

## Kluver-Bucy syndrome in systemic lupus erythematosus

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**Summary.** The Kluver-Bucy syndrome has not been previously reported as a complication of systemic lupus erythematosus (SLE). A 48-year-old female is described who sustained several cerebral infarcts, some of which were bilateral, due to SLE vasculopathy. She developed a complex behavioural picture consisting of global aphasia, left-side neglect, hyperorality, hypermetamorphosis and hypersexuality. She displayed appropriate emotional reaction to visually presented objects, indicating that her Kluver-Bucy syndrome could not be explained by lack of visual recognition.

**Key words:** Kluver-Bucy syndrome – Systemic lupus erythematosus

### Introduction

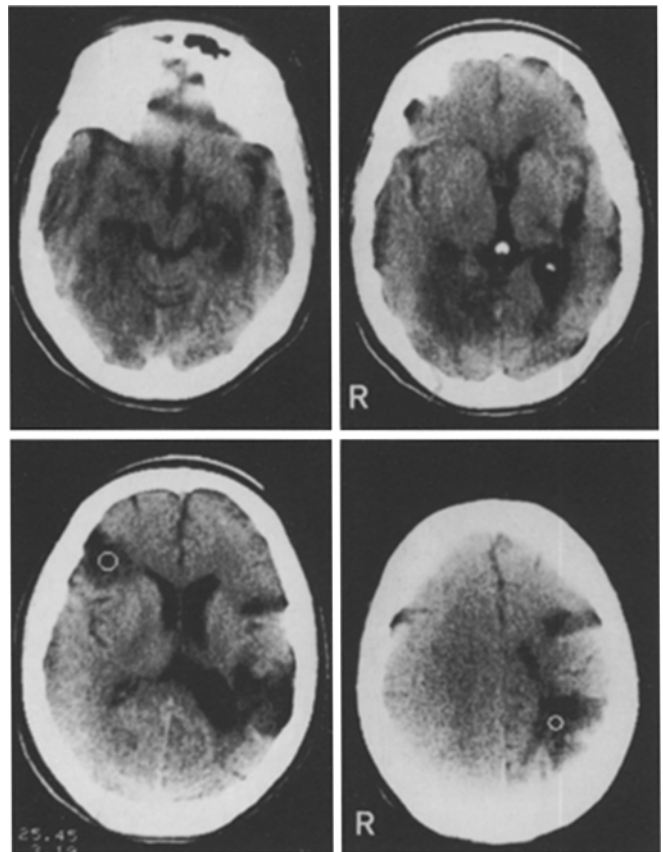
The Kluver-Bucy (K-B) syndrome as described in 1937 [7] consists of visual agnosia, hypermetamorphosis, hypersexuality, placidity and bulimia. It has been reported in man following bilateral temporal lesions secondary to several different aetiologies [8, 10]. We describe a case of K-B syndrome due to systemic lupus erythematosus (SLE).

### Case report

Two years before admission to our hospital a 48-year-old female developed difficulty in recalling events and word finding, as well as lack of efficiency in performing her housework. Six months later she had a sudden episode of right-sided weakness, which partially recovered in the next few months. An increase in her speech disturbances was noticed, with progressive difficulties in comprehension, as well as a decrease in her verbal output with production of frequent paraphasias. She also experienced increasing difficulty in carrying out daily activities. Two months later there was an abrupt increase in her hemiparesis and speech difficulty. Six months after, psychomotor and emotional incontinence were noticed. She then became bedridden. Her past medical history included symmetrical, non-deforming, non-erosive, sero-negative polyarthritis of several years' duration and mild hypertension treated with methyl-dopa.

On admission, she only had sporadic polyarthralgia. Her blood pressure was 160/90 mmHg. Malar rash and livedo reticular were evident on her forearms and thighs. No neck bruits were detected and the results of a cardiac examination were normal. On neurological examination she was found to be alert and cooperative, but periods of placidity would alternate with others in which she was agitated and produced stereotyped utterances. Speech was non-fluent, with palilalia, con-

sisting mainly of stereotypes. She could not name objects or repeat words. Occasionally, simple orders were carried out. She showed alexia, agraphia, constructional, and facial and limb apraxia. There was also neglect for the left hemispace. She displayed a compulsive tendency to touch and manipulate all objects placed nearby, putting them to her mouth and touching them with her lips, even potentially dangerous objects such as an open safety pin. Emotional reaction to visual stimuli was investigated by presenting plastic models of different animals. She always displayed appropriate emotional behaviour, such as reacting with cheerfulness to a small dog and screaming at a rat. Jaw jerk was exaggerated. Grasp, groping and snouting reflexes were elicitable bilaterally. There was a right spastic hemiparesis with brisk tendon reflexes and a Babinski sign on that side. Walking was impossible without assistance. During her stay in the hospital she displayed altered sexual behaviour, showing a stereotyped, non-selective tendency to touch and kiss other persons, both male and female.



**Fig. 1.** CT without contrast medium, showing several cortical infarcts (left temporo-parietal, rolandic and uncus; right temporo-occipital and frontal)

Laboratory tests disclosed mild anaemia (Hb 11.5g/100 ml), persistent leucopenia (3900 WBC/mm<sup>3</sup>), a platelet count of 165000/mm<sup>3</sup>, a high erythrocyte sedimentation rate (49 mm in 1st hour), and an antinuclear antibody titre of 1/500 by indirect immunofluorescence (homogeneous pattern). Complement factors were normal except for a mild increase of C<sub>3</sub> (144 mg/dl, normal range 50–120). VDRL and lupus anticoagulant were negative. CSF was normal. Muscle biopsy (deltoid) showed intimal proliferation and mononuclear infiltrates in small vessels, with occlusion of some of them. ECG and echocardiogram were normal. EEG showed mild diffuse slowing. CT disclosed several cerebral infarcts: two large ones, in the left temporo-parietal and right temporo-occipital regions, and several smaller ones (left rolandic, left uncus and right frontal) (Fig. 1). Angiography showed diffuse narrowing and distal occlusions of several cortical branches of the middle cerebral arteries. Extracranial supra-aortic vessels appeared normal.

She was placed on prednisone (60 mg/day). A decrease in her erythrocyte sedimentation rate was observed (30 mm in 1st hour) and there were no new developments. She was discharged on prednisone (30 mg/day) and 1 year later her condition was still stable.

## Discussion

The full picture of the K-B syndrome is rarely seen in man [8]. It is generally accepted that the diagnosis of K-B can be made when at least three of the typical symptoms are present, as in this patient, who showed increased oral tendency, hypermetamorphosis and hypersexuality. Our patient had bilateral temporal infarcts, this observation being in agreement with Poeck's statement [10] that bilateral lesions of the temporal lobe are *sine qua non* for the occurrence of this syndrome. K-B can be produced by a variety of aetiologies of damage to both temporal lobes, but has not been previously reported in association with SLE. Our patient fulfilled the diagnostic criteria of SLE [11], as in addition to multifocal CNS involvement she had facial rash, arthritis, leucopenia and antinuclear antibodies. Ischaemic stroke in SLE can have different pathophysiological mechanisms, such as small-vessel vasculopathy secondary to hypertension, embolism from endocarditis, vasculitis due to circulating immune complexes, platelet injury and damage to the vascular endothelium caused by antiphospholipid or anticardiolipin antibodies [5, 6]. In our patient there was no clinical or echocardiographic evidence of endocarditis and the lupus anticoagulant was not detected. Angiography showed multiple stenosis and occlusions of medium-sized arteries, suggesting that vasculitis was the most probable cause of the cerebral infarctions.

Poeck [10] stated that the oral compulsion is the most characteristic symptom of the K-B syndrome and that it has a phylogenetic and ontogenetic basis, corresponding to a stage of development when human infants begin to grasp objects with their hands as well as with their mouth. Geschwind [4], on the other hand, thought that the increase in oral activity was due to visuo-limbic disconnection between the olfactory and gustatory cortex and visual cortex.

Patients with visual agnosia due to bilateral temporo-occipital lesions [1] do not usually display features of the K-B syndrome, neither do amnesic patients with bilateral temporal lesions [9]. These features seem to indicate that most of the features of this syndrome are not secondary to visual

perceptual or visual memory defects. The ability to recognize objects in cases of K-B syndrome has not been reliably studied. Bakchine et al. [2] made a thorough neuropsychological study of a case of K-B syndrome due to herpes II encephalitis and concluded that their patient behaved as if he had lost all mnemonic traces of the objects, whatever sensory modality was used to test him. Although our patient was severely aphasic, could not identify objects visually and used most of them incorrectly, she demonstrated appropriate emotional reactions to models of different animals, proving that at least some visuo-limbic pathways were intact and that the other features of K-B syndrome could not be interpreted as due to visuo-limbic disconnection or to impaired visual recognition.

Hyperorality, hypermetamorphosis and hypersexuality can be viewed as a tendency to react indiscriminately to all stimuli or as a disinhibited primitive form of exploratory behaviour. Following Bear's model [3], most of the features of the K-B syndrome, specially those demonstrated by our patient, can be interpreted as an imbalance between the dorsal or parieto-frontal cortical visuo-limbic system from the ventral or temporo-frontal system. Bitemporal lesions causing the K-B syndrome damage the striate – peristriate – inferotemporal visual cortex – amygdala, hippocampus – orbital frontal cortex ventral visuo-limbic pathway. This system's main limbic function is the regulation of drive-relevant responses and the acquisition and storage of visuo-emotional associations. Deprived of this information and regulation the dorsal parietofrontal system, whose essential function is the surveillance of environment for drive-relevant stimuli, becomes hyperactive, reacting in a non-specific fashion, thus leading to an uncontrolled increase in oral, tactile and sexual activity.

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