ISSN 2394–806X
IJHRMLP, Vol: 01 No: 01 January, 2015

REVIEW PAPER

Agnosia, Apraxia and Disconnection Syndrome

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Accepted (Revised) November 29, 2014

ABSTRACT

Cognitive motor disorders other than language and memory deficit that usually occurs as a squeal or presenting manifestation to stroke or other neurodegenerative diseases are not very uncommon. With the advancement of newer functional imaging studies we are now having better ideas regarding involvement of specific cortical areas and network in connection with different cognitive dysfunction. Although no specific pharmacological treatments are available, the underlying causes should be treated. Occupational therapy and cognitive rehabilitation may be helpful in some of these patients. In this review we will discuss the current knowledge on agnosia, apraxia and disconnection syndrome.

***Key Words**: Disconnection Syndrome, Aphasia,White Matter Tract*

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INTRODUCTION

Agnosia and apraxia refers to rare neuro cognitive disorders resulting from stroke or degenerative diseases of central nervous system. Agnosia is a failure of recognition that cannot be attributed to elementary sensory deficit, mental deterioration, attentional disturbances, aphasic misnaming or to unfamiliarity with external stimuli. Agnosia is characteristically modality specific. Apraxia refers to difficulty performing motor acts which cannot be accountable by lack of understanding of the task, weakness or proprioceptive loss. Disconnection syndrome, first described by Karl Wernicke, as disorder related to disruption of white matter association tract connecting two cortical areas of same or opposite hemisphere. The four classical disconnection syndromes described are: conduction aphasia and visual agnosia as described by Wernicke, apraxia by Lipmann, and pure alexia by Dejerine.

DISCUSSION

Agnosia is a relatively rare neuropsychiatric symptom first introduced by Sigmund Freud in 1891, to denote disturbance in the ability to recognize and name objects, usually in one sensory modality, in the presence of intact primary sensation[1]. General public were familiar with the termagnosia from Oliver Sack's story "The Man Who Mistook His Wife for a Hat". Criteria for diagnosis of agnosia[2]: 1) failure to recognize an object; 2) normal perception of the object, excluding an elementary sensory disorder; 3) ability to name the object once it is recognized, excluding anomia as the principal deficit; 4) absence of generalized dementia.

To diagnose agnosia, the examiner must establish that the deficit is not a primary sensory disorder, as

documented by tests of visual acuity, visual fields, auditory function and somatosensory function, and not part of a more general cognitive disorder, such as aphasia or dementia, as established by bedside mental status examination.

Visual agnosia refers to loss or impairment of the ability to recognize things visually with preserved ability to recognize them through touch or hearing in the absence of impaired primary visual perception or dementia[3]. It is divided by Lissauer into two subtypes: appreceptive and associative. In appreceptive visual agnosia there is perceptual defect distorting the visual image so that patient fails to appreciate the whole object but they can pick out feature of an object correctly such as lines, angles, colours[4]. This type of visual agnosia usually occurs in patients with bilateral occipital lesions. The patient may be able to see parts but not the whole, e.g. she may not be able to distinguish a circle from a square. Associative visual agnosia refers to global inability to identify objects but patient can readily identify the same object using other sensory modalities such as tactile or auditory modality occurs in lesions with bilateral posterior hemisphere, often involving the fusiform or occipitotemporal gyri and sometimes the lingual gyri.

Visual object agnosia (optic aphasia) is an associative visual agnosia causing an inability to recognize things that is not due to visual impairment, cognitive deficit, inattention, aphasic misnaming, or unfamiliarity. Patient may be able to see the object, even describe it, but have no idea what it is or what it is called. It must be distinguished from anomia. The patient with anomia cannot recognize the object when presented by another modality (e.g. touch). The anomic patient able to demonstrate the object by gesture (e.g. appropriately applies a comb to her hair, yet not be able to call it comb). The patient with agnosia doesn't recognize the comb as a comb and has no idea what to do with it.

Finger agnosia refers to loss or impairment of the ability to recognize, name, or select individual fingers of the patient's own hand or hand of the examiner. Testing for finger agnosia is usually combined with assessment for the right left confusion. Finger agnosia and right left confusion along with agraphia and acalculia, make up Gerstmann's syndrome and lesion is likely in the region of dominant angular gyrus[5].

Prosopagnosia is the inability to recognize familiar faces. The patient may recognize face as a face but cannot associate it with a particular individual, usually occurs lesions involving bilateral occipitotemporal areas especially lingual, fusiform and parahippocampalgyri. Facial perception is localised to fusiform gyri but recognisation of familiar facies requires anterior temporal memory stores. Prosopagnosia can be described on the basis of disconnection syndrome hypothesis as there are interruptions of fibres passing from the occipital cortex to the memory storage centre[6]. Astereognosis is loss of the ability to recognize and identify an object by touch despite intact primary sensation modalities. Although this represent deficit of cortical sensory loss, it can be described as apperceptive tactile agnosia. It is tested by asking the patient to identify, with eyes closed, common objects placed into her hands (e.g. coin, key). It usually indicates a lesion involving contralateral parietal lobe, may also occur with lesions involving anterior corpus callosum and thalamic radiation. In auditory agnosia patient is unable to recognize sounds (e.g. striking a match, ringing a bell), but can recognize the objects by sight or touch. The site of lesion is in the posterior part of the temporal convolution of the bilateral hemisphere (mostly dominant) especially the primary auditory cortices in Heschel's gyri. Tactile agnosia refers to a disorder of object recognization via tactile modality e.g. touch. There are several mechanisms for tactile agnosia, varies from right hemispheric lesion to disconnection syndrome. Autotopagnosia is loss or impairment of ability to name and recognize body parts. Phonagnosia, which is analogous to prosopagnosia in the visual modality is the failure to recognize familiar people by their voices due to lesion of right parietal lobe. Simultagnosia, first described by Wolpert in 1924, is the ability to perceive only one object at a time. It is usually occurs due to lesion of left occipital lobe.

Apraxia (Greek, praxis—action) is the inability to carry out a high level, familiar, and purposeful motor act in the absence of any weakness, sensory loss or other deficits involving the affected part[7]. There are many variant of apraxia, we are going to discuss the types of most clinical importance.

A loss of the ability to make precise, independent but coordinate movements is called limb-kinetic apraxia. It can be tested by asking patients to rotate a coin between their thumb and index and middle finger as rapidly as

they can. Patients with limb kinetic apraxia have trouble rotating the coin and appear slow and clumsy, often also dropping the coin. This type of apraxia often occurs in the limb contralateral to hemisphere lesion.

Failure to adopt the correct posture of the arm and hand, to move the limb correctly in space and at the correct speeds, or to properly orient the limb to the goal of the action is called ideomotor apraxia (IMA). It is one of the most common types of apraxia. In right handed person, IMA almost always is associated with left hemisphere lesions, but in left handed person, IMA usually is associated with right hemisphere lesions. IMA is also associated with lesion of some other structure of brain e.g. corpus callosum, inferior parietal lobe, premotor area, rarely with basal ganglia and thalamus (pulvinar).

The inability to correctly order a series of movements to achieve o goal is called ideational apraxia. To test for ideational apraxia the patients should be tested for their ability to perform multistep sequential tasks. For example, when asked to demonstrate how to mail a letter, the patient may seal the envelope before inserting the letter. Ideamotor apraxia most often is associated with degenerative dementia but also may be seen with focal lesion of left hemisphere.

In constructional apraxia, the patient is unable to copy geometric forms of any complexity because of impaired visuospatial skill. She may be able to draw a square but not a 3-dimensional cube. In dressing apraxia, the patient loses the ability to clothing correctly. A useful test for dressing apraxia is to turn one sleeve of the hospital gown inside out, and then ask the patient to put it on. Patient with dressing apraxia often gets confuse. Both constructional and dressing apraxias are found due to lesion of non dominant parietal lobe.

Disconnection syndrome: Disconnection syndrome, first described by Karl Wernicke (1848-1904), as disorder related to disruption of white matter association tract that connect different cortical areas with sparing cortical areas of origin[8].Wernicke was the first to suggest such a pathoanatomic mechanism might exist when he described leitungsaphasie (conduction aphasia). Almost a century later, Geschwind[9, 10] expanded and popularized the concept, describing several new examples. In Geschwind's theory, lesion of association cortex if extensive enough, act to disconnect primary receptive or motor areas from other region of the cortex in the same or in the opposite hemisphere. Some of the disconnection syndrome we are going to discuss here are: alexia without agraphia, pure word deafness, conduction aphasia,agnosia and apraxia.

The syndrome of alexia without agraphia was described by Dejerine in 1892, also known as pure word blindness, central aphasia, pure alexia[11, 12]. By definition, alexia is acquired inability to read. The patient is able to speak, understand and write, both spontaneously and to detection, but cannot understand or copy the written word. There is an associated right homonymous hemianopia or right upper quadrantopania. These patients have lesion of left occipital lobe with involvement of splenium of corpus callosum often due to infarction of the territory of the posterior cerebral artery.

Patient with pure word deafness are unable to comprehend spoken language although they can read, write, or speak in a relatively normal manner. The syndrome is "pure" in the sense that it is relatively free of aphasic symptoms found with other disorders affecting language comprehension. This disorder was first described by Kussmaul(1877). Lichtein (1885) defined the disorder as "the inability to understand spoken words as an isolated deficit unaccompanied by disturbance in writing or understanding of the printed words". This is a rare defect and responsible pathology is typically bitemporal or dominant temporal, causing disconnection of primary auditory cortex from the left hemisphere Wernicke area.

Conduction aphasia was described byKarl Wernicke in his thesis"The aphasic symptom complex" which contained a description of the disconnection syndrome, who also called it leitungsapharie[8]. Prototype of all other disconnection syndrome, conduction aphasia is characterised by poor repetition with preservation of other language function, i.e. patient is fluent and comprehension is unaffected. Wernicke originally postulated that a lesion disconnecting Wernicke's and Broca's area would produce this syndrome. Geschwind later pointed to the arcuate fasciculus, a white matter tract travelling from the deep temporal lobe, around the sylvian fissure, to the frontal lobe, as the site of disconnection. The etiology is most often an embolic occlusion of terminal branch of middle cerebral artery.

In agnosia, which has been already described, there is a lesion that spared visual cortex but involved its white

matter output, results in visual sensory images being disconnected from other brain area. The consequences will be inability to recognise common objects presented visually.

In apraxia (ideomotor), the patient is unable to perform a complex command (e.g. salute, wave good bye) with the involved extremity. The patient sometime may substitute a hand or finger for the imagined object (e.g. raking her fingers through her hair instead of showing how to use a comb). Liepmannin in his classical description on apraxia, presented a diagnosis of mixed aphasia and dementia[13, 14]. A striking feature of the patient was that although his spontaneous movements were normal, when asked to perform or copy gesture with his hand (e.g. point to your nose) or manipulate imaginary object (e.g. how to use a comb), he did so in an absurd manner. Since the patient had no visual impairment and no paralysis, Liepmann hypothesised a disconnection of visual, auditory and somatosensory areas from motor area. Later, he described that a lesion localised to the left parietal lobe (dominant) disconnected the left hand area from visual, somatosensory and auditory input, leading to bilateral apraxia whether, a lesion of the anterior portion of corpus callosum disconnected the right hemisphere from the left leading to unilateral left hand apraxia.

CONCLUSION

Disconnection syndrome, as originally outlined by Wernicke and his associates as disorders of higher function resulting from a disconnection of association white matter tract lesion, still exists today with the expansion of association with different other diseases.Also, agnosia and apraxia, representing neurocognitive disorders, can produce significant everyday impairment. Clinicians should be aware of these key symptoms, characteristic neuroradiological findings and neuropsychological based approach that will help to learn more about these rare complex disorders.

Conflict of Interest: Nil.

Contribution of Authors: We declare that this work was done by the authors named in the article and all liabilities pertaining to claims relating to the content of this aticle will be borne by the authors.

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