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Gaisbock's Syndrome: A Rare Cause of Hypertensive Emergency

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Abstract: Gaisböck Syndrome is a condition characterized by apparent polycythemia, in the absence of an elevated red cell mass, because of plasma volume contraction. There is no true polycythemia. It is a coincidence of polycythemia and hypertension. As these patients are followed up with persistently elevated hematocrit values, it becomes evident that this entity is distinct from polycythemia vera. This is important since patients who have had neither an elevated total red blood cell volume, nor polycythemia vera, have been treated by phlebotomy and radioactive phosphorus. This error in diagnosis and treatment is more common than is realized. We present a case of a 40 year old male who presented with elevated hematocrit levels with hypertension and was finally diagnosed to be a case of gaisbock's syndrome.
Keywords: Gaisbock's syndrome, hypertension, polycythemia Vera, hematocrit.

INTRODUCTION

Gaisböck Syndrome otherwise known as chronic relative polycythemia or erythrocytosis has been variously referred to as [1], "stress" erythrocytosis, benign polycythemia, benign erythrocytosis[2], spurious polycythemia [3,4], pseudopolycythemia [5], and apparent polycythemia [6], which occurs due to relative plasma volume contraction. Gaisbock's syndrome is a peculiar entity characterized by elevated hematocrit and hemoglobin levels in a patient without splenomegaly, leukocytosis and thrombocytosis.

Patient suffering from this syndrome usually had elevated blood pressure, elevated blood viscosity, plasma proteins, serum cholesterol, uric acid and increased excretion in urinary sodium. The reduction in plasma volume with relative increase in red cell count seemed related to elevation of blood pressure [7, 8].

CASE

A 40 year old male presented to us with chief complaints of headache since 15 days and fatigue since 10 days. There was no history of fever, rhinorrhea, neck pain, photophobia, abnormal mentation, diplopia, seizures and any kind of stress. There was no past history of hypertension, diabetes mellitus, and migraine. He was farmer by occupation and was not disturbed by headache. No history of smoking, alcoholism, tobacco chewing. Family history was not significant.

On examination, patient was normal built, BMI was 22.41 Kg/m ^{2}, oriented to time, place and person. Pulse was 102/min, regular, no radio femoral delay, without any special character. Blood pressure was 220/120 mm hg in left arm, 236/128 mm hg in left lower limb. JVP was normal, there was no edema feet.CVS examination revealed a forceful apex and S4 on auscultation. Other system examination was within normal limit.

Routine investigations revealed hemoglobin 20.9 g/dl, RBC counts- 6 lakhs/cumm, hematocrit -55.9,WBC counts -8400/cumm, platelet count-2,39000/cumm. Serum uric acid levels were 7.9 mg/dl and serum cholesterol levels were 248 mg/dl. Liver function test, Kidney function test,Blood sugar levels were within normal limits.USG abdomen and pelvis and bilateral renal artery Doppler study were normal.

The blood pressure was controlled by nitroglycerine infusion over 24 hours and later on patient was switched to tablet amlodepin 10 mg and tablet telmisartan 40 mg once a day. In view of abnormally elevated hemoglobin and hematocrit, serum

erythropoietin levels were done and patient was evaluated for janase kinase mutation (JAK 2) .Erythropoietin levels were normal and patient was negative for JAK 2 mutation. A final diagnosis of hypertension induced Gaisbock's syndrome was made. After 2 weeks the hematocrit came down to 44.5 and patient was called for follow up after 2 months. The clinical profile at the time of discharge is shown in table 1.

Table-1: Showing the comparision of Variables on admission and on discharge

Symptoms	On admission	On discharge
Headache	present	relieved
Fatigue	marked	improved
Signs		
Pulse	102/min	84/min
Blood pressure	220/120 mm hg on upper limb 236/128 mm hg on lower limb	140/80 mm hg on upper limb 150/90 mm hg on lower limb
Cardiovascular system	S 4 heart sound present	absent
Investigations		
Hemoglobin	20.9 g/dl	14.0 g/dl
Hematocrit	55.9%	44.5%
Uric acid	7.9 mg/dl	6.8 mg/dl
Total cholesterol	248 mg/dl	235 mg/dl

Table-2: shows discussion of Differential diagnosis with diagnosis of exclusion in our case.

Differential diagnosis	Points of exclusion in our case
Polycythemia rubra vera	-No splenomegaly -JAK2 mutation negative -Normal whole blood count and platelet count
Secondary polycythemia	-Non smoker -Not residing at any high altitude area -No congenital cyanotic heart disease and respiratory disease -Cardiovascular and respiratory system examination normal -Chest xray normal -No signs of hypoxia (arterial blood gas analysis normal)
Final diagnosis	Points in favor
Gaisbock's syndrome	-Elevated hematocrit (55.9) -Polycythemia RBC counts- 6 lakhs/cumm -Hypertension -Elevated serum uric acid levels, -Hypercholesterolemia -Erythropoietin levels normal,JAK 2 mutation absent

DISCUSSION

Gaisbock's syndrome is a peculiar entity characterized by elevated hematocrit and hemoglobin levels in a patient without splenomegaly, leukocytosis and thrombocytosis. The syndrome was described by Gaisbock in 1905. Patient suffering from this syndrome usually had elevated blood pressure, elevated blood viscosity; plasma proteins, serum cholesterol, uric acid and increased excretion in urinary sodium [4, 5, 9].The reduction in plasma volume with relative increase in red cell count seemed related to elevation of blood pressure. In one series of 215 patients referred with a diagnosis of polycythemia vera [10], 18 (8.3%) were believed to have chronic relative erythrocytosis. Patients with relative polycythemia or erythrocytosis are usually male, obese individuals. The mean age of presentation is usually less than that of patients with primary polycythemia Vera [6].

Other features reported to be strongly associated with relative polycythemia are hypertension, smoking, alcohol abuse and occasionally chronic renal disease [11, 12].

There are two major groups of patients with relative polycythaemias. The first includes patients associated with significant degrees of dehydration due to an obvious cause like; gastrointestinal fluid losses, therapeutic diuresis, and hypercalcaemia where there is substantial extracellular fluid loss. The aetiology of the increase in haematocrit does not usually present a diagnostic challenge. The second group is associated with a chronic progressive increase in haematocrit. These patients are frequently middle-aged, mildly hypertensive, obese males with considerable stress who present with persistent polycythaemia. Characteristically, they appear plethoric but without any of the other typical features of polycythaemia vera. The

cause for the contraction in the plasma volume is poorly understood, but one school of thought explains it by autonomic dysregulation with changes in venous capacitance [13].

The usual range of haemoglobin in these individuals is between 18 and 20 g/dl with haematocrits ranging from 49 to 55 per cent. Most of these patients seek medical evaluation for an unrelated condition, and are incidentally found to have increased haemoglobin and haematocrit values [13] like in our case who presented with symptoms of elevated blood pressure.

The optimal management of relative polycythemia is unknown. As noted previously, phlebotomy increases cerebral blood flow even in patients with relative polycythemia; whether it is of symptomatic benefit is less clear. It should probably be avoided. Theoretic arguments can be made that contracting the blood volume further in these patients who already are normovolemic or slightly hypovolemic may impair tissue perfusion. Satisfactory control of hematocrit can be obtained in at least two thirds of patients by reduction of excess weight, improved hypertension control, avoidance of diuretics, and reduction if not cessation of smoking [11]. Potentially leukemogenic to reductive therapy, such as radioactive phosphorus or oral chemotherapeutic agents, is probably never indicated.

Gaisbock's syndrome is one of the rarest causes of hypertension. Our patient presented with hypertensive emergency making this case even rarer. It is important to keep this rare condition in mind while dealing with cases of polycythemia without splenomegaly and with hypertension so as to avoid unnecessary phlebotomy and other treatments of polycythemia Vera.

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