

OCULAR MANIFESTATIONS OF NARCOLEPSY*†‡

BY

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THE narcoleptic tetrad consists of narcolepsy (irresistible sleep or sustained drowsiness), loss of muscle tone, sleep paralysis, and hypnogogic or hypnopompic hallucinations.§ Patients may have all four components or may have only narcolepsy, the feeling of irresistible sleep or sustained drowsiness. It is by no means rare, 500 cases being seen in 5 years at the Mayo Clinic (Yoss and Daly, 1957), and ophthalmologists should therefore be aware of the ocular complaints. The onset is usually in the second or third decade and males may slightly predominate. The pathogenesis and theories have been described by various workers (Kennedy, 1929; Daniels, 1934; Levin, 1943; Yoss and Daly, 1957; Keefe, Yoss, Martens, and Daly, 1960; Goode, 1962; Dale and Langworthy, 1964). Briefly, the prevailing theories are a conditioned Pavlovian response or a lesion in the reticular activating system.

Diplopia is a common, but often overlooked symptom, and may be the only early manifestation. Kennedy (1929) reported the case of a male who had diplopia at age 22, sleep paralysis beginning at age 25, and cataplexy beginning at age 26. Patients may present with complaints of "weak eyes", "eye strain", or of their eyes "tiring easily" because they must "rest their eyes" after reading for short periods. They may note difficulty in staying awake while driving and may visit an ophthalmologist for a refraction hoping that this will alleviate the problem.

Case Report

A 23-year-old white male medical student first complained of blurring of vision becoming frank diplopia in September, 1965. These attacks occurred usually in the morning and during lectures but could be controlled by a conscious effort. However, shortly after the first occurrences, the patient noted an ever-increasing desire to sleep immediately after the diplopia. At no time was there any numbness or paralysis, nor were there hypnogogic or hypnopompic hallucinations, sleep paralysis, or cataplexy. He denied severe headaches; there was no history of encephalitis.

As the attacks were occurring once or twice a day, usually in class, he was seen in the health office and was referred to the ophthalmology clinic in December, 1965. No diagnosis was made and no medication was given, but because of his history of diplopia and blurring while reading (though diplopia first began on looking at distant objects) convergence exercises were prescribed. These had little effect and he was seen in the ophthalmology clinic again in February, 1966, when his phorias and vergences were found to be extremely variable, even at the same sitting.

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§ Hypnopompic and hypnogogic refer to that state just before complete awakening (hypnopompic) or complete sleep (hypnogogic). Sleep paralysis is that state in which a patient on awakening is fully conscious but cannot move.

Examination

REFRACTION: Right -5.25 D sph., +1.25 D cyl., axis 90°
 Left -5.75 D sph., +0.5 D cyl., axis 90°

VISUAL ACUITY (with glasses): Right 20/10—2
 Left 20/15

HEAD POSTURE: Normal

NEAR POINT OF CONVERGENCE: 0-60 mm.

EXTRA-OCULAR MOVEMENTS: Full versions and ductions

PHORIAS (Prisms, Maddox rod) with correction:

Distance 3-15 prisms ×

Near 0-04 prisms × LH 1-2 prisms (Maddox ring)

On prolonged cover: ×4 prisms ×'16 prisms LH ½ to 1 prisms, without correction
 ×1½ prisms ×'7 prisms No hyper (Maddox rod) with correction

VERGENCE AMPLITUDES (Break point, prisms):

	B1	B0
Distance	4-6 prisms	12-40 prisms
Near	6 prisms	12-40 prisms

Wirt stereoscopic vectographs: All correct (40°)

The patient was then seen in the department of neurology and the diagnosis of narcolepsy was confirmed by Dr. P. Scheinberg. 10 mg. methylphenidate hydrochloride (Ritalin) twice daily was prescribed but because of agitation while on this dosage the patient began to regulate himself and found that he did relatively well on 10 mg. daily, suffering only occasional attacks.

He was discharged with instructions to report periodically for follow-up.

Discussion

Diplopia has been found as a symptom of narcolepsy by Daniels (1934), Levin (1943), Yoss and Daly (1957), Keefe and others (1960), and Dale and Langworthy (1964), either with or without catalepsy. Keefe and others (1960) found that ten of 25 patients had intermittent diplopia and a further nine of that same 25 had blurring of vision when reading.

It is reported that a conscious effort to stay awake usually results in diplopia, but in the present case, a conscious effort to fuse tended to cause more sleepiness. Levin believed that it was analogous to a generalized paralysis. Daniels (1934) reported five cases of diplopia before falling asleep and others noted it as a common symptom questioning narcoleptics.

A few patients have been diagnosed as cases of multiple sclerosis, myasthenia gravis, or hypothyroidism (Yoss and Daly, 1957; Keefe and others, 1960; Dale and Langworthy, 1964), but when a symptom appears that may be due to sustained drowsiness one should suspect narcolepsy.

The criteria for diagnosis (Yoss and Daly, 1957) show that sudden precipitous sleep is not the usual symptom, a sustained drowsiness being more commonly the rule. Since analeptic drugs have come available, it is important to diagnose the condition and begin treatment to relieve or at least modify this previously non-treatable illness.

The extreme variability in measurements at the same sitting may cause the ophthalmologist to suspect malingering, but these variations are due to the variability of a narcoleptic's physical status at any given time. Keefe and others (1960) reported that patients with phoria greater than 8 prism dioptres usually have a tendency to diplopia while those with less than 8 prisms of phoria tend to have blurred vision. Measure-

ments in such cases, however, may be of little value. Most patients appear to improve on therapy but this too is hard to evaluate because of the variability in their physical status.

Summary

The case is presented of a 23-year-old white male medical student who presented with intermittent diplopia followed by an urge to sleep. Correct diagnosis of narcolepsy enables treatment to be given. The extreme variability in eye muscle measurements is noted, with a brief discussion of points of interest to ophthalmologists and orthoptists.

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