SAS Journal of Medicine
Abbreviated Key Title: SAS J Med
ISSN 2454-5112
Journal homepage: https://saspublishers.com

OPEN ACCESS

Ophtalmology

Bilateral Retinal Vein Occlusion as the First Presentation of a Myelodysplastic Syndrom: A Case Report

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DOI: 10.36347/sasjm.2022.v08i04.023 | **Received:** 11.03.2022 | **Accepted:** 19.04.2022 | **Published:** 25.04.2022

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Abstract | **Case Report**

Purpose: To describe a case of myelodysplastic syndrom associated with bilateral retinal vein occlusion. ***Observations:*** A man in his 50s presented flashes and progressive drop of vision in both eyes with and feeling tired. He had no history of ophtalmic pathology. His medical history was negative for diabetes and hypertension. His initial best-corrected visual acuity was 20/200 OD and 20/25 OS. Anterior segment examination was normal. In fundus examination he had bilateral peripapillary flame-shaped and intraretinal hemorrhages in both eyes with macular hemorrage in the right eye. There was no vessel tortuosity. Fluorescein angiography of both eyes showed a good venous filling, no leakage, and no neovascularization.

Keywords: Myelodysplastic syndrom, retinal vein occlusion.

INTRODUCTION

Myelodysplastic syndrome (MDS) is an hematopoietic stem cell malignancy characterized by ineffective and dysplastic haematopoiesis leading to blood cytopenias and by a high risk of progression to acute myeloid leukaemia (AML) [1]. Ocular findings may be the first presentation of this disease and should be suspected in cases of bilateral retinal vein occlusion.

CASE PRESENTATION

A 50 years old man, with a negative medical history, presented with a progressive decrease vision in both eye OD >OS, in a context of deep asthenia. His visual acuity reduced to 20/200 OD and 20/25 OS, anterior segment examination was normal, and dilated fundus finds bilateral peripapillary hemorrhages and macular retrohyaloid hemorrhage in the right eye, without uveoretinal inlammatory manifestations. General examination finds many ecchymotic spots in the upper and lower limbs.

Fluorecein angiography shows central retinal vein in both eyes without retinal ischemia. The blood cells count finds pancytopenia, and the bone marrow biopsy shows a myelodysplastic syndrome with a high rique of acute leukemia transformation, in addition the haemostasis, hepatic and renal assessment were normal.

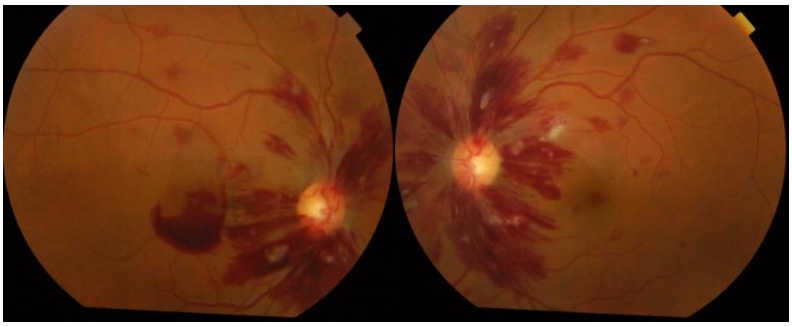

Figure 1: Central vein retinal occlusion with macular hemorrhage in the right eye

DISCUSSION

The myelodysplastic syndromes (MDS) are a disorders with variable degrees of cytopenias, morphological dysplasia and risk of progression to acute myeloid leukemia (AML). Although MDS comprises heterogeneous subcategories these share a common origin in the hematopoietic stem and progenitor cell compartmen [2].

The clinical presentation are caused by cytopenia. According to the Swedish Registry 11% and 42% of newly diagnosed patients had hemoglobin levels <8 g/dL and 8-10 g/dL, respectively, and 50% needed erythrocyte transfusions, 40% had platelet counts below $100x10^9$/L, 5% received platelet transfusions, and 20% had neutrophil counts $<0.8x10^9$/L [3].

Symptoms of anemia, such as dyspnea and fatigue, are in the first line fatigue and drop of vision as first presentation, with Bleeding complications, infectious manifestations are also frequent.

Diagnosis of MDS requires a review of a peripheral blood (PB) smear, a representative bone marrow aspirate (as well as an iron stain on the aspirate smear), and an adequate bone marrow biopsy. persistent and clinically unexplained cytopenia; significant morphologic dysplasia of hematopoietic elements ; cytogenetic and/or molecular genetic evidence of clonal hematopoiesis a detailed assessment of blood and bone marrow morphology is essential to classify the disease, and provides critical information in patient risk stratification MDS is frequently associated with a wide range of autoimmune diseases such as Sweet's syndrome, relapsing polychondritis, polymyalgia rheumatica, Sjogren's syndrome, systemic lupus erythematosus, seronegative arthritis and systemic vasculitis [4, 5].

Mekinian *et al*., [6] described the largest series to date of 123 patients with MDS-associated systemic inflammatory and autoimmune diseases (SIADs). They found the diagnoses of SIADs and MDS were concomitant in 38 (31%) patients. SIADs were classified as systemic vasculitis in 39 (32%) cases, connective tissue disease in 31 (25%) cases and inflammatory arthritis in 28 (23%) cases. Among the vasculitic diseases, polyarteritis nodosa and giant cell arteritis were most frequent. There were six cases of possible BD which were reported to have shown 'incomplete' criteria. No reference was made to the specific type of criteria applied [6]. Our patient doesn't have any auto imune disease association.

Multiple therapeutic options are available. But the only potentially curative treatment is allogeneic stem cell transplantation that necessite patient eligibility and it is associated with substantial morbidity and mortality [7, 8]. Counseling should be aimed at personalizing the decision regarding transplantation in a shared process that gives full consideration to the patient's values and wishes. Once eligibility for transplantation has been established, appropriate planning of the therapeutic program can be initiated.

CONCLUSION

Our case highlights the importance of a complete ophthalmological assessement including fundus examination in case of persistent asthenia; it can reveal potentially life-threatening diseases, such as MDS.

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