Chapter 12

Senile Dementia of the Neurofibrillary Tangle Type

CQ 12-1

What kind of disease is senile dementia of the neurofibrillary tangle type (SD-NFT)?

Answer

SD-NFT is a subtype of senile dementia characterized by numerous neurofibrillary tangles (NFTs) mainly in the hippocampus but very few senile plaques. This disease is often diagnosed clinically as Alzheimer's disease dementia.

Comments and evidence

1. Concept

SD-NFT is a subtype of senile dementia characterized by the presence of numerous NFTs mainly in the hippocampus, and the incidence increases with aging. SD-NFT is reported to constitute 1.7-5.6% of autopsy cases of dementia in older people and account for 20% of dementia cases in those over 90 years of age [1]. Among the dementia cases in the Hisayama study, 2.9% was classified as SD-NFT, and the percentage was 4.9% when limited to autopsy cases [2]. On the other hand, the term "primary age-related tauopathy (PART)" has been proposed as a pathological term for the condition in which NFTs are chiefly distributed in the medial temporal lobe while senile plaques are absent or very scarce [3, 4]. Clinically, this is perceived as a concept of spectrum including the states from normal cognitive function to dementia through mild cognitive impairment.

2. Clinical features

SD-NFT is considered to be a pathological condition of accelerated aging process of the brain. The clinical features are as follows [1, 3, 5]:

(1) Common in the late old age
(2) Progresses slowly
(3) Memory impairment as onset symptom
(4) Other cognitive impairments and personality changes are relatively mild
(5) Rare appearance of delirium and extrapyramidal symptoms
(6) Imaging study shows atrophy of hippocampal region and expansion of inferior horn of lateral ventricle.

3. Differential diagnosis

Many cases of SD-NFT are diagnosed during life as Alzheimer's disease dementia, and many pathological findings of SD-NFT overlap with those of argyrophilic grain dementia and vascular lesions.

Although SD-NFT shares many common clinical features with late-onset Alzheimer's disease dementia, such as memory impairment as the core symptom and lesions enhanced in the medial temporal lobe, SD-NFT is a more slowly progressive disease, and amyloid PET is useful for differentiation.

Both argyrophilic grain dementia and SD-NFT are classified as late-onset tauopathy, and the two share common features including lesion in the medial temporal lobe and memory impairment as onset symptom. However, in argyrophilic grain dementia, although memory impairment is the main feature, other symptoms such as irritability, behavioral abnormalities, and personality change are a characteristic feature and useful for differentiation. In addition, the medial temporal lobe atrophy in argyrophilic grain dementia is asymmetric, which is also a differentiation feature. The medial temporal lobe atrophy is anterior-dominant in argyrophilic grain dementia but is relatively prominent at the posterior side in SD-NFT . These features are helpful in differential diagnosis [1, 3, 5].

4. Treatment

In clinical practice, SD-NFT is often diagnosed as Alzheimer's disease dementia and treated with cholinesterase inhibitors. There are currently no therapies that have proven efficacy for SD-NFT [1, 3, 5].

References

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