Klüver-Bucy syndrome

What is Klüver-Bucy syndrome?[1]

Klüver-Bucy syndrome is a neuro-behavioural syndrome associated with bilateral lesions in the anterior temporal horn or amygdala. Heinrich Klüver and Paul Bucy first described the syndrome in 1937 after experimental work where they removed rhesus monkeys' temporal lobes.[2] They found that the monkeys developed:

- Visual agnosia - they could see, but were unable to recognise familiar objects or their use.
- Oral tendencies - they would examine their surroundings with their mouths instead of their eyes.
- Hypermetamorphosis - a desire to explore everything.
- Emotional changes - emotion was dulled and facial movements and vocalisations were far less expressive. They lost fear where it would normally occur. Even after being attacked by a snake, they would casually approach it again. This was called 'placidity'.
- Hypersexuality - a dramatic increase in overt sexual behaviour, including masturbation. They may even attempt copulation with inanimate objects.

The syndrome in humans is due to bilateral destruction of the amygdaloid body and inferior temporal cortex, most commonly due to herpes simplex encephalitis (HSE). It shares visual agnosia and loss of normal fear and anger responses in common with the monkey model but one also sees loss of memory with dementia, distractibility and seizures. The hypersexuality tends to be less overt than in the monkeys but may be public and unacceptable.

How common is Klüver-Bucy syndrome? (Epidemiology)[3]

It is a very rare disorder and most of the literature relates to animal models rather than human cases:

- Most literature relating to humans is isolated case reports and few papers report more than a small number of cases.
- It is likely to become more common as a consequence of greater survival following HSE, as antiviral agents improve and are readily available.

Risk factors

The most common cause is HSE but it has also been associated with other infections such as:

- Tuberculous meningitis.
- Listerial meningoencephalitis.[4]
- Primary cerebral Whipple's disease.[5]
- Herpes simplex encephalitis.[1]

Other causes include:

- Head injury (not necessarily very severe).
- Dementia, especially frontotemporal dementia (Pick's disease) but also Alzheimer's disease.[6]
- Surgical lesions.
- Post-epilepsy.[7]
- Cerebrovascular disease.

It has occasionally been described in children.[8]

Clinical features

NB: we rarely, if ever, see the full syndrome in humans.

In adults

- Emotional blunting: there is a flat affect and poor response to emotional stimuli (placidity).
- Hyperphagia: there is a strong compulsion to place objects in the mouth, probably to gain oral stimulation and to explore the object to counteract the visual agnosia, rather than due to hunger. Nevertheless, there is bulimia and there will be marked weight gain unless diet is restricted. Actions may include socially inappropriate licking or touching.
- Visual agnosia: there is an inability to recognise objects or faces visually. This is also called 'psychic blindness' and may account for the oral compulsion.
- Increased sexual behaviour: individuals with Klüver-Bucy syndrome lack social sexual restraint with profuse and inappropriate sexual activity.

In children [8]

- It usually follows HSE and develops on regaining consciousness and activity.
- Altered emotional behaviour, changes in dietary habits, hyperorality and hypersexuality have been reported as present in all, while psychic blindness and hypermetamorphosis occurred in only a few.
- Marked indifference and lack of emotional attachment towards their family.
- Apathy and easy distractibility are rare.
- Bulimia and a strong urge to put items other than food into the mouth are common.
- Hypersexuality presents as frequent holding of genitals, intermittent pelvic thrusting movements and rubbing of genitals to the bed on lying prone. Usually sexually inappropriate behaviour in children is taken as indicative of sexual abuse. There was no suggestion reported that they had been abused and it is thought that their ignorance of sex led to a different pattern from adults.

Diagnosis[3]

KBS is diagnosed clinically by the presence of its characteristic symptoms. Brain MRI is used to confirm the diagnosis by demonstrating bilateral temporal lobe mutilation. Other investigations will be directed at establishing the underlying cause.

Differential diagnosis

The differential diagnosis usually relates to pinpointing the actual site of the lesion(s) and to cause.[9] Where psychiatric symptoms are predominant, the presence of Klüver-Bucy syndrome suggests a primarily organic cause.

Management of Klüver-Bucy syndrome

General points

- Patients need careful monitoring to prevent bulimia and consequent obesity but also to prevent uninhibited and inappropriate sexual activity, which has been reported as leading to criminal conviction for at least one patient.[10]
- Sudden behavioural or emotional changes after HSE treatment may be indicative of a relapse and should prompt a longer course of aciclovir.[11]

Pharmacological

Selective serotonin reuptake inhibitors (SSRIs) have been shown to be of value but carbamazepine may be better.[8]

Prognosis

Cognitive and behavioural disturbances after HSE are often severe but improvement can occur over a long time and residual disabilities vary from major to fairly mild.[12] [13] The loss of memory is consistent with the hypothesis that medial temporal lobe structures mediate memory consolidation.

Prevention of Klüver-Bucy syndrome

A paper from India concluded that HSE is often misdiagnosed, leading to late treatment.[14] Important factors influencing mortality and morbidity are early aciclovir therapy, age, the immune status of the patient, duration of illness and consciousness level before initiation of therapy.

Further reading

- Klüver-Bucy Syndrome; National Institute of Neurological Disorders and Stroke
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