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Hydroxychloroquine Induced Toxicity Presenting As Bulls Eye Maculopathy: A Case Report

Authors
Jitesh Jeswani[1], P.K. Deshpande[2], S.A. Deoke[3], R. Khirsagar[4]
[1]Junior Resident, [2]Associate Professor, [3]Associate Professor, [4]Assistant Professor

Abstract
Hydroxychloroquine is a common medication used for rheumatological inflammatory conditions. Retinal toxicity from hydroxychloroquine is rare and irreversible. We present a case of a 38 year old women , diagnosed case of Rheumatic heart disease (Mitral valve Replacement done) with Rheumatoid Arthritis , taking Tab hydroxychloroquine 200 mg BD since 2 years and developed hydroxychloroquine induced retionopathy diagnosed by evidence of Bulls eye Maculopathy . The appearance of maculopathy in such a short duration can be traced to the high doses that the patient was receiving inappropriate for her height.

Introduction

The beneficial role of antimalarial drugs in Rheumatoid arthritis, SLE and Sjogren's syndrome together with their antithrombotic and lipid decreasing action and lack of life threatening toxicity has lead to increase use of these drugs . One of the complication of hydroxychloroquine toxicity is retinal damage if taken either in a high dose - > 6.5mg/kg , prolonged use >6 years , with underlying renal or liver disease or preexsisting retinal disease.

Case Report

A 38 year old women with a BMI of 15.22 kg/m^2 , diagnosed case of Rheumatic heart disease (Mitral valve Replacement done) with Intractable seizures since the age of 7years and diagnosed to have Rheumatoid Arthritis 3 years back presented to the OPD in congestive cardiac failure with restenosis of mitral valve. She had evidence of active arthritis of bilateral wrist, small joints of hands and knee joints in the form of swelling and deformities in the form of swan neck deformity (index finger) and button hole deformity (little finger) in bilateral hands (Figure 1) which was confirmed on x ray of both hands which confirmed the diagnosis by the presence of joint space narrowing: symmetrical or concentric, marginal erosions: due to erosion by pannus of the bony , involvement of the proximal joints in a bilaterally symmetrical distribution, predilection for PIP and MCP joints (especially 2nd and 3rd MCP) boutonniere and swan neck deformities (figure 2)

Figure 1: hands showing classical deformities of RA

Figure 2: X ray both wrist and hands.

The patient was on Tab hydroxychloroquine 200 mg BD since past 2 years; her baseline fundus examination during previous admission was suggestive of subscapsular cataract. She was advised regular follow up but she did not come for workup. During present admission she complained of diminution of vision since last 6 months which was gradually progressive more in the right eye. On retinal examination by retina specialist she had evidence of a telltale sign of hydroxychloroquine toxicity characterized by bilateral change in the retinal pigment epithelium of the macula that gives the commonly described appearance of a bull's -eye Maculopathy diagnostic of Hydroxychloroquine toxicity[1]. (Figure 3), following which hydroxycholoroquine was stopped and she was started on DMARDS . Other differential diagnosis of Bulls Eye maculopathy are: Benign concentric annular dystrophy, Central areolar choroidal dystrophy, Chronic macular hole , Cone and cone-rod dystrophies .

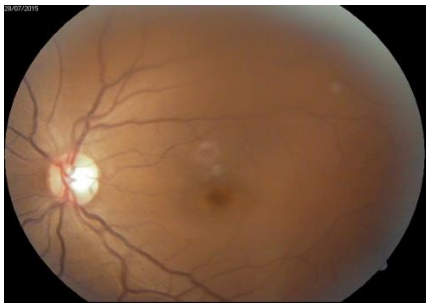

Figure 3: Retina examination showing Bulls eye maculopathy.

Discussion

Retinal toxicity from hydroxychloroquine is rare, but even if the medication is discontinued, vision loss may be irreversible and may continue to progress. It is imperative that patients and physicians are aware of and watch for this drug's ocular side effects. And before treatment is initiated with hydroxychloroquine, a complete ophthalmic examination should be performed to determine any baseline maculopathy .Patients diagnosed to have bulls eye maculopathy must undergo fundus autofluorescence (FAF), multifocal electroretinogram (mfERG) and spectral domain OCT (SD-OCT), however in this patients it could not be done.

Several factors have been associated with the risk of developing hydroxychloroquine retinopathy. One of the most important appears to be dosage with debate over whether daily intake vs. cumulative dosage is most significant. Recent studies indicate that cumulative dosage may be a more important consideration than daily dosage[2]. However, since higher daily dosage will obviously lead to the toxic cumulative dose more rapidly, daily dosage is still important to consider. Higher daily dosage also leads to higher concentration of the drug in the RPE, which could lead to more aggressive tissue damage. Previous reports indicate that toxicity is rare if dosing is less than 6.5 mg/kg/day. To avoid over dosage, especially in obese patients or those of short stature, dose should be based on height, which allows for an estimation of ideal body weight[2]. (The drug clears slowly from the blood, so basing dosage on weight puts obese patients at risk.) The typical daily dosage for most indications is 200 mg to 400 mg per day. Daily dosage is recommended not to exceed 400 mg.

Hence in this case the cause of hydroxychloroquine induced maculopathy was due to the inappropriate dose for her height.

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