Optic nerve sheath meningioma with globe invasion

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Case Report

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ABSTRACT (40 words)

An 82-year-old female presented with painless and progressive visual loss of right eye for two years. Ophthalmological examination revealed choroidal detachment in her right eye. Orbita and brain magnetic resonance imaging showed right optic nerve sheath meningioma with globe invasion.

KEYWORDS: Optic nerve sheath meningioma, choroidal detachment, globe invasion, magnetic resonance imaging, ultrasonography

INTRODUCTION

Optic nerve sheath meningiomas (ONSMs) are histopathologically benign tumors. ONSMs grow slowly but they can cause complete vision loss without treatment. ONSMs are usually unilateral and originate from arachnoid cells of the optic nerve sheath. They can be detected from orbit to intracranial space.

ONSMs usually occur in adult women and clinical presentation include progressive visual loss, proptosis, color blindness, and relative afferent pupil defect. In the early stage, papilledema may be seen and it may result in optic atrophy. Mortality risk of ONSMs are null.

Radiological findings are very useful to detect ONSM. First choice scan technique is magnetic resonance imaging (MRI). In MRI; tubular enlargement of the optic nerve, enlarged optic canal, and calcification within the tumor can be seen. [1]

In this case, we aimed to present an ONSM patient with intraocular extension and choroidal detachment.

CASE REPORT

The patient was informed about the study and was invited to be a part of it. The patient was also instructed that participation was totally voluntary and not participating in this study would have any negative effect on her treatment and relationship with their physicians. Before initiating the study, written consent was obtained from the patient. The study conformed to the tenets of the Declaration of Helsinki.

An 82-year-old Turkish female patient presented with a two-year history of progressive visual loss in her right eye. She gave a history of diabetes mellitus, hypertension, and coronary bypass surgery 3 months ago. Her best-corrected visual acuity on the Snellen chart was 20/400 in the right eye and 10/20 in the left eye. There was a right relative afferent pupillary defect. Bilateral nuclear cataract grade two was detected in anterior segment examination. The intraocular pressure on Goldmann applanation tonometry was 14mmHg on both eyes. There was no proptosis and restriction of movement in all directions of gaze. A dilated fundus examination of the right eye revealed retinal elevation (Figure 1). In her left eye, chorioretinal atrophy was detected. Orbital ultrasonography examination was performed. The choroidal detachment was detected and there was subretinal tumor suspect (Figure 2). An MRI (gadolinium contrast) scan of the brain and orbits was performed and post-contrast T1-weighted axial MRI with fat

saturation and the axial T1-fat suppressed without gadolinium contrast MRI scan showed diffuse enlargement and contrast enhancement of the right intraorbital optic nerve sheath, and intrabulbar extension was detected (Figure 3, Figure 4, Figure 5, and Figure 6). In the coronal T2 scan, we detected hyperintense lesion which was around the optic nerve (Figure 5). The coronal T1 weighted fat suppressed with gadolinium contrast MRI scan showed us intense contrast-enhancing lesion around the optic nerve (Figure 6). The patient was referred to the neurosurgery department with a prediagnosis of right ONSM. They confirmed the diagnosis and planned surgery but the surgical intervention was not a good choice because of the age and cardiovascular disease. Patient took a total dose of 50 Gy fractionated stereotactic radiotherapy over the course of 8 weeks.

DISCUSSION

ONSMs constitute about 2% of all orbital tumors and 1-2% of all meningiomas. [2] After gliomas, ONSMs are the second most common optic nerve tumor. Middle-aged women are the most affected population in the world but ONSMs may occur in childhood especially who have Neurofibromatosis type 2. [3] The typical clinical symptom is progressive visual loss. The tumor grows around the optic nerve and makes compression so clinical findings are usually seen in optic disc. Clinical findings include optic nerve head edema, optic atrophy, optociliary collateral vessels, relative afferent pupillary defect, dyschromatopsia, and proptosis.

Last decades, MRI (especially with gadolinium-enhanced fat-suppression sequences) has become a gold standard scanning technique for diagnosis of ONSMs. The other scanning technique is high- resolution computed tomography. Nowadays, the most popular treatment method is radiotherapy especially in early and progressing stage. Some authors suggest surgery If tumors invade to the intracranial space. [4,5] The other treatment options are surgery and radiotherapy and simply follow-up observation.

Our case is atypical because she had an intrabulbar extension and choroidal detachment. ONSM may apply mechanical pressure to the choroid. Also, ONSM may compress the retinal and choroidal veins. When the these veins are chronically compressed and venous pressure in the eye rises. Venous stasis occur. These conditions may cause ONSM choroidal detachment. In our case, it is well seen the orbital involvement in the axial MR image. We thought that the reason for the choroidal detachment was an orbital invasion. In the literature, the intraocular extension of the ONSMs are very rare. In a review of 5000 orbital meningiomas, Dutton reported that intraocular extension rate was 3.8%.[6] He detected that tumor may invade optic disc, sclera, choroid, and retina. Schittkowski *et al*. presented a patient with an intraocular spread of ONSM in 1999.[7] Some authors described the choroidal invasion. Schatz *et al*. reported a patient who had ONSM and choroidal neovascular membrane(CNVM) but this patient had age-related macular degeneration in both eyes. [8] Tirkey *et al*. reported that similar case who had ONSM and CNVM but they said that there was no predisposing factor. [9] Sekeryapan *et al*. showed choroidal folds in patient with

ONSM and they thought that the choroidal fold formation was probably due to stretching of the optic nerve. [10] In our patient, we detected choroidal detachment. Because of the intraocular invasion which is seen in MRI, we thought that the reason for the choroidal detachment was ONSM.

CONCLUSION

ONSM should be considered in a patient with progressive visual loss and intraocular invasion possibility of the ONSM should be kept in mind.

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Authors Contributions:

SC: design of the work, acquisition, analysis, interpretation of data, drafting and, writing of the manuscript

CS: writing of the manuscript and acquisition

TS: writing of the manuscript and acquisition

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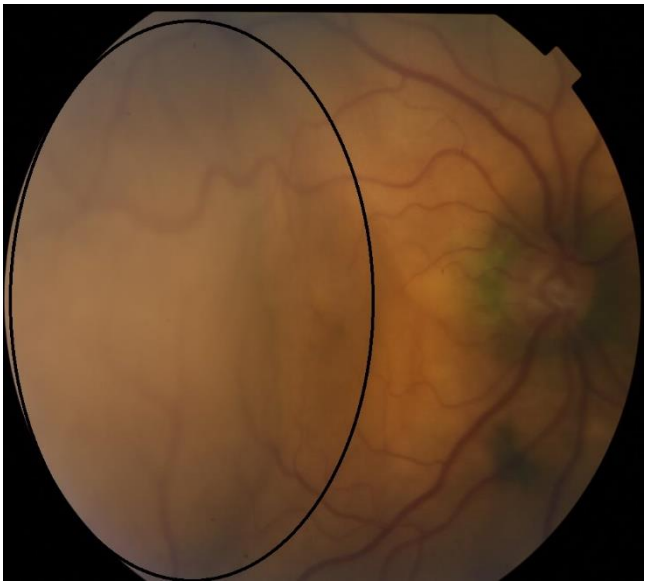

Figure 1: Choroidal detachment in the right eye (black circle)

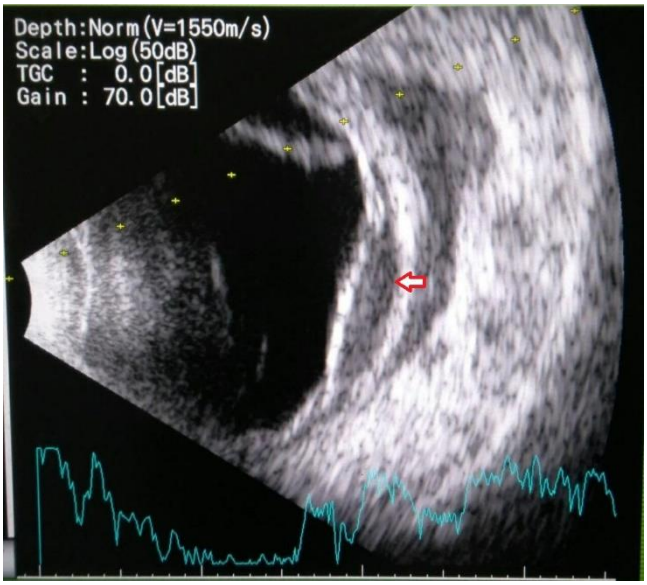

Figure 2: Orbita Ultrasonography (red arrow shows choroidal detachment)

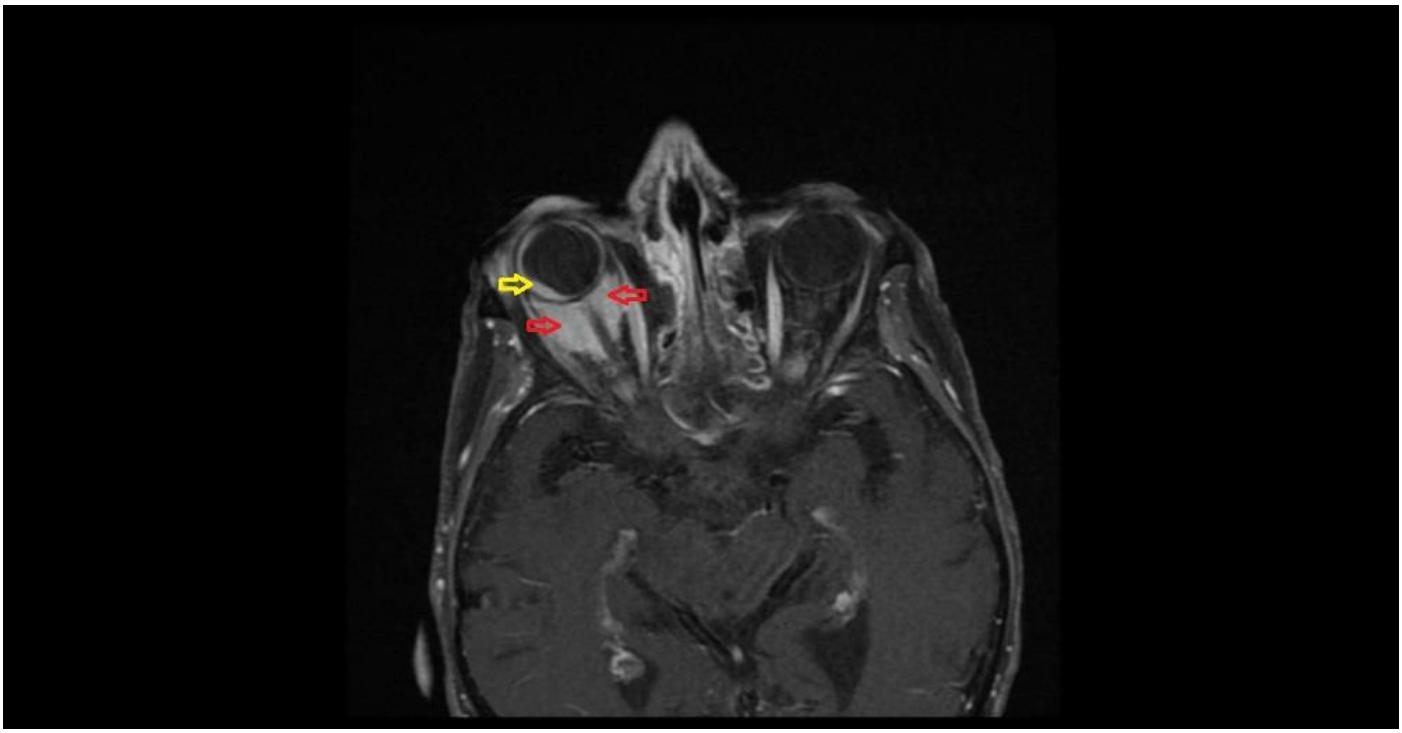

Figure 3: The axial T1-weighted fat suppressed with gadolinium contrast MRI scan of optic nerve sheath meningioma (red arrow) and globe invasion (yellow arrow)

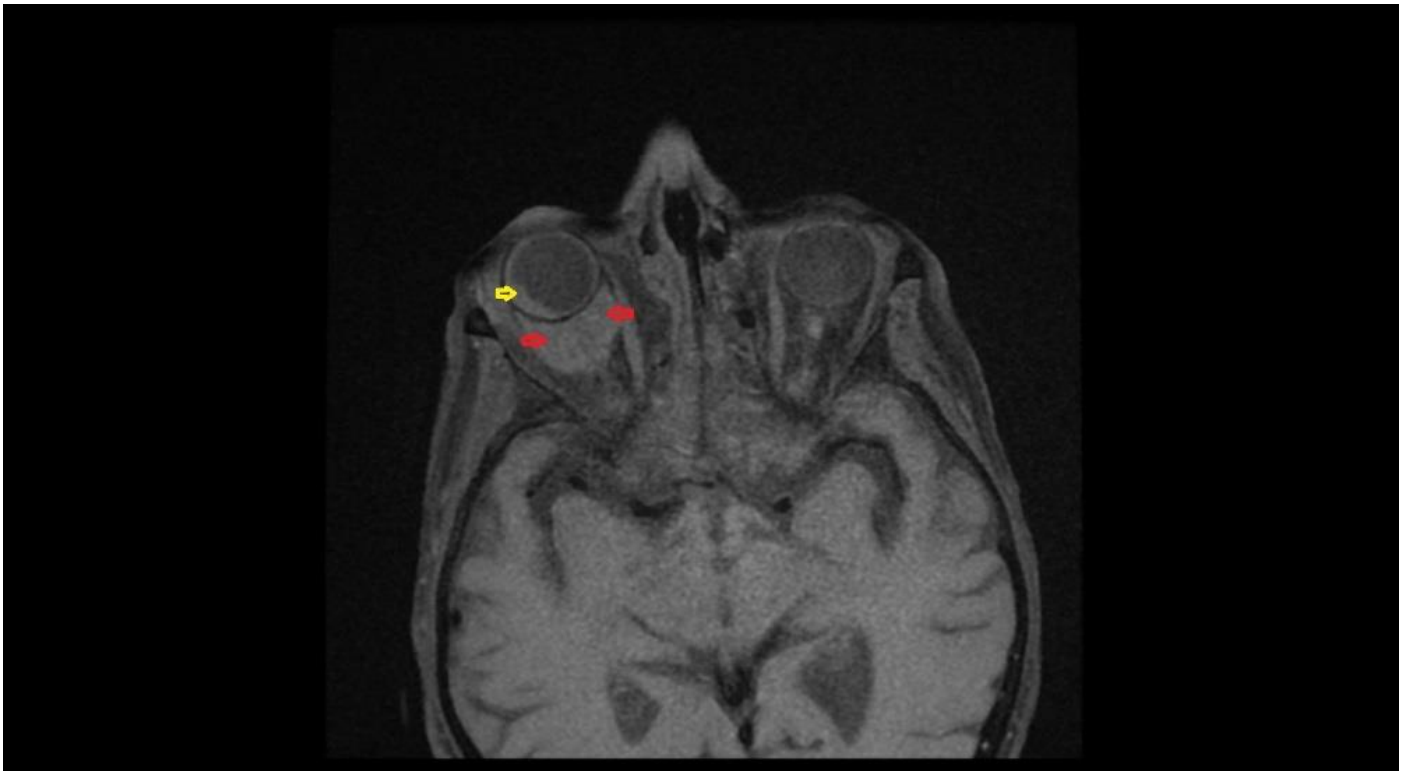

Figure 4: The axial T1-fat suppressed without gadolinium contrast MRI scan of optic nerve sheath meningioma (red arrow) and globe invasion (yellow arrow)

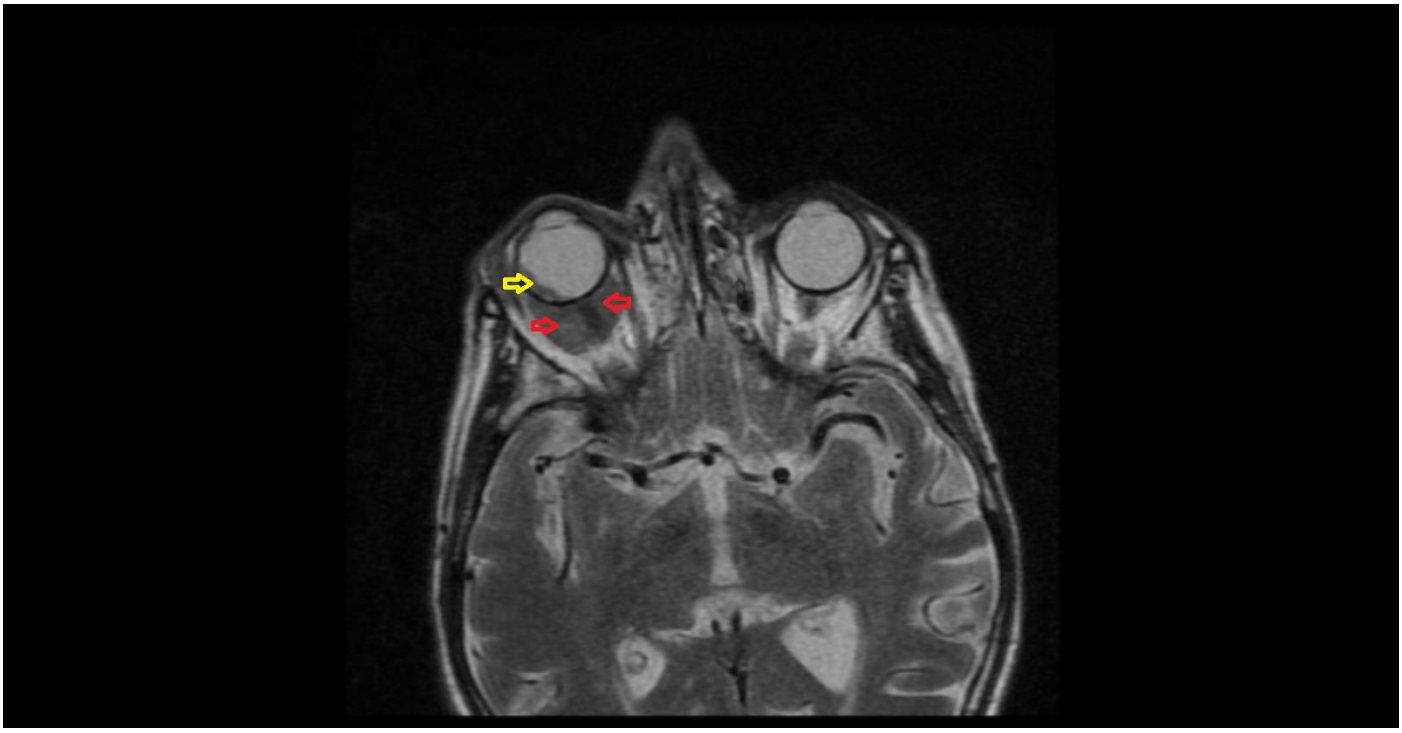

Figure 5: The axial T2 MRI scan of optic nerve sheath meningioma (red arrow) and globe invasion (yellow arrow)

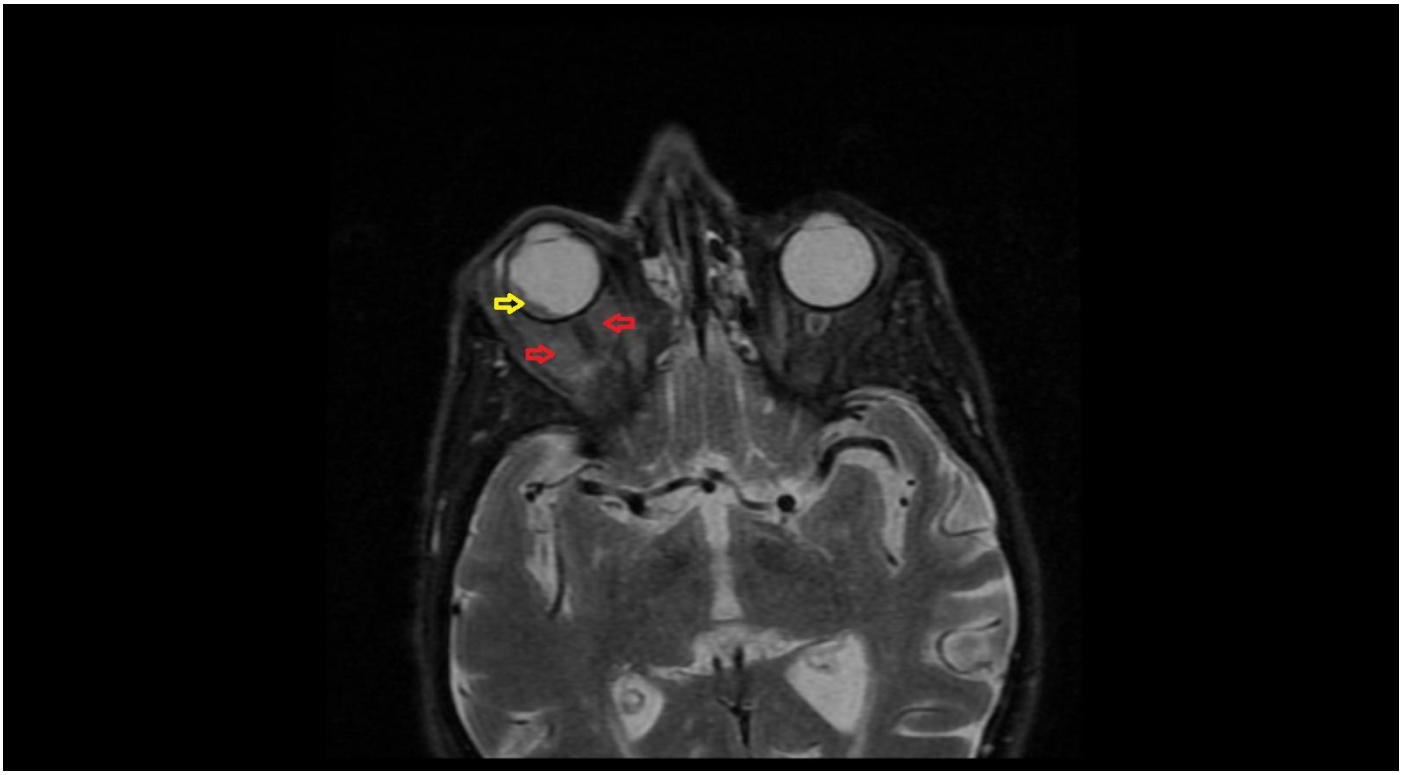

Figure 6: The axial T2-weighted fat suppressed MRI scan of optic nerve sheath meningioma (red arrow) and globe invasion (yellow arrow)

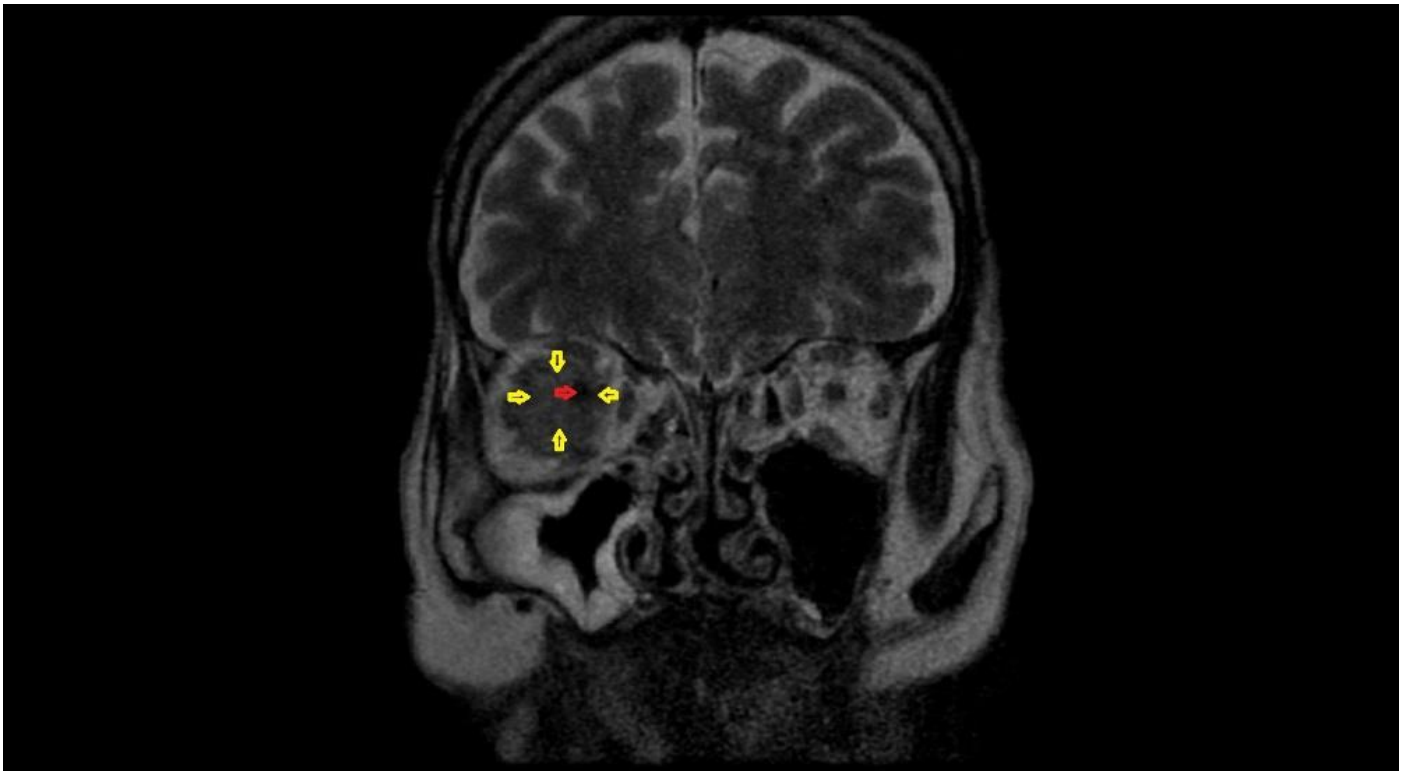

Figure 7: The coronal T2 MRI scan of optic nerve sheath meningioma (yellow arrow) and optic nerve (red arrow)

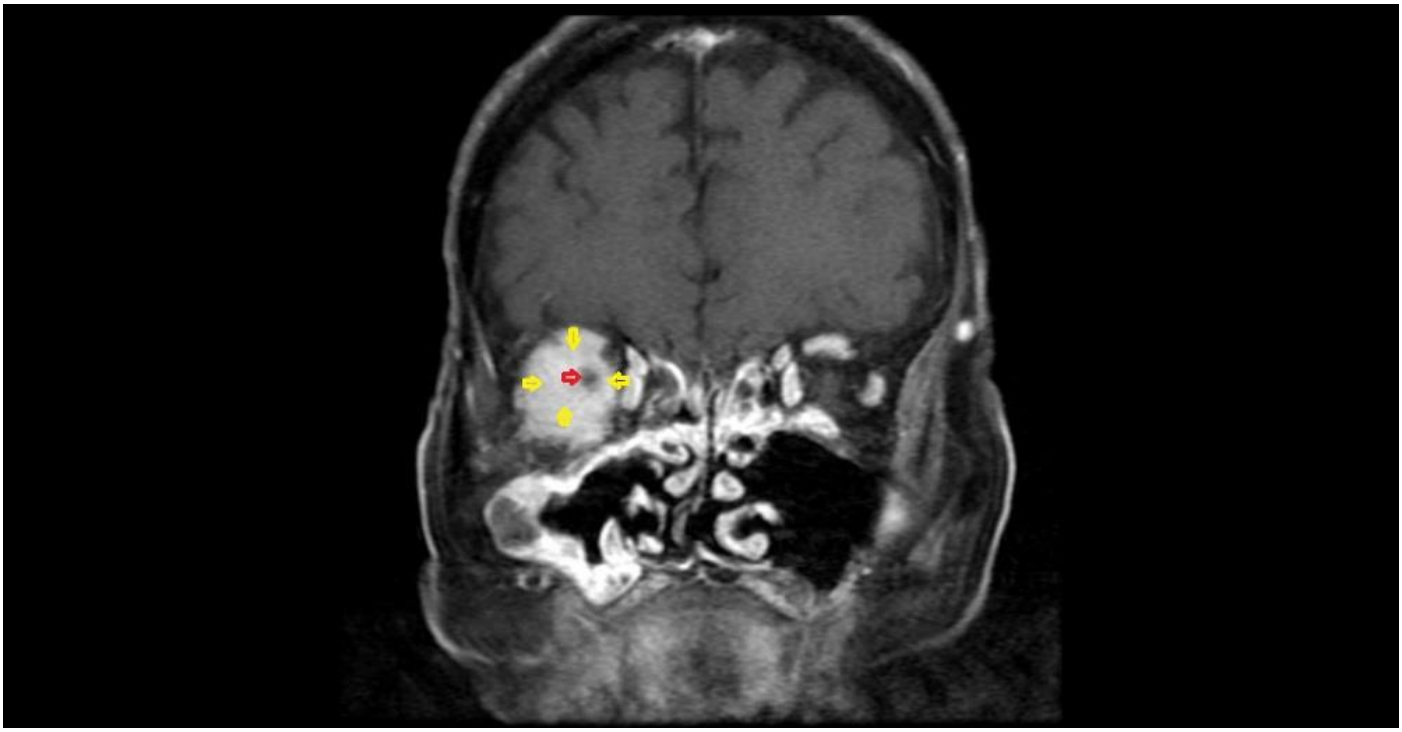

Figure 8: The coronal T1 weighted fat suppressed with gadolinium contrast MRI scan of optic nerve sheath meningioma (yellow arrow) and optic nerve (red arrow)