

Case study

NARCOLEPSY AND AERONAUTICAL APTITUDE

Abstract:

Narcolepsy is a rare chronic neurological hypersomnia, differentiated into two independent nosological entities: type 1 and type 2 narcolepsy. Through a clinical observation, the authors recall the diagnostic criteria of narcolepsy and the role of the aeronautical physician in the screening of this pathology.

Keywords : narcolepsy, excessive daytimesleepiness, aeronautical expertise

INTRODUCTION

Narcolepsy is a rare chronic neurological hypersomnia characterized by excessive daytime sleepiness with uncontrollable sleep episodes, inconsistently associated with cataplexy and other symptoms considered minor (1, 2). Its repercussions on vigilance and its psychological impact make this pathology incompatible with aeronautical activity.

CASE PRESENTATION:

Mr. M A, 45 years, presents to the center of medical expertise for aircrew (CEMPN) in Rabat for a flight engineer admission medical examination. He worked as an aircraft ground mechanic for 18 years. During the interrogation, he declared that he had been followed in neurology for hypersomnia for 10 years.

The onset of his illness dates back 15 years with complaints of chronic helmet headaches that interfered little with activity without nausea, vomiting or visual disturbances. Our mechanic is referred to a neurology consultation where the doctor notes a yawn in the sitting position, then falling asleep in the lying position during the clinical exam.

The interrogation a posteriori rules out sleep deprivation, drug intake, stigmata of sleep apnea syndrome or depressive syndrome and finds the notion of incoercible sleep accesses even in a stressful situation with access of generalized cataplexy provoked by laughter. Furthermore, there is no notion of sleep paralysis, hypnagogic or hypnopompic hallucinations.

An additional assessment is requested, in particular a cerebral MRI which proved to be normal (no hypothalamic cerebral lesion). Polysomnography (PSG) shows

shortened REM sleep latency with sleep onset latency on Multiple Sleep Latency Test (MSLT) of 6 minutes and 2 falling asleep in REM sleep.

The diagnosis of narcolepsy type 1 is made and Modafinil-based treatment is started in combination with sleep hygiene measures (avoidance of sleep debt, caffeine consumption and scheduled naps)

The evolution is marked by the disappearance of the cataplexy after 3 years and the patient has since been lost to sight. During this period, Modafinil is taken irregularly and then stopped spontaneously (side effects such as insomnia persist despite the reduction in dosage).

During his flight engineer assessment, he reported stabilization of his illness for 7 years by sleep hygiene measures alone.

The Maintenance of Wakefulness test (MWT) finds normal average sleep onset latency; his file is presented to the derogatory commission of the Royal Air Forces which declares him fit by derogation with restrictions.

DISCUSSION

Reminder of narcolepsy

The 3rd International Classification of Sleep Disorders (ICSD-3, AASM 2014), differentiated narcolepsy into two independent nosological entities: narcolepsy type 1 (NT1) and narcolepsy type 2 (NT2).

• **narcolepsy type 1 (NT1)** called narcolepsy with cataplexy or "Gélineau's disease" in the old nomenclature is a rare (20 to 30 cases per 100,000 subjects) (3) and disabling affection in young subjects, the age of onset of the disease is distributed around two peaks around 15 and 30 years (2).

Its pathophysiology is based on the loss of neurons producing hypocretin/orexin in the posterior hypothalamus secondary to an autoimmune process with the absence of hypocretin 1 in the cerebrospinal fluid. An association with the HLA DQB1*06:02 typing is very frequent (> 92%) but not very specific since it is present in 20% of the general population (excellent negative predictive value) (4).

NT1 is characterized by two major symptoms:

- **Excessive daytime sleepiness** is often the initial symptom with uncontrollable episodes of sleep several times a day, brief and often restoring a normal level of

wakefulness for a few hours. Hypersomnolence occurs preferentially in monotonous situations or relative inactivity (2).

- **Cataplexy** is the most specific sign of NT1; it corresponds to a brief sudden loss of muscle tone in full consciousness triggered by emotions, especially positive ones such as laughter, surprise. It can be either generalized causing the collapse of the subject, or localized affecting only the muscles of the neck, face or limbs, defining partial cataplexy (3-5).

Other symptoms are often associated but not specific (2) such as

- **sleep paralysis** corresponding to the persistence of REM sleep atony on waking, when the subject is already conscious. They are usually very brief (a few seconds) but stressful (6)

- **hypnagogic** (when falling asleep) and **hypnopompic** (on awakening) hallucinations: are visual, auditory and sometimes somesthetic hallucinations, at sleep/wake time transitions, which readily accompany episodes of sleep paralysis.

- Poor quality sleep (dyssomnia) with very rapid falling asleep but frequent nocturnal awakenings and parasomnia.

- Weight gain may be observed (especially in children), Periodic limb movement disorder (PLMD) and parasomnias; depression is a common comorbidity (6).

• **Narcolepsy type 2 (NT2)** shares the same clinical symptoms as narcolepsy type 1 except for the presence of cataplexy with the same neurophysiological criteria. Nighttime sleep is often less fragmented and weight gain less frequent. Its prevalence and pathophysiology remains poorly understood and are the subject of controversy (7).

The diagnosis of narcolepsy combines clinical, polysomnographic (PSG, MSLT) and biological signs (3).

The 3rd edition of the International Classification of Sleep Disorders (ICSD-3) defines NT1 by the presence of the following criteria:

The presence of somnolence for more than 3 months, associated with:

- either the presence of cataplexy, an average sleep onset latency at (MSLT) of less than 8 minutes and at least 2 falling asleep in REM sleep at MSLT and/or during nocturnal PSG, corresponding to the onset of REM sleep within 15 minutes of falling asleep
- Or a hypocretin level of less than 110 pg/mL in the cerebrospinal fluid.

The diagnosis of narcolepsy type 1 is a definite positive diagnosis.

NT2 is defined in ICSD-3 by the presence of sleepiness for more than 3 months

- In a patient without cataplexy;
 - having an average sleep onset latency at MSLT of less than 8 minutes with at least 2 falling asleep in REM sleep at MSLT and/or nocturnal PSG;
 - The level of hypocretin > 110 pg/mL or not measured in the cerebrospinal fluid;
- The hypersomnolence cannot be explained by another cause such as sleep deprivation, sleep apnea syndrome, circadian rhythm disorders, or the effect of a drug or substance.

The diagnosis of NT2 is therefore a diagnosis of elimination.

This hypersomnolence is sometimes transient and may be the cause of significant phenotypic variations. Some patients initially diagnosed with narcolepsy without cataplexy may, sometime after the initial assessment (several months or years), present the diagnostic criteria for idiopathic hypersomnia, no longer have any criteria for hypersomnia, or even develop cataplexy (3); The diagnosis of NT2 is therefore not definitive, requiring reassessment during follow-up.

The treatment of narcolepsy types 1 and 2 is currently symptomatic, it associates:

- behavioral measures: regular sleep hygiene, anticipated naps of short duration < 15 min;
- psychological support if needed;
- Adapted drug treatment to any associated comorbidities (cardiovascular, metabolic, endocrine, neuropsychiatric, sleep disorders). These are molecules that act :
 - on vigilance (modafinil, methylphenidate, pitolisant..)
 - on cataplexy: antidepressants, preferably stimulants ones (fluoxetine, venlafaxine, etc.) or sodium oxybate also having a favorable effect on dyssomnia, sleep inertia and daytime alertness. (6)

narcolepsy and aeromedical aptitude

In aeronautical activity, somnolence is the main human factor involved in the occurrence of air accidents (8). The aeronautical doctor must be attentive to the symptoms that may lead to the suspicion of a pathology responsible for excessive

daytime sleepiness (EDS) such as narcolepsy which remains insufficiently advanced (3).

During medical visits, the interrogation is an essential step in the expertise. The medical expert must be able to detect, by careful questioning, unrecognized daytime sleepiness (case of our aspiring flight engineer) or omitted to be declared voluntarily or not.

Many symptoms can suspect the diagnosis of narcolepsy.

- Severe daily and recurrent drowsiness in a passive situation, but also in a more active situation
- An irrepressible need to sleep with difficulty fighting against this need.
- Several naps or sleep episodes during the day, short, refreshing,
- Generalized or partial loss of muscle tone in response to positive emotions (such as laughter, surprise) indicating the presence of cataplexy.

Suspicion of excessive daytime sleepiness in aircrew justifies a polysomnographic recording possibly followed by a MSLT in the context of temporary unfitness.

On the aeronautical aptitude plan, narcolepsy is a chronic neurological pathology belonging to the group of rare hypersomnolences of central origin. There are no civil or military regulatory texts dealing with this pathology in particular, however, its occurrence in aircrew is synonymous with unfitness given the increased risk of air accidents and the uncertain evolutionary potential of the narcolepsy type 2.

However, the case of our flight engineer candidate is special; he has been followed for type 1 narcolepsy for 10 years with clinical stabilization for 7 years without psychological repercussions.

The stabilization of the disease by behavioral measures alone with the absence of objective somnolence confirmed by the maintenance of wakefulness test (MWT) which objectifies the absence of sleep at 40 minutes in non-pilot aircrew (no high aeronautical risk function) motivated, justifies the presentation of his file to the appeal authority (Royal Air Force Derogation Commission) which grants him fitness by derogation in medical standards with restrictions, in particular no external operations, exemption from guards with regular quarterly clinical monitoring by the unit's doctor using Epworth sleepiness scale (figure 1) and annually at the neurophysiology center (systematic MWT) and at the center of medical expertise for aircrew.

CONCLUSION

Narcolepsy is a rare neurological pathology responsible for excessive daytime sleepiness and thus threatening flight safety. The role of the aeronautical doctor is mainly to detect this condition by careful questioning, supplemented if necessary by a

polysomnographic recording and by Multiple Sleep Latency Test. Rehabilitation remains exceptional, depending on the aeronautical function and the absence of objective sleepiness on the Maintenance of Wakefulness Test.

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How Sleepy Are You?

How likely are you to doze off or fall asleep in the following situations? You should rate your chances of dozing off, not just feeling tired. Even if you have not done some of these things recently try to determine how they would have affected you. For each situation, decide whether or not you would have:

- No chance of dozing =0
- Slight chance of dozing =1
- Moderate chance of dozing =2
- High chance of dozing =3

Write down the number corresponding to your choice in the right hand column. Total your score below.

Situation	Chance of Dozing
Sitting and reading	•
Watching TV	•
Sitting inactive in a public place (e.g., a theater or a meeting)	•
As a passenger in a car for an hour without a break	•
Lying down to rest in the afternoon when circumstances permit	•
Sitting and talking to someone	•
Sitting quietly after a lunch without alcohol	•
In a car, while stopped for a few minutes in traffic	•

Total Score =

Analyze Your Score

Interpretation:

0-7: It is unlikely that you are abnormally sleepy.

8-9: You have an average amount of daytime sleepiness.

10-15: You may be excessively sleepy depending on the situation. You may want to consider seeking medical attention.

16-24: You are excessively sleepy and should consider seeking medical attention.

Figure1: Epworth sleepiness scale

The total score can range from 0 to 24, and values above 10 are considered abnormal, indicating excessive sleepiness (9)

UNDER PEER REVIEW