The role of the cerebellum in the regulation of language functions

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Summary

The present paper is a review of studies on the role of the cerebellum in the regulation of language functions. This brain structure until recently associated chiefly with motor skills, visual-motor coordination and balance, proves to be significant also for cognitive functioning. With regard to language functions, studies show that the cerebellum determines verbal fluency (both semantic and formal) expressive and receptive grammar processing, the ability to identify and correct language mistakes, and writing skills. Cerebellar damage is a possible cause of aphasia or the cerebellar mutism syndrome (CMS). Decreased cerebellocortical connectivity as well as anomalies in the structure of the cerebellum are emphasized in numerous developmental dyslexia theories. The cerebellum is characterized by linguistic lateralization. From the neuroanatomical perspective, its right hemisphere and dentate nucleus, having multiple cerebellocortical connections with the cerebral cortical language areas, are particularly important for language functions. Usually, language deficits developed as a result of cerebellar damage have subclinical intensity and require applying sensitive neuropsychological diagnostic tools designed to assess higher verbal functions.

Key words: cerebellum, language

Introduction

Until the beginning of the 20th century, the cerebellum was considered to be a part of the central nervous system responsible for the regulation of motor functions, such as visual-motor coordination, muscular tone and diadochokinesis. It was then that...
investigators began to recognize the cerebellum’s role in speech control (the first description of ataxic dysarthria in the form of explosive and chanted speech with pauses between syllables and words), but it was still studied only with regard to motor functions [1]. Not until the second half of the 20th century was the possible role of the cerebellum in the regulation of cognitive and emotional functions reported [2]. It is worth to mention about the Polish contribution to this research and the article on the cerebellar diaschisis published in 1997 [3]. In 1998, upon a study conducted on a group of patients with cerebellar damage, Schmahmann and Sherman [4] described a set of symptoms that they termed the cognitive affective cerebellar syndrome (CCAS). Apart from cognitive, visual-spatial and emotional function impairment, language dysfunctions such as anomy, agrammatism and dysprosody constitute a significant part of this syndrome.

Since this study appeared, the interest in this brain structure’s role in psychic function regulation suddenly increased resulting in numerous papers that support the cerebellum’s participation in such processes as working memory [5], acquiring new material [6], executive functions [7], visuospatial functions [8], and affective regulation [9, 10]. Eventually, also the language sphere was meticulously analyzed in this context with the application of both neuroimaging examination of healthy subjects and clinical groups with cerebellar lesions of various etiologies. This paper presents the contemporary hypotheses concerning the mechanisms of the cerebellum’s engagement in language processes with particular reference to their non-motor aspects.

**Neuroanatomical connectivity between the cerebellum and cortical language areas**

The reflection on the possible role of the cerebellum in the regulation of language functions was triggered by the discovery of the main neural connections between this brain structure and the frontal areas, specifically the Broca’s area in the left (usually dominant for speech) hemisphere of the hominid brain. Fundamental for this subject were studies performed by Leiner [11] who was the first to propose that multiple bilateral cerebellocortical connections present in man but not in less developed primates may translate into the actual engagement of the cerebellum in cognitive and language functioning of man. Leiner [12] called attention to the long unacknowledged and underestimated fact that during the phylogenetic development of man the growth of the cerebral association areas was accompanied by a parallel rapid growth of the lateral portions of cerebellar hemispheres and the dentate nucleus, and connections important for cognitive functions were developed between these areas. These emergent neural tracts between the neocerebellum and the frontal lobe comprise not only motor cortex (areas 4 and 6, according to Brodmann) but also other frontal and prefrontal regions, including the Broca’s area (areas 44 and 45, according to Brodmann). Each cerebellar hemisphere receives and sends information to the contralateral hemisphere [11]. Corticocerebellar tracts connecting the lateral parts of cerebellar hemispheres with the
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Frontal lobe consist of ascending and descending pathways. The ascending pathway (the fronto-ponto-cerebellar projections) consists of two tracts: one runs from the frontal cortex to the pontine nuclei in the brainstem (the corticopontine tract) and then via mossy fibres to the lateral portions of cerebellar hemispheres (the ponto-cerebellar tract). The other tract runs from the cerebral cortex to the nucleus ruber, from which the central tegmental tract runs to the medial olivary nucleus and to the external portions of the cerebellar hemispheres. The ascending pathway (the cerebello-thalamic-frontal projections) begins in the dentate nucleus in the cerebellum and runs to the anterior nucleus of the thalamus via the cerebellothalamic tract and further to the contralateral areas of the frontal lobe via the thalamocortical pathway [13]. The motor cortex receives stimuli from the dorsal portions of the dentate nucleus, while the prefrontal cortex from its evolutionarily younger ventrolateral portions. Studies on the role of cerebellar connections with cortical areas crucial for language resulted in coining the term “linguistic cerebellum” and proved its participation in numerous non-motor aspects of cognitive process [14].

The cerebellum and verbal fluency impairment

Hubrich-Ungureanu et al. [15] used functional magnetic resonance imaging (fMRI) to examine the activation of brain parts in right – and left-handed healthy volunteers performing a silent verbal fluency task that consisted in generating in their mind a word from a specific semantic category. As expected, in right-handed subjects the task caused cortical activation in the left frontoparietal region. Simultaneously, a significant activation of the contralateral, that is the right cerebellar hemisphere was observed. Furthermore, a reverse activation pattern that engaged the right cerebral cortex and the left cerebellar hemisphere was observed in left-handed subjects with atypical right hemisphere dominance for language.

Due to these observations the authors concluded that the cerebellum’s part in language processes is inverse in terms of sides to the activation of the cerebral cortex, in accordance with the crossing of the corticocerebellar connections. Further evidence for the cerebellum’s participation in semantic and phonological fluency was provided by studies in which transcranial magnetic stimulation (TMS) was applied [16]. Healthy individuals were subjected to inhibiting continuous theta burst stimulation of the left or right cerebellar hemisphere while performing semantic fluency tasks (animals and vegetables) and phonological fluency tasks (words beginning with letters F, A and S). It was observed that the transcranial stimulation of the right cerebellar hemisphere causes a decrease in the number of connections between clusters (classes of semantically or phonologically congruous words), which is the measure of cognitive flexibility.

The relationship between the cerebellum’s functioning and the processes of semantic and phonological accessibility of a word was also confirmed in clinical trials in patients with both focal brain injuries and neurodegenerative brain lesions. Important
was the first neuropsychological case report of a 41-year-old man with a vascular injury in the right cerebellar hemisphere. The patient did not display deficits in standard neuropsychological batteries. On the contrary, he demonstrated a high level of conversational skills [17], but when asked to generate verbs in response to a noun he gave semantically inadequate names of actions or words that were not verbs. Moreover, he was unable to notice and correct numerous language mistakes that he made.

A few years later, Leggio’s team [18] carried out a study of verbal fluency in a larger group of patients. Subjects with cerebellar damages obtained lower results than a control group in evaluation of both semantic and phonological fluency. Furthermore, patients with focal cerebellar damages performed considerably poorer than patients with generalized atrophy of this brain part, especially if the lesion was located in the right cerebellar hemisphere. Cerebellar damage had a particularly negative impact on phonological fluency, while semantic fluency was relatively well retained. Later, in a group of patients with neurodegenerative cerebellar injuries, Stoodely and Schmahmann [19] observed not only verbal fluency impairment, but also a considerably lower speed of name retrieval in response to specific viewed stimuli.

The cerebellum and grammar/syntactics

Syntactics can be defined as a set of rules governing the structure of a grammatically correct utterance and its reception. Silveri, Leggio and Molinari [20] were the first to put forward a hypothesis concerning the cerebellum’s participation in grammar processing. Examining a patient with a focal vascular injury of the right cerebellar hemisphere, the investigators observed that his utterances contained temporary grammatical violations (agrammatisms) in the form of skipping or substituting nouns necessary in a sentence as well as elisions. This research direction was continued by other scholars [21–23]. Justus [24] discovered that patients with cerebellar injury have difficulties identifying and correcting grammar mistakes made by other people, which made him hypothesize that cerebellar lesions negatively influence not only the expressive syntactic processes but also grammatically correct reception of an utterance.

Explaining this phenomenon, Schmahmann [25] assumed that cerebellum’s role in the regulation of language functions is analogous to its role in motor performance. As in the case of motor functions the cerebellum’s role consists of predicting movement direction and preventing dysmetria (underestimation or overestimation of a distance to an object), in the case of language functions it is responsible for the prediction of the final “linguistic result”, that is identification and control of potential mistakes before the ready utterance occurs. Schmahmann described the impairment of cerebellar mistake control within the scope of non-motor processes as a dysmetria of thought.

Studies conducted by Friederici [26] and Stowe and Doedens [27] support the above hypothesis. In the former, a significantly higher cerebellar activation was observed in fMRI (left gyrus I, lobule VI) during the reading of grammatically incorrect sentences (atypical word sequence), which was in contrast with the reading of sentences contain-
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The observed coexistence of language disorders including phonological, lexicosemantic, and syntactic processes in patients with cerebellar damage made the researchers consider whether there is such a condition as cerebellar aphasia. Mariën et al. [28] described a case of a 73-year-old patient with vascular injury in the right cerebellar hemisphere who displayed an aphasic speech disorder manifested by a relatively retained ability to naming in response to a visual stimulant, phonological skills and repetition, and significantly impaired spontaneous speech, programming of elaborate utterances, their initiation, presence of fragmentary sentences, severe difficulties in word generativity as well as deficits in reading and writing skills. The above deficit pattern was analogous to that observed in transcortical motor aphasia (TCMA), developed as a result of injury in the left frontal region. An MRI examination, however, did not reveal other than cerebellar damages of the central nervous system. This evidently stronger engagement of the right cerebellar hemisphere in language functions made Mariën propose the notion of the “lateralized linguistic cerebellum” [29].

Cerebellar aphasia has also been reported by other authors [30–32], who studied not only individual cases but also groups of patients [33]. Despite the above-mentioned literature, cerebellar aphasia still arouses controversy. Drawing on results obtained in other studies, a considerable amount of scholars deny the existence of such a phenomenon. What is more, they question any influence of cerebellar damage on later language deficits altogether [34, 35].

The cerebellum and dyslexia

It has been more and more emphasized that the ability to read fluently requires synergy between cortical and subcortical regions of the nervous system [14]. It has been suggested that in this process the cerebellum plays the part of a “conductor” who oversees the eye-brain-voice coordination during reading and supervises the process of acquiring proficiency in reading fluently, starting from the automatisation of the grapheme-phoneme conversion, and ending with speech internalization essential in silent reading [14]. This is why many developmental dyslexia hypotheses are based on the assumption that cerebellar dysfunction accounts for the reading automatisation deficit [36]. In literature, the emphasis is put not only on the deficit of cerebellocortical connections but also on the presence of anomalies in the very structure of cerebel-
lum in dyslectic individuals [37]. In a group of patients with dyslexia, Fawcett and Nicolson [38] demonstrated the presence of subtle, specifically cerebellar neurological symptoms, so-called “cerebellar soft signs”, such as: motor disorders, impairment of movement automatisation, balance dysfunction and lower information processing speed. They proposed the cerebellar deficit hypothesis as the main cause of difficulties in automatisation of learned skills such as articulation, reading, spelling and phonological abilities in dyslectic children.

Baillieux et al. [39] noticed that the cortical as well as the cerebellar activation pattern in dyslectic children during noun-verb association was diametrically different from that observed in the control group. While in the latter a well known and described focal activation pattern was observed in the bilateral frontal and parietal cortical regions as well as in the posterior cerebellar regions during performance of such tasks, in dyslectic children the activation of cerebellar cortex was diffuse and scattered (gyri I, II, cerebellar lobules VI, VII, vermal lobules I, II, III, IV, VII). Further evidence for the cerebellum’s participation in reading is also provided by clinical trials, but these are scarce. Moretti et al. [40] noticed that patients with cerebellar damage displayed a lower level of reading correctness making mistakes with regard to both letters and words.

The cerebellum and agraphia

Writing disorder (agraphia) has a twofold etiology. Firstly, it can be a result of motor-sensory injuries that impair the fluency of movements necessary to write letters, giving the picture of peripheral agraphia. Secondly, they can stem from neurological impairment of psychic functions, including the language system. In such case they are referred to as central agraphia [41]. Among cerebral areas responsible for hand writing noted are connections between parietal areas, dorsolateral and medial prefrontal cortex and thalamus, which are dominant in terms of speech. The recent clinical trials and neuroimaging studies suggest a significant role of the cerebellum in the mentioned network [42]. Silveri et al. [43] described two patients with vascular injury in the cerebellum in whom they observed spatial dysgraphia characterized by fragmentary and dysmetric movements during writing. Frings et al. [44] in turn report macrographia observed in six children who previously underwent a surgery of a posterior cranial fossa tumor. De Smet et al. [32] reported three cases of patients with vascular cerebellar injury who had a similar pattern of writing disorders resulting from apraxic agraphia, so-called pure agraphia treated as another example of peripheral agraphia. What is more, these patients occasionally displayed difficulties in recognizing the shape of certain graphemes.

Pure agraphia was also observed in a patient with cerebellar disorders by Mariën [41]. It has been assumed that this type of agraphia is related to dysfunctions in movement planning necessary for fluent and correct hand writing, with purely motor and sensory functions retained in the dominant limb. Consequently, the process of writing is marked by hesitant movements, stoppages, weird, irregular and imprecise movement
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The role of the cerebellum in the regulation of language functions is described as dysfunction of the system of converting information contained in graphic motor engrams (imaging movement during writing – a function of the parietal lobes) in accordance with the appropriate innervation of specific muscles involved in the writing process (movement program – premotor-frontal region) [45]. Both De Smet et al. [32] and Mariën et al. [41] have reported a decreased cortical hypoperfusion observed in SPECT scans in the above described patients with no structural cortical damage and a coexistent hypoperfusion in the damaged cerebellum. It has been suggested that in these patients agraphia is a result of broken connections between the cerebellum and the premotor region of the frontal lobe responsible for attentional processes and motor planning.

Cerebellar mutism syndrome

Cerebellar mutism syndrome is part of a wider posterior fossa syndrome. It has been mainly observed in the pediatric population, most often in children who underwent a neurosurgery of a brain tumor [46]. This syndrome is characterized by severe language, cognitive and behavioral-affective disorders (emotional instability). A lack of verbal activity (apart from automatic sounds produced during coughing and yawning) has been observed [7]. This condition is usually preceded by a short postoperative period of 1–2 days of normal speech. The causes of cerebellar mutism syndrome are sought in the perioperative contraction of vessels supplying the cerebellum and the pons that leads to ischemia followed by brain swelling in these parts, as well as in bleeding or mechanical damage caused either by the surgery or an infiltrating tumor. The mechanism underlying this phenomenon is in turn explained by suppression of mental initiation of any kind of actions, including movements involved in articulation and speech. A retrospective DTI study conducted by McEvoy et al. [47] suggest that patients with postoperative cerebellar mutism syndrome display a significantly decreased fractional anisotropy in the superior cerebellar peduncle (with no significant differences in the middle and inferior cerebellar peduncles or the cerebellar white matter) as compared to patients with no language impairment. Quite often, cerebellar mutism syndrome subsides spontaneously and speech is relatively unblocked, although in most cases ataxic dysarthria remains [48].

Recapitulation and practical implications for neuropsychological diagnosis

Knowledge of cognitive-behavioral syndrome after cerebellar damage is becoming more and more common, as evidenced by the growing interest in this issue by researchers and clinicians. The description of cognitive consequences after cerebellar damage is included in the program of education of clinical psychologist and neuropsychologist. Hence, patients with cerebral damage are increasingly subject to neuropsychological diagnosis and rehabilitation. While cognitive and emotional deficits described in patients
with cerebellar lesions are not as intense as dysfunctions resulting from cerebral damage, it does not mean that they are non-existent or that they do not hinder patients’ daily functioning [23]. In terms of speech, if the ataxic dysarthria is not extremely evident, cerebellar language dysfunctions, having subclinical intensity, may also remain long unidentified by standard tools designed to diagnose aphasia. No sufficiently sensitive neuropsychological screening tool assessing the non-motor cognitive-language functions of the posterior cerebellar lobule has yet been developed. The cerebellar language difficulties usually become apparent only in linguistically demanding situations, e.g., in a hurry, stress or when the subject of a conversation is complex. Therefore, neuropsychological tests dedicated to this group of patients should include tasks engaging higher language processes. Highnam and Bleile [49] suggest that such a battery of tests should be extended to include tasks allowing the assessment of verbal fluency (both semantic and phonological), verbal working memory, retrieval of names during naming of pictures including a measurement of the retrieval time, and narrative tasks allowing the assessment of the type of mistakes made and the capability of self-correction. Although these tasks involve many different cognitive processes, they are difficult to attribute solely to the cerebellar specificity, however, erroneous performance of these tasks along with other features of cerebellar deficits may suggest cerebellar syndrome.

Finally, it is worthy of note that language skills are strictly determined by the efficiency of other cognitive functions, which according to the recent studies also depend on the cerebellum. These cognitive processes certainly include working memory and executive functions, which have been widely studied with respect to their dependence on the functioning of the cerebellum in the recent years [5, 50–52]. The role of the cerebellum in the regulation of these processes is a subject deserving a separate study. It ought to be noted, however, that the neuropsychological examination of patients with cerebellar damage should also include an evaluation of these functions.

References

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