Complete remission of narcolepsy after surgical treatment of an arachnoid cyst in the cerebellopontine angle

Narcolepsy (excessive daytime sleepiness with cataplexy) has rarely been associated with intracranial or extracranial brain lesions. Such lesions have predominately been in the brainstem or diencephalon but their functional relation with the mechanism of narcolepsy remains unclear. Recent evidence confirming a strong linkage between narcolepsy and some phenotypes of HLA in the Japanese population has not yet been related to a detailed account of the brainstem disorder responsible.

Intracranial extraintracranial or extracranial lesions with symptomatic narcolepsy are extremely rare. The present case is unique in describing reversible narcolepsy in a patient with a left cerebellopontine angle arachnoid cyst in whom surgical decompressive treatment of the cyst has induced complete remission of all symptoms.

A 24 year old woman rapidly developed excessive daytime sleepiness when 14 years of age, with repeated attacks several times a day without any other narcoleptic symptoms. At that time, she was disturbed by frequent quarrels between the parents. Her parents divorced and she was 18 years old. While she was a high school student (16–18 years old), other narcoleptic symptoms became apparent. She had frequent oppressive presleep dreams, such as the fall of an aeroplane on to her head (hynagogic hallucination). She often felt paralysed during the presleep stage (sleep paralysis). She never complained of disturbed nocturnal sleep, even during periods when excessive sleepiness occurred, often during the daytime. A sudden change of emotion—for example, when startled by a loud sound or in a rage against her brother—caused transient motor paralysis, and sometimes she dropped down on to her knees (cataplexy). She finished high school and was employed by a city bank where her job was often interrupted by excessive daytime sleepiness. In August 1992 she consulted our department and CT showed a large cyst in the cerebellopontine angle. Magnetic resonance imaging (T1; figure) confirmed a cerebellopontine angle arachnoid cyst with shift of the brainstem, which prompted her immediate admission. General physical examination was normal except that she was obese (75 kg for 157 cm height). Neurological examination was normal. A multiple sleep latency test was not performed and an EEG examination during the daytime showed bilateral alpha rhythm, alpha blocking by eye opening, and paradoxical alpha blocking during stages 1 and 2 of sleep; REM sleep was not seen, nor any presleep symptoms. Auditory brainstem response showed normal latency on both sides. Cisternography with contrast medium given by lumbar puncture showed non-filling of the cyst after three hours. Blood analysis was normal. She was HLA-DR2 positive. Cerebral angiography showed no anomaly in the vertebrobasilar system other than an avascular mass lesion in the left cerebellopontine angle.

An operation performed in September 1992 at the age of 22 years was to marsupialise the arachnoid cyst as widely as possible into the surrounding cisterns. The arachnoid membrane was hypertrophied, and the cisterns were adherent.

Within four weeks of the operation, the narcoleptic symptoms completely disappeared. After two months, she returned to her job and has been asymptomatic since.

A few cases of reversible symptomatic narcolepsy have been reported. Matsuda et al described a patient with ischemic infarction in the upper ventral brainstem who developed hypersomnia and cataplexy after treatment with sodium valproate and phenytoin. These two symptoms disappeared after discontinuation of the antiepileptic drugs. Onofrj et al reported hypersomnia and cataplexy in a case of left midtemporal primary B-cell lymphoma. The radiotherapy and immunosuppressive treatment induced complete disappearance of the symptoms and regression of the tumour. Presumably, lymphoma cells might have infiltrated the brainstem. Rubinstein et al reported a case of CNS sarcoidosis presenting with hypersomnia. Computed tomography showed an 8 mm round area of contrast enhancement located in the hypothalamus. Whole brain irradiation resulted in dramatic clinical improvement with disappearance of the hypothalamic lesion on CT. These three cases relate the narcoleptic symptoms to destructive lesions in the brainstem or diencephalon. The complete brainstem natal cerebrospinal fluid has not been mentioned in any of these reports.

Symptomatic narcolepsy secondary to cerebellopontine angle extra-axial mass lesions such as acoustic neuroma, meningioma, etc, has not been reported to our knowledge. This arachnoid cyst in the cerebellopontine angle region was a benign mass that distorted the brainstem. Brainstem dysfunction of ischaemic origin might have caused the symptomatology in the present case. This could explain the reversible narcolepsy except that there were no other brainstem symptoms or signs.

Several functional networks in the brainstem have been postulated to play a part in the genesis of REM sleep. More than one region may be responsible for the genesis of narcolepsy. Possibly individual variation of the brainstem vascular system might be involved in ischaemic processes for the incidental initiation and development of the narcoleptic tetrad in this unique subject.

Hence, distortion of the brainstem seems to be able to cause the same narcoleptic tetrad as the idiopathic type (which is believed to be irreversible), and the genetically predisposed group, suggesting that a pathological process in the brainstem is common to them all.

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