hemoglobin levels in the absence of splenomegaly, leukocytosis, or thrombocytosis. Gaisbock first described the condition in 1905. Patients with this syndrome typically have high blood pressure, blood viscosity, plasma proteins, serum cholesterol, uric acid, and salt excretion in the urine. [7, 8, 9] The decrease in plasma volume associated with a relative rise in red cell count appeared to be linked to an increase in blood pressure. Most patients with relative polycythemia or erythrocytosis are obese men. Patients with secondary polycythemia frequently present at a younger age than those with primary polycythemia vera. Hypertension, smoking, alcohol misuse, and chronic renal illness are all known to be highly related to relative polycythemia. [2]

Patients with relative polycythemias fall into two categories. The first group includes patients who have significant dehydration because of an evident cause, such as gastrointestinal fluid loss, therapeutic diuresis, or hypercalcemia with significant extracellular fluid loss. The cause of a rise in hematocrit normally does not provide a diagnostic issue. The second group is linked to a steady increase in hematocrit over time. These are usually middle-aged, mildly hypertensive, obese individuals who are under a lot of stress and have persistent polycythemia. They have the plethoric appearance of polycythemia vera but none of the other characteristics. The origin of the decrease in plasma volume is unknown; however one theory explains it because of autonomic dysregulation and alterations in venous capacitance. [14]

The best way to treat relative polycythemia isn't known. At least two-thirds of patients can achieve satisfactory hematocrit control by losing weight, improving hypertension control, avoiding diuretics, and reducing, if not quitting smoking. Venesection is rarely necessary, but it should be considered if the patient's PCV levels are high enough to put them at risk of thrombotic events (i.e., > 54 mL/dL) or if they are suffering signs of cardiovascular ischemia. Even in individuals with relative polycythemia, phlebotomy increases cerebral blood flow; however, whether it is of clinical benefit is less evident. It's probably best to avoid it. Theoretically, further reducing the blood volume in these patients who are already normovolemic or slightly hypovolemic could compromise tissue perfusion. [2, 11]

Krishnamurthy P et al [13] studied large inpatient database through the national registry and identified all patients aged 18 years or more who were diagnosed with polycythemia. They concluded that polycythemia was associated with an increased prevalence of hypertension and the presence of polycythemia was a strong determinant of hypertension even after accounting for other potential confounding risk factors. They also analyzed that polycythemia remained strongly associated with hypertension, even after adjusting for other confounding risk factors and was associated with increased in hospital mortality and sudden cardiac death.

One of the rarest causes of hypertension is Gaisbock's syndrome. Our patient had a hypertensive emergency, which made this case even more unusual. To prevent needless phlebotomy and other polycythemia vera therapies, bear this unusual disease in mind when dealing with patients of polycythemia without splenomegaly and with hypertension.

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Patient Consent Declaration: Patient's consent not required as the patient's identity is not disclosed or compromised.

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