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Radiation Induced Optic Neuritis: A Role for Steroids?

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Abstract

Introduction: Radiation induced optic neuropathy is a devastating complication of radiotherapy to the anterior visual pathway, resulting in significant, permanent vision loss.
Presentation: A 55-year-old male presented with 5 days of decreased vision in the right eye who had previously been treated for squamous cell carcinoma of the face. Ophthalmologic examination and MRI showed findings consistent with radiation induced optic neuropathy. The patient was treated with high dose IV corticosteroids followed by an oral taper.
Results: The patient's baseline vision in the right eye was 20/20 prior the event. On the initial presentation the patient's visual acuity had decreased to only counting fingers in the right eye. Immediately following the steroid treatment the patient's visual acuity had improved to 20/40, and two months later was measured to be 20/50.
Conclusions: High dose corticosteroids may play a role in the treatment of radiation induced optic neuropathy.

Background

Radiation of the brain or face can have devastating effects on the eye and the optic pathway. Radiation retinopathy is the most commonly seen complication, usually presenting between 6 months and 3 years following radiation treatment and is commonly monitored for in patients who have received radiation treatment in a region close to the eyes. A less commonly seen complication which can cause severe or complete vision loss in this patient group is radiation induced optic neuropathy (RION). It most commonly presents 10-20 months following treatment, although cases have been documented as much as 9 years following treatment.

Both complications have similar factors increasing the risk of occurring after treatment. The most significant of which is the cumulative doses of radiation that exceed 50 Gy or single doses to the anterior visual pathway of 10 Gy or more. These values vary significantly in the literature and cases of radiation retinopathy have been documented with as little as 15 Gy of cumulative treatment. Increased age is also a risk factor in both diseases. In the case of RION pre-existing compression of the optic nerve by the tumor has also been shown to increase the risk of developing the disease.

The diagnosis of RION is based off clinical findings as well as imaging of the optic pathway. Patients will show decreased visual acuity, visual field deficits, and decreased color vision. MRI is the imaging modality of choice showing optic nerve enhancement on T-1 or T-2 weighted images.

RION is thought to cause permanent vision loss with no proven beneficial treatment at this time. Treatment with corticosteroids and anticoagulation have generally been unsuccessful in large case series. There is some evidence of consistent improvement after early treatment with hyperbaric oxygen, but the difficulty in obtaining this treatment has limited the research in this area.

Case Presentation

A 55-year-old male presented with 5 days of decreased vision in the right eye. Two years prior to presentation, he was diagnosed with squamous cell carcinoma of the skin and treated with a cumulative radiation dose of 70 Gy. ENT and oncology were monitoring the patient for a squamous cell carcinoma of the maxillary sinus without current treatment at that time. On ophthalmologic exam, his baseline visual acuity of 20/20 had decreased to counting fingers. Dilated fundus exam (Figure 1) and fluorescein angiogram (Figure 2) showed optic disc edema and hemorrhage. MRI orbits (Figure 3) showed no interval increase in size of the squamous cell carcinoma with slight enhancement of the optic nerve and laboratory workup was unremarkable. The patient was diagnosed with radiation induced optic neuropathy. After risks and benefits were discussed with the patient, he agreed to a trial of high dose steroid therapy. The patient received three days of intravenous solumedrol (1 mg/kg/day), followed by a twelve day taper of oral prednisone (starting at 80 mg). Ten days after steroid initiation, the patient's visual acuity improved to 20/40. Two months later, his visual acuity remained improved at 20/50.

Imaging

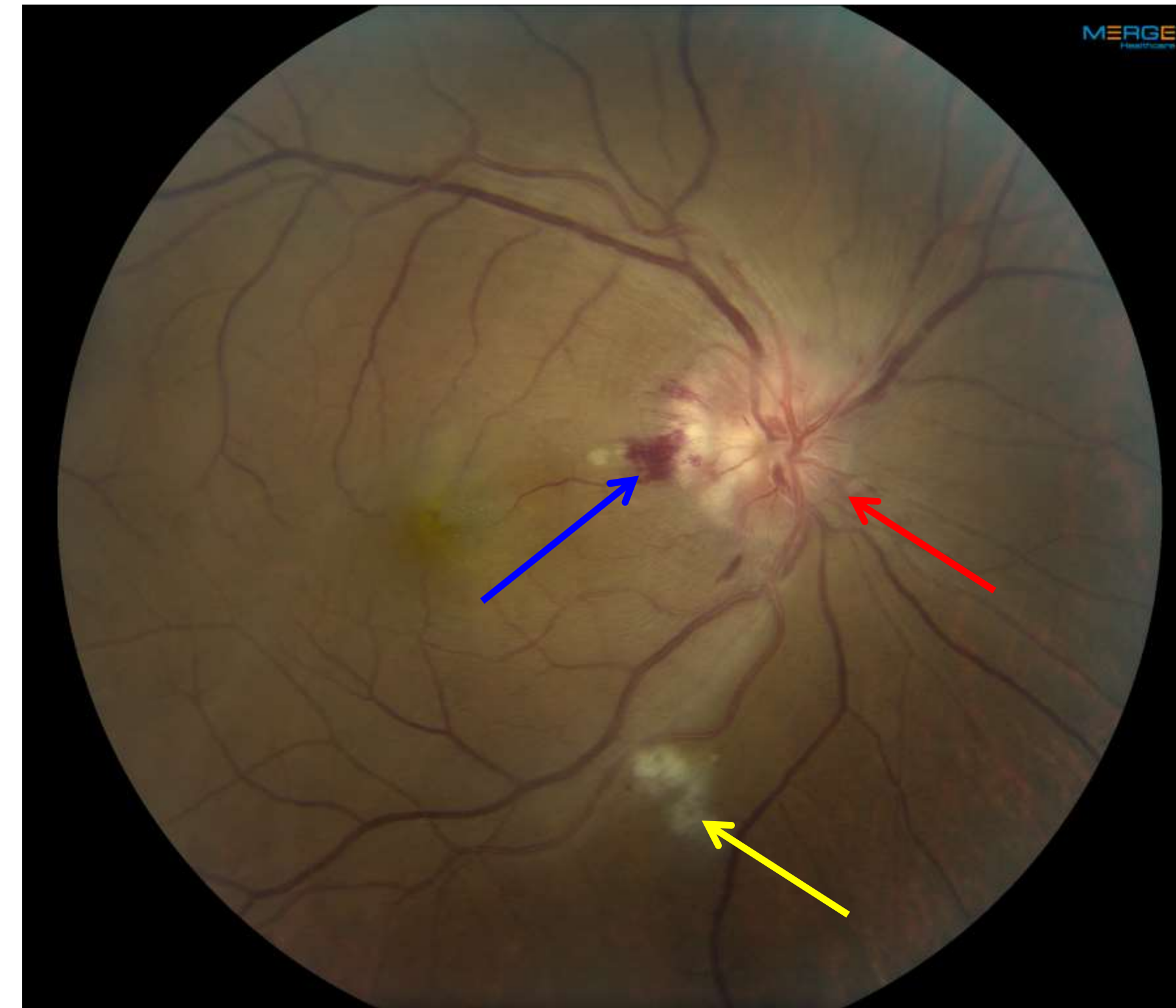


Figure 1. Fundus Photo of the right eye showing blurred disc margins consistent with disc edema (red arrow), a disk hemorrhage (blue arrow), and cotton wool spot consistent with radiation retinopathy (yellow arrow)

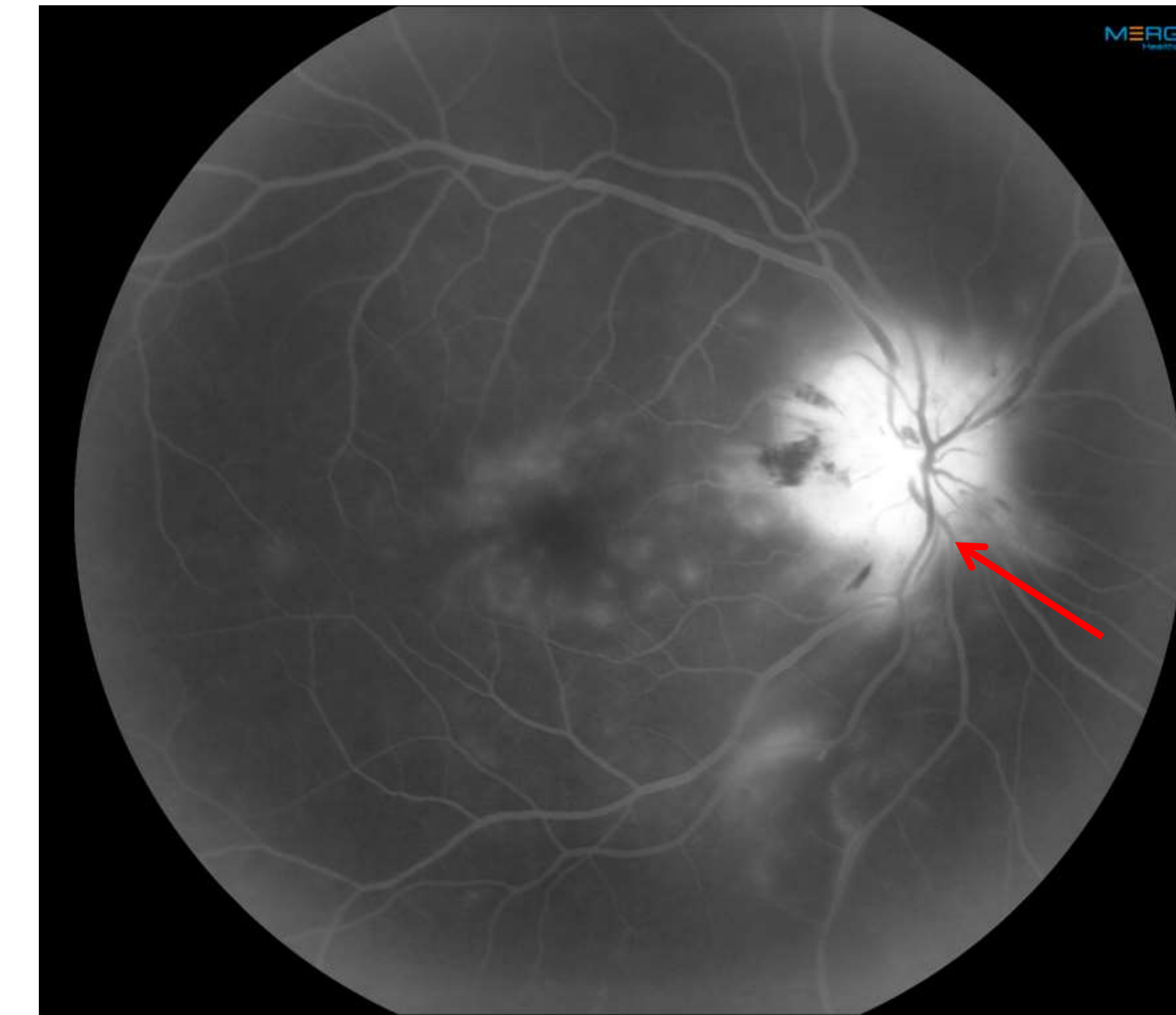


Figure 2. Fluorescein angiogram showing hyper fluorescence and leakage around the optic nerve consistent with disc edema (red arrow)

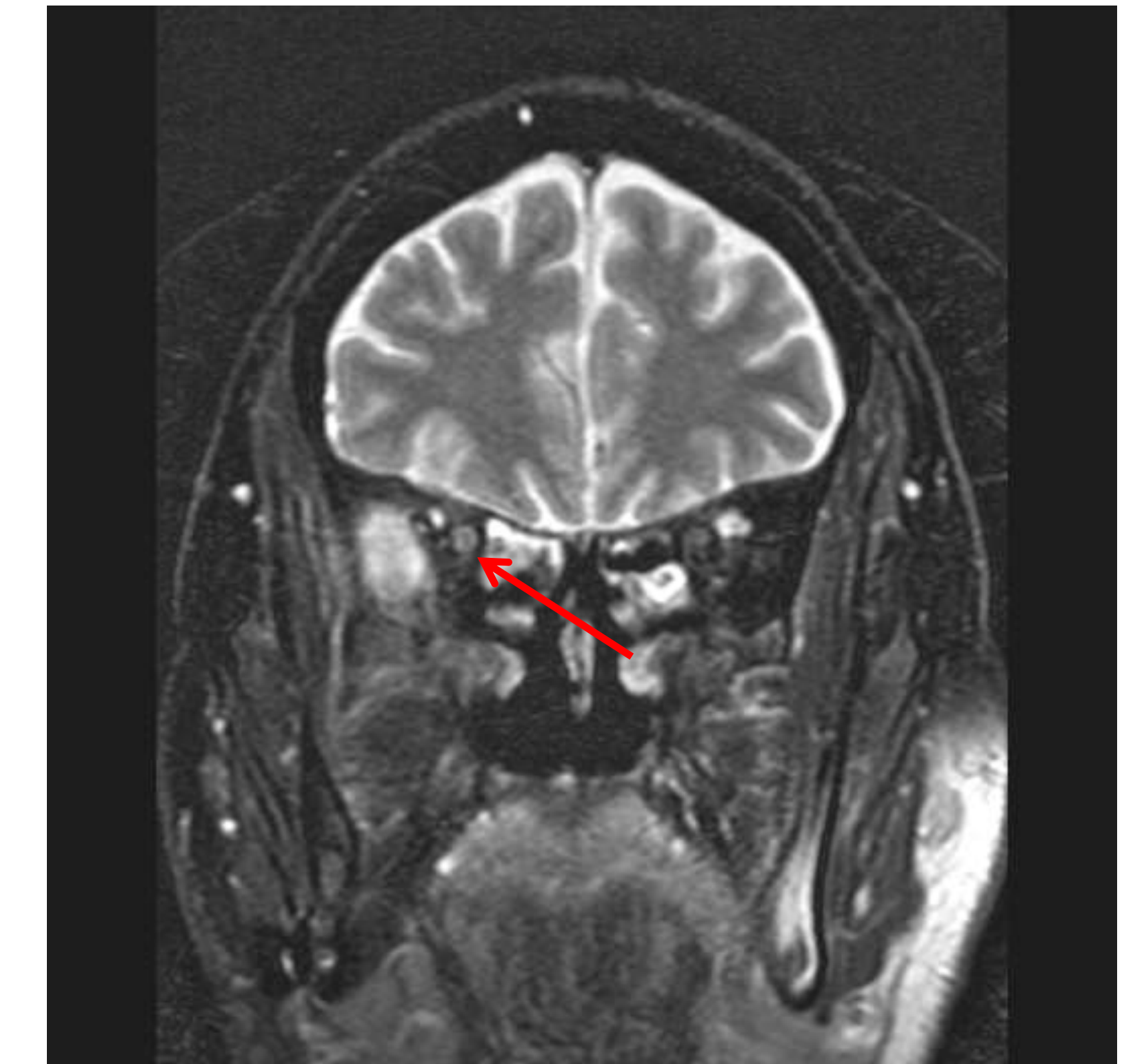


Figure 3. T2 post contrast MRI of the brain and orbits showing slight enhancement of the optic nerve within the optic canal (red arrow)

Discussion

Radiation therapy is a widely utilized therapy for treatment of many cancers of the skin of the face, the sinuses, and intracranial tumors when adequate resection is not a reasonable or possible treatment. Guided radiotherapy can limit the radiation exposure to sensitive tissues such as the retina and optic nerve, but radiation retinopathy and radiation induced optic neuropathy (RION) are complications of radiation treatment that must be discussed and monitored for following treatment. As seen with this patient, the complications may be limited as seen with his asymptomatic radiation retinopathy, but the complications may also be severe resulting in significantly decreased vision, as he presented with his RION, potentially leading to complete loss of vision. These complications commonly occur within the first two to three years as in this case, but there are documented cases of patients presenting as much as nine years after radiation treatment, highlighting the importance of long term ophthalmic monitoring.

Given that the rate of RION is relatively low, there is not a clear consensus at this time regarding a definitive treatment modality. Systemic corticosteroids, intra-vitreous anti-VEGF injections, and hyperbaric oxygen have all been trialed as potential therapies with mixed results and no modality showing consistent benefit. Systemic corticosteroids have been approached as a potential therapy given the success that has been seen using them as treatment for a cerebral radiation necrosis. However, their success has been limited in the treatment of RION and there are some small population retrospective studies showing that patients have no signs of benefit from systemic corticosteroid treatment.

Recently a study was published that performed a retrospective review of 73 patients who had received systemic corticosteroid therapy for RION. 38 of the patients had responded to treatment with the corticosteroids and 35 of the patients showed no benefit. From this patient data a predictive model was created which was able to obtain a high degree of accuracy in determining which patients could benefit from treatment. Elevated BUN was found to be significantly related to a greater response to treatment. The patient in this case had a BUN of 21 at the time of presentation.

The current understanding of RION is limited given its relatively low prevalence. Given that there is no clear intervention that has been shown to halt or reverse the vision loss in affected patients, further research in this area with larger prospective studies is needed. Although some studies have dismissed systemic corticosteroids as a potential treatment for RION, as seen with this patient there may be a subset that significantly benefit from early treatment with high dose corticosteroids.

Conclusions

- RION is a devastating complication of radiation treatment to the brain or face
- Patients who have need radiation treatment to the face or brain should be warned of ocular complications and possible decreased or loss of vision
- There is not a widely accepted treatment at this time, although systemic steroids, anti-VEGF injections, and hyperbaric oxygen are all being trialed at this time
- As seen with this patient there appears to be a subset of patients that can significantly benefit from treatment with high dose corticosteroids
- Further evaluation of recently developed predictive models may allow stratification of patients into those that may benefit from treatment with corticosteroids
- Early intervention after presenting symptoms appears to be crucial as seen in this case and as documented in the literature
- Treatment with high dose corticosteroids should be considered in all patients presenting with RION

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