

Witzelsucht after Right Putaminal Hemorrhage: A Case Report

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Abstract- Witzelsucht is a tendency to tell inappropriate and poor jokes. It usually occurs after a focal lesion involving orbitofrontal cortical or paramedian thalamic regions, especially on the right side. Here we report a 56-year-old man developing witzelsucht and hypersexuality after a right putaminal hemorrhage. The hematoma extended to the sublenticular part of posterior internal capsule and mesencephalon. The hemorrhage might have disconnected the fibers in the ascending reticular systems, and the fibers between paramedian thalamus and orbitofrontal cortex, and thus could be responsible for the patient's rare clinical manifestations.

Key Words: Humor, Hypersexuality, Orbitofrontal lobe, Paramedian thalamus, Putaminal hemorrhage, Witzelsucht

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INTRODUCTION

The German "witzelsucht" means a tendency to tell inappropriate and poor jokes. Patients with witzelsucht have a habitual routine of telling jokes and pointless stories filled with jocularly in socially inappropriate circumstances, and applying many witticisms and quips to conversations, while being oneself inordinately entertained thereby. But, surprisingly, they are paradoxically insensitive to humor. More than 100 years ago, witzelsucht was described following right frontal tumors⁽¹⁾. Here we present a case with witzelsucht and hypersexuality after a hemorrhagic stroke.

CASE REPORT

A right-handed 56-year-old man, KS, with 8 years of formal education, was a factory owner. He had a history of hypertension without medication for many years, but no history of stroke or psychiatric illness. KS suffered from an acute onset of left hemiparesis, sensory loss on the left side of his body, and dysarthria in a morning after playing mahjong overnight. The paresis of extremities progressed rapidly to dense paralysis within several minutes, and he became dull. On arrival at the emergency room, KS presented with, in addition to the aforementioned symptoms, evident left central

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type facial palsy, dysphagia, and a dense visual field defect on his left side. KS was oriented and responded properly, although slowly. He did not undergo surgical intervention because the hematoma was small.

In the following days, KS became alert gradually. He would complain of nasal obstruction, headache, and request oral intake to replace tube feeding which made him feel uncomfortable. On the 5th day, KS could speak for a longer period of time, yet developed inappropriate jokes and exaggerated similes. At that time, KS was alert and cooperative, and had no disorientation, delusions, hallucinations, or emotional liability. He had mild dysarthria and dysphagia, but no abnormal crying or laughter. He was euphoric, outspoken, prankish, and was so talkative that an interruption was usually needed to pull the conversation back to the topic or to complete a test. KS frequently spoke to attract others' attention with a tendency of exaggeration. He also talked a lot of seeming witticisms and quips. On some occasions, he showed no smiles or laughter to the jokes he talked which made everyone laugh loudly, while on other occasions, he was not able to appreciate jokes from the others. However, the content of his chats was not bizarre. For instance, KS called his attending physician "Kong-Ming", who was a famous prime minister in ancient China, and the residents and clerks "Kong-Ming's generals and soldiers". He also interpreted the medical team as a troop to fight his disease. KS had no anosognosia or asomatagnosia. Sometimes, he used right hand to raise the weak left arm repeatedly for rehabilitation, and asked doctors to do their best to take care of his weak limbs. He was concerned about the functional deficits, but talked of them humorously. Freehand copying of representational drawings, such as a clock-face or a flower-head, was fair and showed no hemispatial neglect.

Hypersexuality developed at the same time. KS would use erotic words when there were women nearby. He harassed young nurses and a middle aged caregiver, with amorous language, and would even touch them from time to time. KS called the caregiver "Honey", and asked her to be his "ladylove". He also wanted her to obey his orders, including being touched by him. He would become childish, or even angry, if she refused his

requests. On one occasion, KS called a young female clerk "a little hen" in his vernacular language. He did not care about the complaints from others, and was unable to correct his inappropriate behaviors. KS's family was surprised at his inappropriate jokes and the hypersexual behaviors, which were quite different from that he behaved before the stroke. Before the stroke, he did talk humorously sometimes. But he had never told jokes incongruous with the context, or behaved impolitely or brusquely to females.

He was transferred to the rehabilitation ward on the 11th day of hospitalization. Sertraline, a selective serotonin reuptake inhibitor, was given the next day at daily dose of 50 mg. Moderate reduction of the aberrant behaviors was noted. The effect, however, lasted for no more than three weeks. Two months later, sertraline was discontinued. The dense left hemiplegia improved gradually in the following two months when he could walk with a cane. The sensory loss on the left side of body also was much improved. However, KS's wife still reported that the endless jokes of KS were not only inappropriate in terms of context, but also erotic or even obscene. Medications with venlafaxine, a serotonin and norepinephrine reuptake inhibitor, were instituted roughly four months after the stroke. Interestingly, the behavioral changes were noticeably reduced after the medication with venlafaxine at a daily dose of 37.5 mg for two weeks. In the following two months, inappropriate jokes and hypersexual behaviors were rarely noticed. No relapse of aberrant behaviors was noted before this report was written.

INVESTIGATION

Two hours after the stroke, a cerebral CT scan was done and showed a hematoma at the right putamen (Fig. A). The hematoma was roughly 8-10 ml in volume and extended into the posterior and lateral portion of thalamus as well as the rostral midbrain (Fig. B). A brain MRI was done on the 11th day of hospitalization, showing extension of the hematoma to the posterior and lateral, but not paramedian, portion of the right thalamus (Fig. C). HMPAO-SPECT was performed also on the

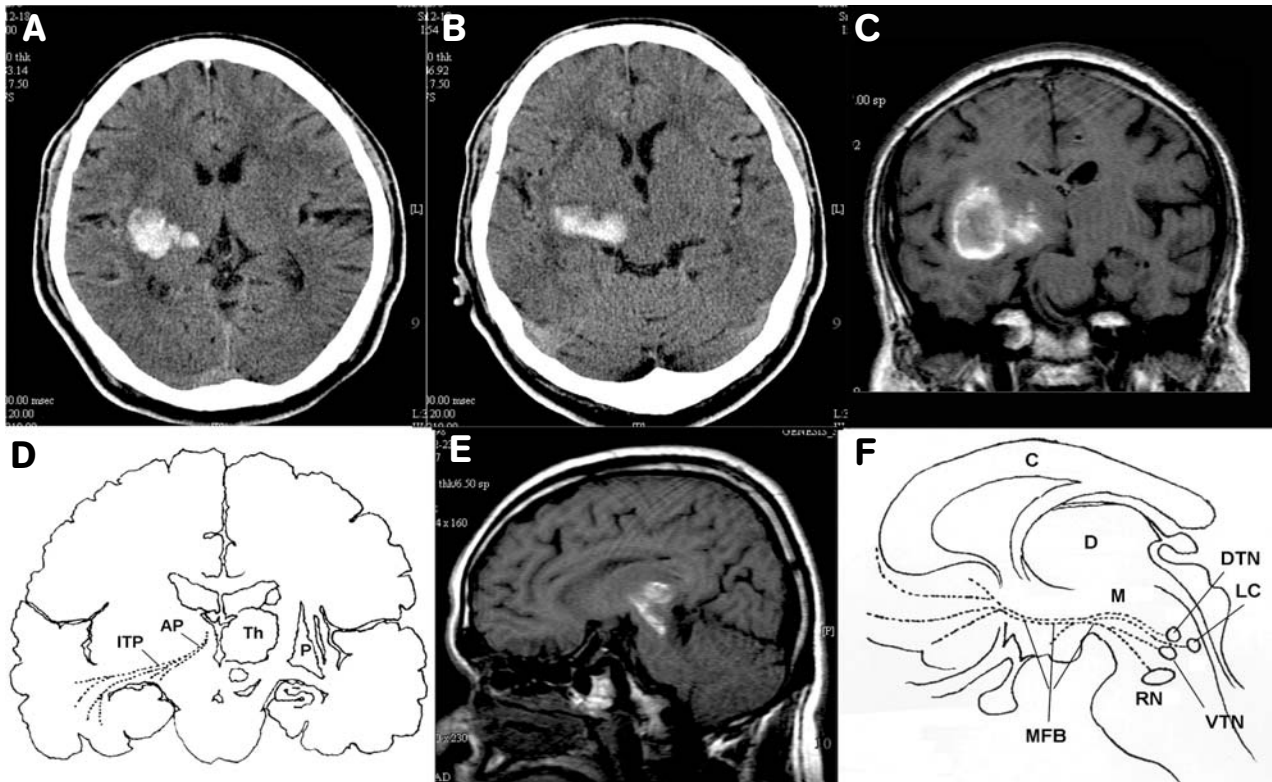


Figure. (A) The Brain CT image shows right putaminal hemorrhage with extension to the posterolateral portion of thalamus and internal capsule, as well as the midbrain (B), two hours after the stroke; (C) The coronal section of T1-weighted brain MRI image shows a hematoma occupying the right subcortical region. Note that the paramedian thalamus is not involved; (D) A sketch depicts the ansa lenticularis and inferior thalamic peduncle on the coronal view of cerebrum; (E) The sagittal section of T1-weighted brain MRI image shows that the rostral midbrain is involved by the subcortical hematoma; (F) A sketch depicts the ascending reticular system and medial forebrain bundle on the sagittal view of diencephalons and mid-brain. Abbreviations for (E) and (F): P: putamen, T: thalamus, AP: ansa peduncularis, ITP: inferior thalamic peduncle, C: corpus callosum, D: diencephalon, M: midbrain, DTN: dorsal tegmental nucleus, VTN: ventral tegmental nucleus, RN: red nucleus, LC: locus ceruleus, MFB: medial forebrain bundle.

11th day, and showed perfusion defect in the right basal ganglion and thalamus, as well as hypoperfusion of right temporal and posterior parietal lobes. There was no reduction of perfusion in bilateral frontal lobes.

Cognitive Ability Screening Instrument (CASI)⁽²⁾ was performed on the 13th day, showing deficits in the domains of recent memory, orientation, abstract thinking, drawing, and verbal fluency. The score of Mini-Mental State Examination (MMSE)⁽³⁾ was 23/30.

DISCUSSION

Witzelsucht: a disorder of humor

Humor is a highly evolved cognitive ability, and,

like the other cognitive functions, it may be impaired by cerebral damages. The disorders of humor include inability to produce or appreciate jokes, and addiction of telling inappropriate jokes⁽⁴⁾. A striking example of the disorders of humor is witzelsucht, which comprises two seemingly paradoxical manifestations: excessive and inappropriate production of jokes, and impaired appreciation of humor. The most frequently mentioned anatomical location of the lesion responsible for witzelsucht is in the orbitofrontal region, especially on the right side⁽⁵⁻⁷⁾. Historically, the frontal lobe dysfunction has been related to personality changes, including pronounced alteration on the production of and response to humor^(8,9). Previous investigations on the effects of cerebral lesion

on humor have suggested the important role of the right hemisphere in the appreciation of humor⁽¹⁰⁻¹³⁾. Shammi and Stuss also argued the unique role of the right frontal lobe in the integration of different cognitive and affective information, including complex human abilities such as episodic memory, self-awareness, and humor⁽⁴⁾. Several cases of witzelsucht with unilateral or bilateral paramedian thalamic lesions⁽¹⁴⁻¹⁸⁾ have also been reported.

The disorders of humor are distinct from disorders of laugh. Laugh is normally an expression of merriment with typical facial movements and clonic contractions of the expiratory muscles⁽¹⁹⁾. Disorders of laugh can occur when this expression is inappropriate or out of control. The most common disorders of laugh are associated with pseudobulbar palsy⁽²⁰⁾. It can be triggered by trivial stimuli, and may be incongruent with the underlying mood, or even intermixed with crying⁽¹⁹⁾.

Anatomical correlation between witzelsucht and subcortical lesions

Our case, KS, had no previous history of psychiatric or personality disorder, and no emotional incontinence either. He was alert and cooperative after the stroke. Thus, his witzelsucht and hypersexuality are most likely consequences of the stroke. However, the hematoma did not involve the frontal lobe on either side (the most frequently reported anatomical locations responsible for such symptoms). Moreover, the paramedian thalamic structures, another previously reported location⁽¹⁴⁻¹⁸⁾, were also not damaged according to the MRI findings. The SPECT study did show a perfusion defect in the right thalamus, a finding perhaps ascribable to the mass effect of the hematoma. However, this defect is unlikely the major etiological factor for KS's witzelsucht because of the persistence of the symptoms for 4 months.

It has been known that emotion and its expression are highly related to the basal forebrain, orbitofrontal region, piriform cortex, and amygdala, all of which are connected with part of the diencephalon such as paramedian thalamus and hypothalamus⁽²¹⁻²⁴⁾. The paramedian thalamic complex comprises mediodorsal, medioventral, and mass intermedia nuclei⁽²⁵⁾. The mediodorsal nuclei consists of three components, namely, magnocellular,

parvicellular, and paralaminar⁽²⁶⁾, and may play a major role in emotional control. The magnocellular component has reciprocal connections with orbitofrontal and piriform cortex⁽²⁷⁾. The connections comprise part of the ansa peduncularis, including the inferior thalamic peduncle and fibers interconnecting amygdala and hypothalamus^(28,29). In the cat, piriform cortex lesions may cause hypersexuality⁽³⁰⁾. We therefore propose that disconnection between the paramedian thalamic complex and the frontal-limbic system might play a major role in the genesis of witzelsucht in KS, as the MRI study clearly showed involvement of the ansa peduncularis and the inferior thalamic peduncle⁽³¹⁾ (Fig. C). The inferior thalamic peduncle goes through the sublenticular part of the posterior internal capsule which is ventral to putamen^(29,31-33) (Fig. D). Consistently, cases with similar symptoms to those of KS have been reported after small lesions in the ventrostriatum and substantia innominata⁽³¹⁾.

On the other hand, ascending reticular systems in the brainstem may also affect emotional expression⁽³⁴⁾. For example, raphe nucleus, locus ceruleus, and some disseminated neurons in the mesencephalic tegmentum send serotonergic, adrenergic, and dopaminergic efferents through the medial forebrain bundle to the orbitofrontal cortex and diencephalon^(35,36) (Fig. F). Specific mesencephalic reticular nuclei such as the ventral and dorsal tegmental nuclei send fibers to join the ascending systems in the median forebrain bundle for emotional modulation^(34,37). Because the hematoma of KS extended down to the mesencephalic tegmentum (Fig. E), it might have interrupted the ascending reticular systems. The temporary responsiveness to sertraline and a sustained relief of symptoms by venlafaxine could support the important role of the ascending serotonin system in the genesis of the behavioral symptoms in KS.

CONCLUSION

This is a case with not only unusual manifestations with a relatively common stroke, but also a unique localization for a special behavioral disorder. We further suggest that the frontal lobe syndrome-like symptoms in this

patient may result from disconnection of the fronto-limbic cortex from its related subcortical structures (including paramedian diencephalon and upper brainstem). These findings also support the essential role of right hemisphere in the production and appreciation of humor, as well as in emotional control.

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