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Case Report

Roth Spots as Debut of Multiple Myeloma

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Abstract

Purpose: To report a case of bilateral Roth spots as an initial presentation of multiple myeloma in a previously healthy patient.

Observations: A middle-aged man presented to the emergency department complaining of acute visual loss on the left eye one day before the consultation developing a central scotoma. The patient denied any medical history. Fundus evaluation showed multiple Roth spots and diagnostic studies were conclusive for multiple myeloma.

Conclusions and Importance: There are several ophthalmological signs and symptoms that can lead us to life threatening conditions. Recognition of these findings can help diagnose systemic diseases earlier and improve life expectancy.

Keywords: Multiple Myeloma; Roth Spots

Introduction

Roth spots are described as white centered retinal hemorrhages. They were first observed in 1872 by Swiss physician Moritz Roth and named "Roth spots" in 1878 by Moritz Litten [1]. They were initially regarded as a pathognomonic feature of subacute bacterial endocarditis, however, nowadays, we know that they can be seen in a variety of conditions including leukemia, anemia, anoxia, carbon monoxide poisoning, prolonged intubations during anesthesia, preeclampsia, hypertensive and diabetic retinopathy, HIV retinopathy, shaken baby syndrome, intracranial hemorrhage from arteriovenous malformation or aneurism and acute reduction of intraocular pressure following trabeculectomy [1,2]. They occur in multiple diseases where retinal blood vessel injury and subsequent bleeding and hemorrhage occur. A common thread found in these conditions is a predisposition for endothelial dysfunction allowing retinal capillary disruption. The white centered lesions are composed of a fibrin-platelet plug and infiltrating red blood cells [3]

Case Report

A 42-year-old man arrived to the emergency department complaining of acute visual loss on the left eye one day before the consultation developing a central scotoma. He had no relevant medical and ophthalmological history. He had positive history for different types of cancer in his family. On ophthalmological examination, best corrected visual acuity was 20/20 in his right eye and 20/400 in his left eye. Anterior segment examination and intraocular pressure were normal. Fundoscopy showed right eye with dilatation and tortuosity of the veins and the presence of flame shaped and dot-blot intraretinal hemorrhages distributed throughout the fundus. Some white centered retinal hemorrhages were also present and were described as Roth spots. Optic nerve and foveola showed no alterations (Figure 1).

Figure 1: Right eye fundus ultra-wide field clinical photo showing flame shaped, dot-blot intraretinal hemorrhages and Roth spots distributed throughout the retina.

Figure 2: Left eye fundus ultra-wide field clinical photo showing a prerretinal hemorrhage on the macula and several Roth spots.

Fundoscopy of left eye with similar characteristics but with more abundant and confluent hemorrhages. A prerretinal hemorrhage was present on the macula, which explains the sudden visual loss of that eye (Figure 2). With these clinical findings a more specific interrogatory revealed that he had presented asthenia, diarrhea, epistaxis, unmeasured fever episodes, weight loss of eight kilograms in two months and medium effort dyspnea. According to the fundoscopic findings associated with systemic symptoms we decided to order some general blood tests witch revealed: hemoglobin 4.8 g/dL (14.5 - 17.7), globulin 11.58 gr/dL (1.9 - 3.7) and calcium 19.9 mg/dL (8.6 - 10.3). We suspect the possibility of multiple myeloma and decided to refer the patient to a hematology hospital. A complete workup was made and the laboratory testing revealed serum protein electrophoresis: gamma 7.60 gr/dL (0.6 - 1.5), serum immunofixation: IgG kappa, serum free light chains: kappa 41 mg/L (3.3 - 19.4). Urine analysis reported Bence Jones proteinuria. Beta 2 microglobulin < 0.5%, serum albumin 3.50 g/dl, fluorescence in situ hybridization (FISH) did not detect t(4:14) and was negative for deletion of the chromosome 17p13. Computed Tomography reported heterogeneous bone structures at the expense of lytic lesions in skull, axial and appendicular skeleton. Bone marrow aspirate showed 60% of plasma cells. Bone biopsy reported a hypercellular bone marrow infiltrated by neoplastic plasma cells (70%) with kappa light chain restriction. The patient was diagnosed with multiple myeloma IgG kappa. Chemotherapy with thalidomide, cyclophosphamide and dexamethasone (TaCyDex) was initiated.

Discussion

There are very few reports on the literature of Roth spots associated to multiple myeloma [4,5]. Priluck., et al. reported a unique finding on the SD-OCT of a 42 year old woman with multiple myeloma which showed Roth spots and subhyaloid hemorrhages in the macula of both eyes. They described large cystic spaces surrounding the subhyaloid hemorrhage suggesting the presence of possibly "serum" in those cystic spaces, and also identified cysts in the outer plexiform layer [4]. In this case, the patient was diagnosed with multiple myeloma, which is a neoplastic proliferation in which bone marrow elements are replaced by malignant plasma cells producing high amounts of a monoclonal immunoglobulin. Therefore, anemia, leucopenia, and thrombocytopenia may be present in advanced disease [6]. The diagnosis of multiple myeloma is often suspected because of one (or more) of the following clinical presentations: bone pain with lytic lesions discovered on routine skeletal films or other imaging modalities, an increased total serum protein concentration and/or the presence of a monoclonal protein in the urine or serum, systemic signs or symptoms suggestive of malignancy, such as unexplained anemia, hypercalcemia which is either symptomatic or discovered incidentally and acute renal failure with a bland urinalysis or rarely the nephrotic syndrome due to concurrent immunoglobulin light chain amyloidosis [7]. It is well known that some multiple myeloma patients present a condition called hyperviscosity syndrome, which is produced by an increase of blood viscosity associated to an increase in plasma components and cellular elements [8]. This produces a significant

increase in the resistance of the blood flow, producing an endothelial dysfunction allowing the vessels of the retina and other parts of the body to bleed. The most common ophthalmic manifestation of hyperviscosity syndrome is venous dilation and retinal hemorrhage, followed by central retinal vein occlusion. Other symptoms include heart failure and neurologic symptoms such as headaches, vertigo, nystagmus, deafness, ataxia, diplopia, paresthesia, stupor, or somnolence, which our patient didn't present [9].

Conclusion

This case emphasizes that multiple myeloma must be considered as a differential diagnosis in patients with Roth spots. It is important to get a multidisciplinary team involved in the treatment of these patients in order to prevent life-threatening complications that multiple myeloma may cause.

Patient Consent

The patient provided both oral and written consent for use of his medical history and images in this publication.

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Conflict of Interest

The authors have no conflicts of interest to disclose.

Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

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