

Curious case of polycythemia vera on unenhanced CT Brain: A rare case report

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Abstract:

Sometimes diffuse hyperdense vasculature is noted on unenhanced CT Brain; one of such rare condition being Polycythemia vera. In this case report, a 20 years old male came to the hospital with chief complains of loss of consciousness, headache and convulsions. Unenhanced CT Brain was performed which revealed, diffuse hyper-density in visualized arteries, superficial venous sinuses, cortical and scalp veins of Hounsfield's Unit (HU) ranging from +60 to +68 (Contrast was not administered, HU of vessels on contrast administration ranges from +200 to +250). On blood examination the report showed raised hematocrit, red blood cells and hemoglobin levels. This showed that the hyper-density in the vasculature was due to raised hematocrit, red blood cells and hemoglobin levels, which is seen in Polycythemia vera.

Conclusion: Polycythemia vera is normally diagnosed on laboratory investigations of raised hematocrit, red blood cells and hemoglobin levels. However, it is seen that unenhanced CT Brain can also aid in the diagnosis polycythemia vera in correlation with blood examination and can also detect it as an incidental finding.

Keywords: Unenhanced CT Brain, Polycythemia vera, Case report

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I. Introduction

Polycythemia vera (PV), also known as "maladie de Vaquez," was originally identified in 1892 by French physician Louis Henri Vaquez [1].

Polycythemia vera (PV) is a myeloproliferative neoplasm defined by aberrant hematopoietic stem cell growth, which can result in life-threatening thrombotic events (1). The WHO criteria for diagnosis include hemoglobin/hematocrit levels, bone marrow biopsy results, and the presence of JAK2 mutations [2].

On non-enhanced computed tomography (NECT) images of the brain, diffuse hyperdensity affecting the Circle of Willis, dural sinuses, and other arteries is occasionally seen. This appearance can be caused by polycythemia, which is accompanied with an increase in circulating red cell mass [3,4]

The imaging results related with polycythemia vera have not been thoroughly documented in the literature. Because the majority of patients of polycythemia vera are asymptomatic until late in the disease or have ambiguous symptoms, a NECT scan for a nonspecific neurologic ailment can help with the diagnosis. We report this case due to the rarity and distinctive presentation of polycythemia vera with extensive hyperdensity of cerebral vasculature and venous sinuses on unenhanced CT brain.

II. Case Report

A 20-year-old male came to the hospital with chief complains of loss of consciousness, headache and convulsions, which were not relieved on medications. He also complained about redness of eyes and sleep deprivation. Clinical work up was done and the patient was advised unenhanced CT Brain for further evaluation.

Unenhanced CT Brain findings:

It revealed, diffuse hyper-density in the visualized arteries i.e bilateral vertebral arteries, basilar artery, arteries of circle of Willis (anterior cerebral arteries, middle cerebral arteries, posterior cerebral arteries and branches of internal carotid arteries), dural venous sinuses including transverse sinuses, internal cerebral veins, superior sagittal sinus, inferior sagittal sinus and falx cerebri. The differential includes severe dehydration; hence blood examination was performed. Possibility of polycythemia vera was to be considered, thus clinical and laboratory correlation was advised.

On blood examination:

The report revealed raised hematocrit (75.8 %), raised red blood cells ($8.5 \times 10^6 \text{ mm}^3$) and raised hemoglobin (25.4 gm/dl) levels. Rest of the report was unremarkable.

On correlation between unenhanced CT Brain findings and Blood report, diagnosis of Polycythemia vera was made.

III. Discussion

Polycythemia is a condition wherein an abnormal elevation occurs in the circulation of red blood cell (RBC) mass. It is detected as an incidental finding. In polycythemia there is increase in hemoglobin and hematocrit levels (Hematocrit is a ratio of volume of RBCs to the total volume of blood) [5].

It is seen from the literature that characterization of the neurological conditions seen in polycythemia are not common. When hematocrit level exceeds 60%, it can cause diffuse hyper-densities of dural venous sinuses and vasculature of the brain including the Circle of Willis [6]. There is a linear relationship between hemoglobin and the density of dural sinuses compared with gray matter, suggesting that increased density of cerebral vessels on unenhanced CT Brain is a sign of high hemoglobin level [7]. Flowing blood at a hematocrit of 45% measures approximately 40HU. In a normal adult, hematocrit ranges from 42-52%, thus, cerebral vasculature appears iso-dense or minimally hyper-dense as the normal adult gray matter measures approximately 25-35 HU [4]. World Health Organization's criteria for diagnosing polycythemia vera is as follows: major criteria includes hemoglobin greater than 16.5 g/dL in women and 18.5 g/dL in men or increased red cell volume such as hematocrit of at least 65% and presence of JAK2 V617F or other functionally similar mutations, such as JAK2 exon 12 mutations; and minor criteria includes bone marrow biopsy showing hypercellularity for age with trilineage growth with prominent erythroid, granulocytic, and megakaryocytic proliferation, or serum erythropoietin levels below the reference range for normal endogenous erythroid colony formation. Diagnosis requires the presence of both major criteria and one minor criterion or the presence of the first major criterion together with two minor criteria [8,9]. Other findings like Splenomegaly, organ infarcts, features of extramedullary hematopoiesis in spleen can be seen on X-ray, USG and CECT abdomen [10].

IV. Treatment

At present there is no cure for polycythemia, so the goal is to keep the hematocrit below 45% by phlebotomy, low dose aspirin and JAK 2 inhibitors [9].

V. Complications

Polycythemia can cause complications like dural venous thrombosis, splenic infarcts, superior mesenteric artery thrombosis, stroke and myocardial infarction, etc. (Attenuation of more than 70 HU is suggestive of venous sinus thrombosis and less than 70HU is suggestive of polycythemia) [11].

VI. Conclusion

Polycythemia vera is normally diagnosed on laboratory investigations of raised hematocrit, red blood cells and hemoglobin levels. However, it is seen that unenhanced CT Brain can also aid in the diagnosis polycythemia vera in correlation with blood examination and can also detect it as an incidental finding.

VII. Legends

Fig. 1: Hyper-density in bilateral Vertebral arteries

Fig. 2: Hyper-density in Basilar artery

Fig. 3: Hyper-density in bilateral MCA arteries

Fig. 4: Hyper-density in PCA arteries

Fig. 5: Hyper-density in Superior sagittal sinus and Cortical veins

Fig. 6: Hyper-density in Internal cerebral veins

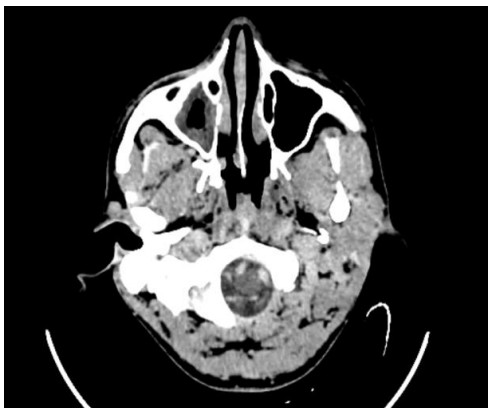


Fig. 1



Fig. 2

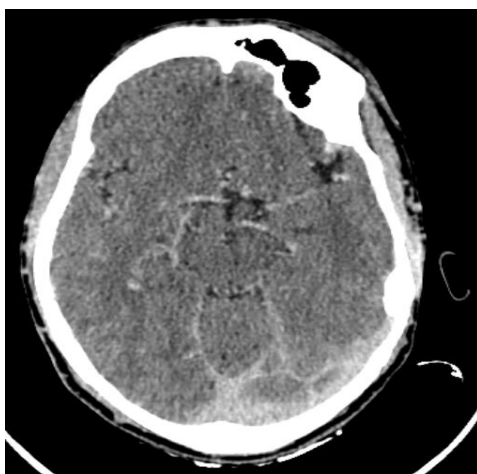


Fig. 3

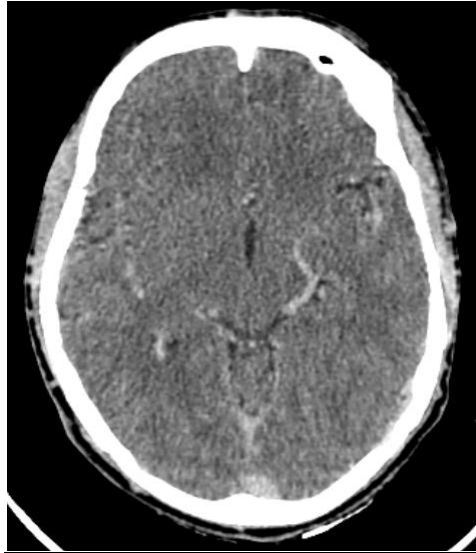


Fig. 4

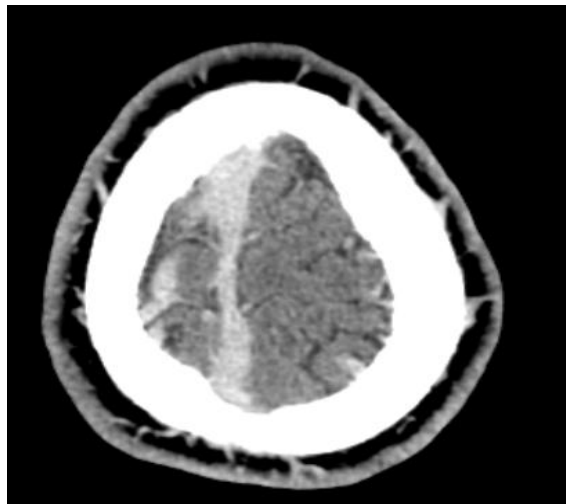


Fig. 5

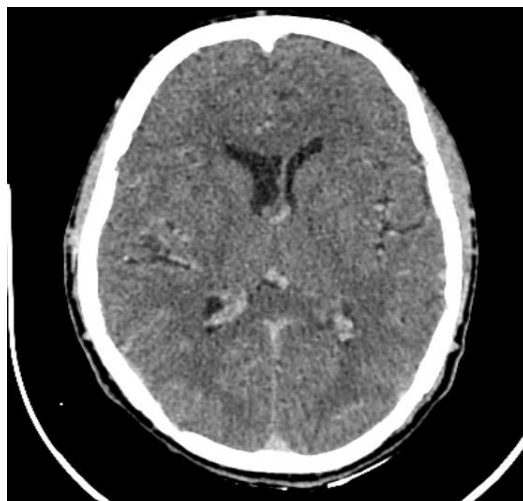


Fig. 6

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