

## Case Report

# Delayed-onset seesaw nystagmus following brain irradiation

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## Summary

Seesaw nystagmus (SSN) is a rare, debilitating neurologic syndrome with few known etiologies. Typically, patients present with oscillopsia subsequent to a recent neurologic insult, with only a handful of patients show-

## Introduction

Seesaw nystagmus (SSN), first reported by Maddox in 1913,<sup>1</sup> is a rare form of nystagmus characterized by abnormal alternating eye movements. Simultaneously one eye elevates and intorts while the opposite eye depresses and extorts. The underlying mechanism of SSN is not fully understood. The theoretical pathogenesis involves a mass lesion or vascular accident disturbing the medial mesencephalic reticular formation, the interstitial nucleus of Cajal (INC) including the area of the rostral interstitial nucleus of the medial longitudinal fasciculus, and the posterior commissure.<sup>2</sup> The dorsal midbrain contains these structures, which are important in the control of vertical eye movements.<sup>2</sup> The INC is involved in the ocular tilt reaction. The medial longitudinal fasciculus serves as the generator of vertical saccades and receives information from the vestibular nuclei, which it projects to both the oculomotor and trochlear nuclei ipsilaterally and contralaterally via the posterior commissure.<sup>2</sup> The waveform of this type of nystagmus can either be pendular (seesaw) or jerk (hemi-seesaw).<sup>2</sup> Jerk SSN is associated with lesions in the region of the INC; pendular SSN is associated with parasellar or chiasmal mass lesions.<sup>3</sup>

To date, the documented causes of seesaw nystagmus include parasellar or mesodiencephalic masses or disease, stroke, trauma, multiple sclerosis, congenital abnormalities (eg, Arnold Chiari malformation), and severe vision loss from cone-rod dystrophies or retinitis pigmentosa, as well as radiation and chemotherapy.<sup>3,4</sup> We present a case of seesaw nystagmus in a patient with

ing delayed onset after trauma. We report a case of seesaw nystagmus in a patient with a large meso-diencephalic mass treated with external beam radiation therapy (XRT), who developed asymptomatic SSN nearly 8 years after treatment.

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## Case Report

In 2003, a 27-year-old Cape Verdean man presented at Boston Medical Center and was diagnosed with a non-germinomatous germ cell tumor of the pineal gland metastatic to the pituitary gland and ependymal lining (Figure 1); at the time he denied any ophthalmic history or complaints. Five years after diagnosis he was treated with XRT, for a total dose of 50 Gy. The earliest data available is from 2013, at which point he was followed by our neurology service. Initial examination was significant for a normal cranial nerve (II-XII) examination, no nystagmus, and bitemporal homonymous hemianopsia on confrontational visual field testing. He was subsequently lost to follow-up.

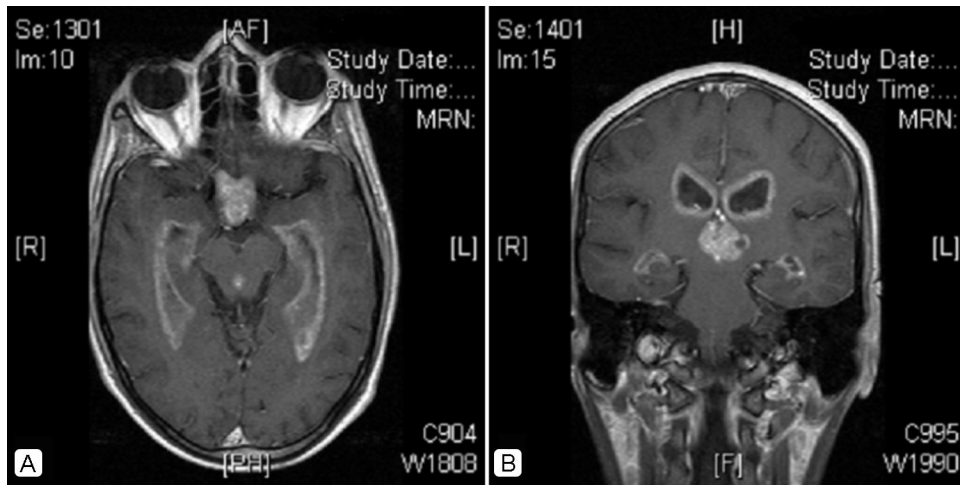
In 2014, he completed his first general ophthalmic examination. At this time, he expressed a gradual subjective decline in vision. His best-corrected visual acuity was counting fingers at 3 feet in the right eye and 20/50 in the left eye. He was again noted to have bitemporal hemianopsia on confrontational visual field testing, rotary nystagmus, ocular hypertension, and bilateral optic nerve pallor. Magnetic resonance imaging of the brain and orbits, with and without gadolinium, did not show tumor recurrence or enhancement at this time. The orbits were unremarkable. On follow-up examination at

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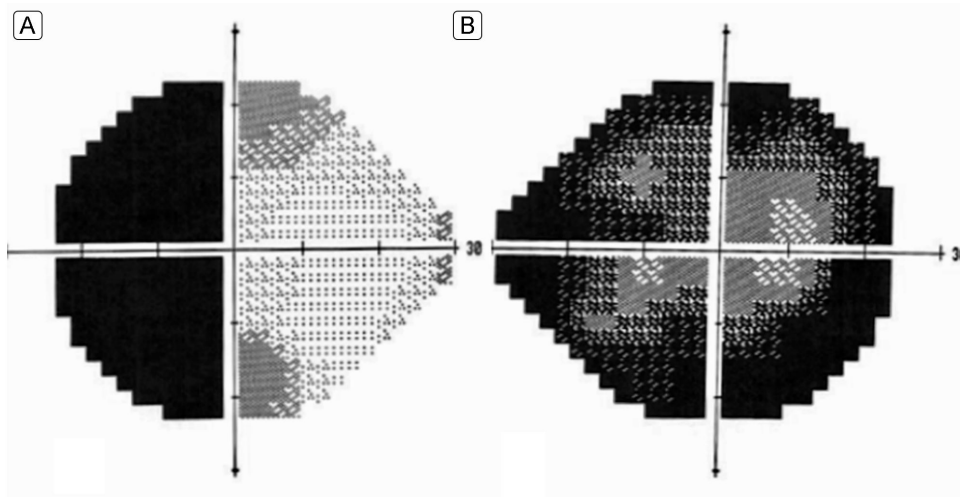
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**Figure 1.** T1-weighted magnetic resonance imaging of the head with gadolinium: axial (A) and coronal (B) views depicting a solid mass in the parasellar region.



**Figure 2.** Humphrey visual fields demonstrating temporal hemianopic loss in the left eye (A) and diffuse loss in the right eye (B).

the neurology service 3 years later, he was again noted to have bilateral rotatory nystagmus.

In 2018, his nystagmus was described as including a vertical component along with the previously noted rotary movements, resembling pendular SSN. At this time he completed a formal Humphrey visual fields test (Figure 2). The patient was referred to neuro-ophthalmology several times but was unable to attend his appointments until 2019. Neuro-ophthalmic evaluation revealed best-corrected visual acuity of 20/400 in the right eye and 20/80 in the left eye. Examination was otherwise stable, with confirmed pendular SSN (see Video 1). Despite his significant SSN, the patient denied any



**Video 1.** Patient with seesaw nystagmus. Examination showing pendular waveforms, with one eye elevating and intorting while the other depresses and extorts; the seesaw nystagmus is seen in all cardinal directions of gaze.

oscillopsia. He has remained in remission to date, with serial MRIs conferring no recurrence.

## Discussion

A lesion of the meso-diencephalon, similar to our patient, is the most common etiology of SSN, although usually with an acute/subacute time to onset. To our knowledge, only 1 prior case of SSN after XRT has been reported in the literature. Epstein et al presented a patient with diffuse large-cell lymphoma, treated with methotrexate and XRT, without an identifiable brainstem lesion.<sup>3</sup> During therapy she developed progressive vision loss and ultimately SSN.<sup>3</sup> Their case supports the theory that brainstem disease may not be a prerequisite to SSN, similar to cases related to retinal diseases like cone-rod dystrophy.<sup>3,5</sup> They also proposed that methotrexate and XRT exposure are possible etiologies of SSN.

There have only been 3 reported cases of delayed-onset seesaw nystagmus in the literature. Eggenberger described 2 of the 3 cases: one patient who developed SSN 21 years after trauma, with imaging demonstrating bilateral frontal encephalomalacia without chiasmal discontinuity, and the other patient who developed SSN with encephalomalacia of the left subfrontal and left anterior temporal lobes 37 years after trauma.<sup>1</sup> The mechanism of delayed-onset nystagmus is not yet completely understood.

Our patient developed SSN nearly 8 years after XRT therapy. Although there are cases that purportedly only associate vision decline with SSN, those patients were found with more severe vision loss compared to our patient, to the level of hand motion in both eyes.<sup>4</sup> Furthermore, despite his severe SSN, our patient remained asymptomatic, without oscillopsia. To our knowledge,

there has only been 1 reported case of asymptomatic SSN.<sup>6</sup>

Our case further supports XRT as a possible etiology for SSN and expands on the limited cases available for investigation of this rare phenomenon. Further investigation is required to explore both the mechanism of delayed-onset SSN as well as a possible neurorestructuring that may allow for adaptation to this movement disorder, preventing a patient from having to endure debilitating symptoms of oscillopsia.

## Literature Search

PubMed was searched in September 2019, without date restriction, for English-language results, using the following terms: *seesaw nystagmus*, *delayed-onset seesaw nystagmus*, *asymptomatic nystagmus*, and *nystagmus after irradiation*. The reference section of each publication was reviewed for additional sources.

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