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Central Retinal Artery Occlusion Revealing a Polycythemia Vera in a Young Adult

Boumaazisara[1*], Belghmaidi Sara[2], Hajji Ibtissam[2], Moutaouakil Abdeljalil[2]

[1]Resident of Ophthalmology, Marrakech University Hospital, Morocco
[2]Ophtalmology Professor Marrakech University Hospital, Morocco

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***Corresponding author:** Boumaazisara
Resident of Ophthalmology, Marrakech University Hospital, Morocco

Abstract **Case Report**

We report the case of yound adult for whom a central retinal artery occlusion was at the first lign of a polycythemia vera.

Keywords: CRAO, polycythemia vera, hematocrit, artery occlusion.

INTRODUCTION

Polycythemia vera is a myeloproliferative neoplasm characterized by elevated hemoglobin and Hematocrit levels with hyperleukocytosis and thrombophilia, resulting in blood hyperviscosity. Ocular involvement can rarely be revealing. We report the case of a young adult in whom a central retinal artery occlusion was at the forefront of the manifestations of the disease.

CLINICAL CASE

A 36-year-old young adult with no medical history presents to the ophtalmology emergency room fo Deep vision loss on his left eye 2 days earlier. Visual Acuity at admission was at hand motion, an affereng pupillary reflex deficit is noted. Biomicroscopic examination of the anterior segment and Tonus are strictly normal. Funduscopy is strongly suggestive of an occlusion of the central artery of the retina, there is an ischemic white edema of the retina, the retinal veins are slender while the arteries are dilated and tortuous. We note the presence of a cilioretinal artery sparing a small intermaculopapillary territory not supporting the macula. There were no visible emboli in the Retinal vasculature. The examination of the right eye was without abnormalities.

Angiography shows lengthening of retina arm time, with fractional filling of the arterial network realizing the aspect of dead tree, we noted a discreet papillary diffusion during the intermediate times not increasing during the sequence. An enlargement of the central avascular zone is noted.

There was no associated choroid ischemia. The intermaculopapillary spared territory fills up normally at different angiographic times. Cardiovascular and carotid exploration was normal, markers of inflammation were normal.

An internal medicine consultation was requested with complete immunological assessment that returned negative. The hematological assessment was disturbed with RBC at 7 million / mm3, hemoglobin 22.6 g%, Hematocrit at 61.1%; platelet rate at 143,000 mm3. The patient is referred to Clinical hematology for additional investigation. Bone marrow biopsy shows a marrow of a richness between 3 and 4 with hyperplasia of the 3 hematopoietic ligns, a densification of the supporting framework without signs of myelodysplasia or malignancy. Serum erythropoietin levels were normal.

The search for the BCR-ABL transcript came back negative, as well as the search for the JAK2 genetic mutation. The hematological caryotype did not reveal any chromosomal abnormalities.

General examination of the patient finds palmar erythrosis without organomegaly or Adenomegaly. The patient was put on aspirin at a dose of 75 mg per day with blood withdrawal at the rate of twice a week to achieve a target hematocrit level of less than 45%. The evolution was marked by a complete repermeabilization of the arterial circulation Objectified by FA, regression of

edema at the cost of retinal atrophy, without Retinal or iris neovascular complications at 6 months of evolution. On the functional plane VA remained at hand motion.

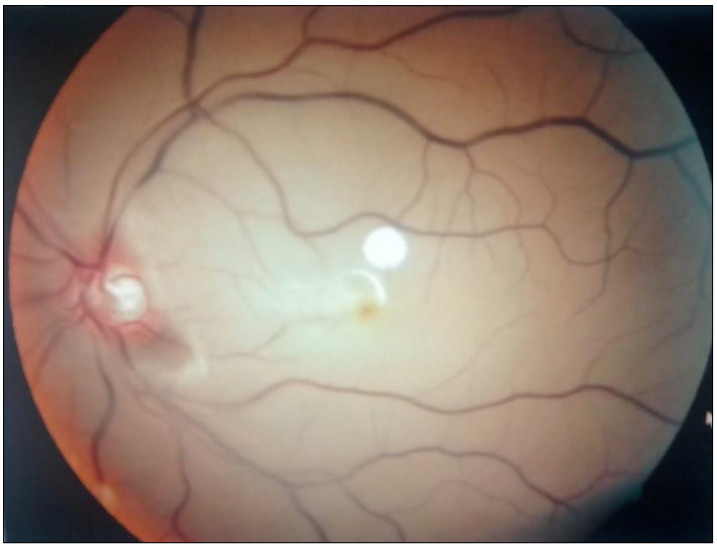

Figure: 1

DISCUSSION

Polycythemia vera is a chronic myeloproliferative neoplasia characterized by an increase in morphologically abnormal red blood cells, associated with increased Risk of bleeding and arterial or venous thrombosis.

In PV, blood hyperviscosity is the cause of a decrease in the deformability of the RBC and an alteration of its adhesion, responsible for vascular occlusions, especially in the case of JACK2 mutation. The thromboembolic phenomena of polycythemia vera are systemic and can be fatal. The classic ophthalmologic manifestations of the disease are sometimes bilateral BRVO, transient retinal vascular occlusions, and Retinal hemorrhages. Only two cases of CRAO revealing polycythemia vera have Had already been reported before our case, including a case of CRAO concomitant with stroke of the Territory of the ipsilateral middle cerebral artery in a 49-year-old subject.

PV can be life-threatening due to its cerebral and cardiovascular thrombotic complications and but also by its risk of progression to myelofibrosis or Leukemia.

In the setting of a CRAO the embolic cause must be seatrched for first, exploration Should be performed to eliminate a giant cell arteritis, the occurrence of an CRAO in a young subject should direct the etiological investigation towards systemic vasculitis.

A hematological assessment must be requested in a systematic way, it is strongly Recommended for people under 60, this assessment includes factor V Leiden, proteins C and S, antithrombin III, Anti-phospholipid antibody, homocysteine level, electrophoresis of proteins at the Search for monoclonal gammopathy in addition to the standard hematological assessment.

Ophthalmologists should have a high index of suspicion for myeloproliferative syndromes when facing a retinal vascular occlusion without local or systemic predisposing factors especially when it occurs in a young subject.

CONCLUSION

In conclusion, patients with CRAO in the absence of local Risk factor must benefit from a systemic evaluation and a complete hematological work-up which may reveal an Etiology as rare as polycythemia vera, as was the case for our patient.

The incidence of prominent ophthalmological manifestations as revealing signs made it possible monitor and prevent systemic complications of the disease.

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