Kluver–Bucy Syndrome

DESCRIPTION

Kluver–Bucy syndrome, an extraordinarily rare neurological disorder, was first documented in primates a little more than a century ago. Research by Brown and Shafer (1888) noted unusual behaviors following bilateral damage or removal of large sections of the temporal lobes in primates. The animals no longer expressed fear or aggression, developed voracious appetites for meat, experienced sensory deficits, and would identify all foreign objects orally.

Beginning in the 1930s with Kluver and Bucy, this syndrome resurfaced under the name of temporal lobe syndrome. Although unaware of Brown and Shafer's work, Kluver and Bucy recorded the same behavioral changes in primates after severe bilateral temporal lobe damage or removal. Kluver and Bucy did acknowledge they were never interested in deciphering the neural or cortical mechanisms responsible for this syndrome, but instead the general role of the temporal lobe in emotional and sensory functioning.

Why this syndrome has been renamed after Kluver and Bucy remains unclear. Nahm (1997) offers the explanation that Brown and Shafer were ahead of their time and Kluver and Bucy's research happens to coincide with a time period where new and exciting advances were being made in brain research. This timing, in turn, permitted their findings to complement the work of others and be more easily accepted and subsequently frequently cited.

Although human examples of Kluver–Bucy syndrome likely existed throughout history, the first human case was not documented until 1955. An epileptic patient following surgery developed an incomplete case of Kluver–Bucy syndrome, which requires the appearance of only three core symptoms. The first definitive human case came 20 years later in 1975. A 22-year-old man exhibited all six core symptoms following a meningoencephalitis infection, likely herpes simplex encephalitis. Since that time, few cases have been documented, attesting to the unusual nature of the syndrome.

NEUROPATHOLOGY/PATHOPHYSIOLOGY

In humans, the precise lesion locations required for Kluver–Bucy syndrome remain unknown, but disruption in several brain areas, including the limbic system, do result in it. Bilateral temporal ablation, as demonstrated by primate research, remains the principle cause of this syndrome. One case in the literature, however, noted Kluver–Bucy syndrome symptoms after just a left temporal lobe resection (Anson & Kuhlman, 1993).

Temporal lobe damage as the main cause of human Kluver–Bucy syndrome has gained support from autopsy studies. These studies report significant lesions in the medial, inferior, and anterior temporal cortex along with amygdalae and hippocampi impairment, specifically the rhinencephalon regions (Lilly, Cummings, Benson, & Frankel, 1983). Overall, human Kluver–Bucy syndrome may require dual hippocampi damage before it can occur.

An alternative explanation of origin proposes that the connecting fibers leading to and from the temporal lobes are disrupted, essentially isolating them from the rest of the brain. Geschwind (1965) used this to define human Kluver–Bucy syndrome as a disconnection between the visual and limbic system. This may explain the syndrome's associated sensory deficits. Further disruption in the connecting fibers with the frontal cortex and limbic areas can also be used to explain the associated memory, emotional, and sexual behavior problems as well.

Specific damage to the temporal lobe or its connections may be the underlying cause of human Kluver–Bucy syndrome, but a host of medical maladies serve as catalysts for this process. They can be grouped into three categories: pathogen, neurological, or other. The pathogenic causes are bacterial or viral and include herpes simplex encephalitis, mycoplasma bronchitis, anoxic-ischemic encephalopathy, tuberculosis meningitis, and methotrexate leukoencephalopathy. Regarding the neurological causes and deficits, human Kluver–Bucy syndrome is viewed, instead, as comorbid or resulting from a primary neurological disorder. Some of the more common neurological disorders are epilepsy, Pick's disease, Reye's syndrome, Alzheimer's disease, Huntington's chorea, and Parkinson's disease. The last category or “other” is unusual since there seems to be no unifying theme. Among this list, there is head trauma, hypoglycemia, arachnoid cysts, carbon monoxide intoxication, postirradiation, and heat stroke.

NEUROPSYCHOLOGICAL/CLINICAL PRESENTATION

Kluver–Bucy syndrome has six core symptoms. These are visual agnosia, excessive oral tendencies, hypermetamorphosis, placidity, altered sexual behavior, and changes in dietary habits. Although humans and primates will display these symptoms, there may be some differences between species as well as minor variations between subjects.

During visual agnosia or psychic blindness, both humans and primates seem to lose their ability to recognize objects visually despite an intact and functioning system. There is some speculation in the literature that sensory agnosias also appear during auditory and olfactory processing, but research in this area is limited. In humans, the visual agnosia usually becomes prosopagnosia where patients experience difficulty distinguishing among the faces of family, friends, hospital staff, and strangers.

Primates suffering from excessive oral tendencies will attempt to identify all objects (e.g., nonfood and food) by licking, biting, and chewing. Once identified, nonfood items are spit out, whereas the food is eaten. Furthermore, nonhuman primates will not typically use their hands to pick up the items but try to use their mouths in a scooping motion.

The excessive oral tendencies in humans are expressed as hyperphagia and bulimia. Humans will lick, bite, and even engage in self-biting. Caution must be exercised with such patients because they will attempt to taste urine (e.g., urophagia) and fecal matter (e.g., coprophagia).

Hypermetamorphosis, the third core symptom, refers to a compulsive need for humans and primates to examine the minute details of every object they encounter while placid. The fourth symptom is a lack of fear, aggression, or interests. Kluver described hypermetamorphosis in primates as appearing as though they were intoxicated from hallucinogenic drugs. Kluver–Bucy syndrome in humans is unique because some patients will be very aggressive whereas others will not, but almost all will be apathetic.

Altered sexual behavior can be extreme in both primates and humans, but it is reported to be one of the least common core symptoms in humans. Primates will excessively masturbate as well as pursue both hetero- and homosexual relations. Some humans will masturbate compulsively though generally only crude remarks and gestures are made. Their attempts for sexual contact are usually unsuccessful. A few humans have reported changing their sexual preference.

The last core Kluver–Bucy syndrome is a remarkable change in dietary habits. Primates, normally vegetarian, will develop an insatiable hunger for meat. Humans will decrease their intake of meat and vegetables for sweets and nonhealthy foods.

Although human Kluver–Bucy syndrome appears more common in teenagers and adults judging by the literature, it can occur in children. Children are capable of having any of the six core symptoms but will exhibit them in somewhat different ways. For example, instead of making crude sexual remarks, they will thrusts their pelvises or rub their genitals against other objects. Children do seem to recognize their parents and other family members, but direct no attention or attraction toward them. Furthermore, children will not respond to any type of behavioral manipulations from family and hospital staff. As of yet, a complete Kluver–Bucy syndrome case has not been documented in a child.

DIAGNOSIS

Given that Kluver–Bucy is a syndrome, recognition and identification of its symptoms constitutes the primary diagnostic practice. It occurs in relation to infection, trauma, or other acute neurological event; thus, diagnostic workup should also seek to elucidate the pathological correlates. MRI and CT are preferred. Depending on imaging results, lumbar puncture with CSF analysis may be utilized to determine the presence of an infectious process.

TREATMENT

Treatment options, other than those that are pharmacological, for human Kluver–Bucy syndrome are extremely limited. The most effective pharmacological agents appear to be the antipsychotics followed by the selective serotonin reuptake inhibitors (SSRIs). The literature regarding the effectiveness of such agents is mixed, and there appears to be no real cure for Kluver–Bucy syndrome.

The antipsychotic, haloperidol, given frequently (e.g., three times a day) does seem helpful in reducing the altered sexual and excessive oral behaviors while reestablishing normal dietary behaviors. During situations when haloperidol is ineffective, it may be combined with other agents such as risperidone, carbamazepine, and propranolol. The best combinations of these agent types and dosages are usually determined through a trial and error basis with each patient. For nontraumatic Kluver–Bucy syndrome examples, a combination of carbamazepine and leuproline is recommended.

Recently, sertraline and fluoxetine, the SSRIs, have been found to be useful in reducing the altered sexual behaviors in Kluver–Bucy syndrome. In one case, risperidone and carbamazepine were ineffective until fluoxetine was added. Once the patient stopped taking just the fluoxetine, the previous Kluver–Bucy syndrome symptoms returned (Slaughter, Bobo, & Childers, 1999).

Recovery from human Kluver–Bucy syndrome can occur, thus indicating an overall positive prognosis. This is unusual because primate examples rarely improve, even after 2-year follow-ups. Generally, anywhere from 1 month to 1 year after symptoms first appear, patients may experience a return to normal eating, sexual, and emotional behaviors. Hyperorality is often considered the slowest to recover from and may only be partial at best. Human Kluver–Bucy syndrome caused by trauma is generally temporary ranging from 7 days to 1 year after the injury.

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