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Cognitive deficits from a cerebellar tumour: A historical case report from Luria's Laboratory

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ABSTRACT

In 1964 an original case report from A.R. Luria's Laboratory of Neuropsychology was published in *Cortex*, being one of the first to draw a link between cerebellum and cognition, by highlighting the manifestation of 'pseudo-frontal' symptoms resulting from a cerebellar tumour. The findings of Luria and his team seem more consistent with modern views about cerebellar interactions with the frontal lobe and its contributions to behaviour than the views prevalent at the time of publication. The paper was originally submitted in Russian, and translated into Italian for its publication by *Cortex*. However, *Cortex* did not preserve the original manuscript in Russian. With the passage of time, and available only to the Italian readership, this case report inevitably fell into obscurity. Hence, we present a translation in English based on the published Italian version of the manuscript and discuss it in the context of Luria's general thinking about information processing in the brain and our current understanding of cortico-cerebellar system. The publication of this article gives readers an opportunity to consider the substantial influence of Soviet neuropsychology on the field internationally under Luria's leadership in the 1960s. It also shows that time is the best judge of ones scientific endeavours, and what may seem implausible today might prove to be valid and worthy of exploration tomorrow.

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1. Commentary

One of the earliest issues of *Cortex* in 1964 contained an original case report that drew attention to 'pseudo-frontal' symptoms resulting from a cerebellar tumour. This report highlights cerebellar contributions to cognition, and in this respect is more in keeping with modern views of cerebellar function than the views prevalent at the time of publication. In this sense it could be considered somewhat ahead of its time.

Patient L presented with neuropsychological symptoms that at first sight appeared to be consistent with frontal lobe pathology. On further inspection, and using an approach that integrated neuropsychology with neurology and psychophysiology, the authors were able to demonstrate a 'pseudo-frontal' rather than a frontal syndrome, and reported that these symptoms related not to frontal pathology, but to a cerebellar tumour.

The authors of this work include Alexander Romanovich Luria, the highly renowned and influential Russian scientist

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and clinician considered to be the founder of neuropsychology in Russia, and his colleagues at the Burdenko Institute of Neurosurgery in Moscow, A.P. Kutsemilova and E.D. Homskaya (1929–2004). This case report is part of a vast research programme undertaken during 1960s in the Laboratory of Neuropsychology headed by Luria. Homskaya, Luria's long-standing collaborator, documented her own work on the frontal lobes in her 1972 book, 'Brain and Activation', recognized as the best in its field and awarded the Lomonosov prize (Fig. 1). She proposed a new electroencephalography (EEG) evaluation method of wave asymmetry that would allow the differentiation of frontal and pseudo-frontal (secondary frontal) symptoms in patients (Glozman and Tupper, 2006; O'Muircheartaigh and Richardson, in press). Research from Luria's Laboratory was tightly linked to the practical work of the Institute through the application of neuropsychological knowledge for topical diagnosis of brain lesions (tumours, haemorrhages, and aneurysms).

Luria's original manuscript submitted to Cortex was in Russian. Cortex translated it into Italian for its publication in 1964. However, the early archives of Cortex were not preserved, and the original manuscript in Russian was not saved (pers. comm., Ennio de Renzi, founder and editor of Cortex at that time). Therefore, we present a translation in English based on the published Italian version of Luria's manuscript for the benefit of a wider readership. Although it was one of the first papers noting the link between cerebellum and cognition, with the passage of time, the paper fell into obscurity due to several reasons: it was a comment on secondary theory linking cerebellum and cognition, and also, it was available only to an Italian readership. We discuss the paper in the context of Luria's general thinking about the distributed nature of information processing in the brain, and its specific relationship with our current understanding of how distributed processing in the cortico-cerebellar system might account for the observations reported by the authors. In the translated manuscript itself we have added accompanying footnotes to help clarify notions that might not be well known to readers and systematically explain Luria's thinking behind the case study.



Fig. 1 – A.R. Luria and E.D. Homskaya (from luria.ucsd.edu/Luria_Pics/Pages/Image20.html).

The study of localised brain damage with neuropsychological methods encompassed only one of the three research areas of Luria's team – the other two being experimental-psychological and psychophysiological study of higher mental functions, and creation of rehabilitation approaches to different neuropsychological dysfunctions (Homskaya, 2001). Luria undertook this specific research interest in the 1920s, during the time of his work with Lev Semyonovich Vigotsky, but it was not until the late 1950s that he began to systematically study deficits of higher mental function in patients with localised brain damage (Fig. 2). It was at this time that Luria's famous monograph 'Higher cortical functions in man and their impairment caused by local brain damage' was published.

The work discussed here exemplifies the challenge that Luria posed to conventional thinking. He challenged the notion that the debate between localisationist and antilocalisationist positions was at all meaningful. Luria's theory of 'systemic dynamic localisation' of mental functions, which was greatly influenced by both Vigotsky and the materialistic traditions of Russian psychology (Pavlov, Sechenov and others), suggested that mental functions are "organized in systems of concertedly working zones, each of which performs its role in complex functional systems, and which may be located in completely different and often far distant areas of the brain" (Luria, 1973, p. 31). In doing so, he acknowledged the specialisation of low-level information processing in discrete brain areas (Petrides et al., this issue, 2012), as well as the requirement for such areas to communicate in order to support higher function (Thiebaut de Schotten et al., this issue, 2012; Catani et al., in press; Yeterian et al., 2012, this issue). Building on this, he challenged conventional ideas about syndromes and deficits. He suggested that since higher functions emerged from the coordinated execution of low-level operations across many brain areas, there was little point in attempting to 'localise' higher functions (e.g., apraxia) to narrow regions of the brain (e.g., the inferior parietal cortex) (Luria, 1973, p. 35). He also proposed that 'primary' deficits could arise from impaired, low-level information processing local to the lesion, whereas there may also be important 'secondary' deficits in higher order functions that require the integrity of several simpler, interacting lower level functions. The authors argued that frontal-like symptoms observed in the patient L (disorientation in time and space, severe abulia, difficulties with motor and cognitive tasks with constant intrusions and interferences) represented such secondary symptoms rather than primary ones (such as nystagmus, gait disturbances, superior colliculus involvement), and arose from the failure to coordinate low-level operations. The approach reflects the influences of Pavlov, who proposed that "optimal, goal-directed activity requires the maintenance of an optimal level of cortical tone" provided by inputs from other brain areas (as cited in Luria, 1973). Luria suggests that subcortical areas are likely to be the major source of such inputs (Krause et al., this issue, 2012; Langen in press, Cubillo et al., in press). Influence was also brought to bear by the work of von Monakow (1914), who similarly proposed that local lesions could give rise to 'diaschisis' (disruptions of physiology in distant areas by virtue of the distributed connectivity in the brain) (as cited in Luria, 1973). These accounts claim that lesions in one part of the brain can result in altered physiological states in distant ones (Catani and Stuss, 2012, this issue). It is significant that in the case of patient



Fig. 2 – A.R. Luria examining a patient during 1960s (from luria.ucsd.edu/Luria_Pics/).

L, Luria and his co-authors do not take the subset of neuropsychological frontal-like symptoms at face value, but instead are careful to consider them in the context of neurological and psychophysiological data, and use these to differentiate between low-level primary symptoms, and higher level secondary symptoms. Their main observations were that (i) symptoms include frontal signs involving disturbances of mental function, but also some signs that did not fit with a classical frontal profile, such as impaired gait and nystagmus, and (ii) abnormalities in the EEG data were present not only in the signals from the frontal lobes, but also in more posterior regions in the vicinity of the posterior fossa. Subsequently, a tumour of the cerebellar vermis was found during surgery, which surrounded both cerebellar hemispheres. This explained the primary symptoms involving impaired gait and impaired eye movements. The authors suggested that the symptoms relating to cognitive function were secondary to these, but importantly, stop short of explaining the mechanism.

Luria and his co-authors did not state anywhere in the paper that the secondary frontal symptoms could be directly attributed to the effects of cerebellar pathology on frontal lobe function, although in our view this is what is implied. Why is this not explicitly stated? Luria and his co-authors did cite neuropsychological evidence to show that their findings were not without precedent (Konovalov, 1957; Shimanskij, 1961;

Shenderov, 1939), but on its own this evidence is not sufficient to make such a claim. At the time of first publication in 1964, conventional wisdom dictated that the cerebellum plays a role in motor behaviour, and there was no convincing evidence of anatomical links between prefrontal areas and the cerebellum, so it would perhaps have been considered bold to make such a claim. Luria and his co-workers would not have known, as we do today, that the cerebellum forms reciprocal and predominantly contralateral connections not only with the motor cortex, but also with the prefrontal cortex (Kelly and Strick, 2003). They also did not have the benefit of many hundreds of functional neuroimaging papers that demonstrate cerebellar activity that can only be explained in terms of cognitive rather than motor demands of tasks (for review papers see Desmond, 2001; Murdoch, 2010; Timmann et al., 2010; Timmann and Daum, 2007). There are also numerous papers that have used imaging techniques to demonstrate crossed cerebellar diaschisis (Baillieux et al., 2010; Botez et al., 1990; Miller et al., 2010; Sönmezoglu et al., 1993), supporting von Monokow's general proposal, and showing specific evidence that cerebellar lesions can profoundly influence the physiological state of large areas of the contralateral frontal lobes. The effects of excising cerebellar tumours on frontal lobe activity and cognitive ability are now well documented, particularly in children (Catsman-Berrevoets and Aarsen, 2010; Davis et al., 2010; Mariën et al.,

2010; Pollack, 2001; Riva and Giorgi, 2000). Recent work provides neuropsychological evidence of cognitive deficits that accompany cerebellar lesions (Leggio et al., 2011; Schmahmann et al., 2007; Schmahmann and Sherman, 1998), with specific impairments in verbal fluency (Stoodley and Schmahmann, 2009), articulatory planning (Mariën et al., 1996; Silveri et al., 1994; Zettin et al., 1997), spatial attention (Townsend et al., 1999), cognitive associative learning (Timmann et al., 2002, 2004), emotional associative learning (Maschke et al., 2002a, 2002b), and mental imagery (Grealy and Lee, 2011). Neuroimaging in healthy subjects provides evidence of cerebellar activity related to verbal working memory (Chen and Desmond, 2005; Desmond et al., 1997, 2005; Durisko and Fiez, 2010; Marvel and Desmond, 2010; Hayter et al., 2007), the acquisition and use of mental skills (Balsters and Ramnani, 2008, 2011; Ramnani, 2006), attention (Akshoomoff et al., 1997; Allen et al., 1997), emotional regulation (Fischer et al., 2000; Schmahmann, 2010; Ploghaus et al., 1999), and mental imagery (Decety et al., 1990). Our revised understanding of organization and information flow in the primate cortico-cerebellar system provides a framework with which one can interpret the authors' observations of patient L's symptoms.

The debate about whether or not the cerebellum influences areas engaged in higher cognitive function (Beaton and Mariën, 2010), or whether or not cerebellar information processing can be attributed specifically to cognitive task demands is now largely settled by a wealth of evidence (but see Alexander et al., *in press*). More recent efforts have focused on the precise role that the cerebellum plays in higher cognitive function, and the ways in which it interacts with areas of the cerebral cortex engaged in the support of such functions. The publication of this article in English gives a wider readership some insights into how Kutsemilova, Luria and Homskaya challenged conventional thinking about cerebellar function long before there was substantial evidence for its role in cognition.

2. Translation of the original article

Neuropsychological analysis of a pseudo-frontal syndrome produced by a cerebellar tumour

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Original Summary

Behavioural disturbances associated with cerebellar tumours are sometimes similar to those due to lesions of the frontal lobes and may therefore be misleading in the diagnosis. We demonstrate how neuropsychological and psychophysiological procedures can successfully supplement the neurological examination in a case of "pseudo-frontal" syndrome due to an arachnoendothelioma of the tentorium cerebelli. Primary involvement of the frontal lobes in this patient was excluded on the basis of the following characteristics of the syndrome: 1. Sudden fluctuations of the behavioural status; 2. Relative integrity of general emotional states; 3. Preserved ability to perform motor tasks without perseveration, and 4. Presence of the activating influence of verbal instructions on the orienting reflex.

It is known that in certain cases tumours of the posterior fossa can give rise to behavioural disturbances that resemble the "frontal syndrome" at a behavioural level. These

psychological changes, which are associated with ataxic disturbances, can sometimes lead to serious misdiagnosis if considered to be the sign of frontal lesion (Kononov, 1957; Shimanskij, 1961; Shenderov, 1939).

Hence, it is of great importance to recognize the symptoms that might be significant for differential diagnosis and distinguish the 'primitive' (behavioural) disturbances of frontal functions from the "pseudo-frontal" or "secondary" frontal symptoms that result from changes in posterior fossa.

Accurate neuropsychological examination provides useful information for differential diagnosis. These show that in cases with frontal lobe lesions, associated with phenomena such as hypertension and cerebral herniation, gross disturbances appear in cognitive processes, consisting in motor inertia, planning deficits, and disinhibition (Filippycheva, 1952; Spirin, 1951; Luria, 1962, 1963).

Certain psychophysiological investigations of patients with frontal lesions showed disturbances of language functions that regulate both involuntary (autonomic) and voluntary (motor) processes (Mesherjakov, 1953; Ivanova, 1953; Homskaja, 1960, 1961; Luria and Homskaja, 1963). Investigations of neuropsychological and pathophysiological processes in patients with "secondary" frontal symptomatology caused by posterior fossa pathology can prove enlightening.

The aim of this study is to use an integrated approach (neurological, neuropsychological and psychophysiological) to investigate a patient with a cerebellar arachnoid endothelioma of cerebellar tentorium presenting with 'pseudo-frontal' symptomatology.

Patient L, 52 years old, Medical Record No. 37644, was admitted on the 5th of March 1964. When first examined, the patient complained of unsteadiness while walking, left frontal migraine, vertigo, and memory deficits. His migraines began a few years earlier. The patient's conditions worsened considerably in August 1963, following a severe thoracic trauma coupled with retrograde amnesia: the migraine became more severe, and nausea appeared together with vomiting, taste disturbances and unsteadiness. The patient was admitted to a neurological ward where he was diagnosed with spontaneous nystagmus, central paresis of the right facial and hypoglossal cranial nerves, a prevalence of deep reflexes in the right limbs, mild ataxia, and bitemporal shrinkage of the visual field with normal fundus. The patient underwent an antiphlogistic treatment followed by amelioration of his symptoms. Two or three weeks after an episode of angina, there was another increase in migraine severity, vomiting reappeared, together with ataxic gait disturbance. The patient became apathetic, began to show memory disturbances, and delivered inappropriate responses to questions. During this period meningeal signs appeared, together with plastic hypertonia in upper and lower limbs, oral automatic reflexes and left hyposmia.

When we first saw Mr. L the patient presented with clear mental disturbances associated with an extrapyramidal syndrome (amimia and changes in muscular tone). No cerebellar signs were apparent. However, his gait was very impaired: the patient walked with small steps and abasia. He had a mild upper gaze paresis, diverging strabismus, his pupils showed no response to light, and there was a weak response to convergence.

EEG results showed brainstem excitability, pathological waves in the posterior fossa, but also some electrical modifications indicating possible involvement of supratentorial medial regions of the brain in the pathological process.

The neuropsychological assessment (A. R. Luria, E. G. Simernitskaja) showed that the patient was lethargic, complained of headache, and was disoriented in time and place: sometimes he claimed to be in Omsk, at the Institute of Microbiology, and at other times he insisted on leaving so that he could meet his wife. Yet, he sometimes correctly asserted he was in Moscow at the Institute of Neurosurgery because of a possible brain tumour. He did not always recognize the examiners, believing them to be his fellow factory workers or “unionists”. Occasionally he did acknowledge the physicians who examined him the day before. He showed obvious disorientation in time, especially in regard to time assessment.

These disorientation phenomena were transient: they were accentuated when the patient was fatigued and on days when there was general deterioration, but these signs sometimes disappeared for short periods.

The patient indicated normal perception of his own deficits, despite his disorientation and general lethargy. He knew he was ill, but he was not able to specify the symptoms. He was aware of his memory problems, and after one test declared: “I cannot hold back anything, while you are reading I understand everything; however, as soon as you stop everything disappears from my memory ... I can understand the text, but shortly after everything dissolves.” Sometimes he reacted emotionally to his mistakes saying: “Look, I am already of no use!”

This severe lethargy coupled with disorientation on the one hand, and preserved awareness of his condition on the other hand, could be observed during the patient’s entire hospitalization.¹

Detailed study of his mental processes revealed the following clinical picture. The patient demonstrated good praxis skills, good finger coordination, and no motor inertia. He performed well at drawing geometrical figures or series of symbols (– + O or – – – Δ+). However, during these tasks, the patient stopped frequently and continued only after repeated stimulation.

Patient had no trouble in reproducing rhythmic sequences (!! !!! or !!! !!!, !!... !!!... or ...!! ...!!), switching easily from one sequence to another. He showed signs of exhaustion and irregular or uneven pacing only after being asked to maintain certain rhythms over an extended time, or to reproduce a rhythm after only verbal instruction. When he noticed his mistakes, he corrected himself, and continued producing the correct rhythm before erring again.

The patient was capable of telling the time by the position of the hands of a clock. He could recognize single figures in an illustration, though sometimes made gross errors: he would take the picture in his hands, oriented the wrong way, and say that he does not recognize what the picture represents, or would deduce the meaning of the image that he perceived

¹ According to Luria, patients with deep lesions of the right hemisphere exhibit disorientation in time and space, or evidence of confusion and disturbances of direct consciousness. However, unlike patient L, these patients lack awareness of their own deficits and are generally poor in self-awareness and self-evaluation – a condition known for many years as anosognosia.

based on the isolated detail randomly chosen. These phenomena should be considered as pseudo-agnosic in character, due to his alterations of conscious state.

He had no language difficulties: he understood the speech directed to him, he named body parts and perceived objects and repeated sequences of 4–5 words. Furthermore, he successfully performed simple arithmetic calculations (31 – 6, 100 – 7, 11 + 9 – 3); however, after completing one task correctly he could not carry on to do the next operation even if it was easier.²

During the examination, the patient’s profound memory deficits became evident. Deficits were present not only in the errors of spatial orientation mentioned above, where they might be confused with disturbances of consciousness, but also in specific experimental tests.

The patient was able to repeat 3–4 words, but failed to recall pairs of words if asked to remember them whilst listening to another pair of words. This effect was also evident in similar tests involving three words. In these circumstances, retroactive inhibition was so marked that the patient was not able to repeat any of the first three words presented.³

Furthermore, these memory deficits could also be observed in the manner in which the patient performed the tasks, forgetting a task only minutes after completing it.

When presented with a prose memory task, he correctly retained the gist of the story (Tolstoy’s ‘The Hen and the Golden Egg’) in immediate recall (“The hen laid golden eggs ... then the farmer decided to kill it ... to get more eggs ... but he found nothing”). However, the patient showed considerable forgetting, perseverations and intrusions if asked to repeat the story for the second time (“A farmer... had these hens that laid good eggs... and another farmer... well things were not so good for him...” – “And?” – “The second farmer decided... that is, not the farmer but his wife... hens decided...” – “What did they decide?” – “To place the eggs from one hen to another hen...”).

Even greater difficulties appeared when the patient attempted to memorize anecdotes of higher complexity; the patient showed

² Luria observed that in patients with frontal lobe lesions, as in patient L, the performance of arithmetical operations was generally unimpaired; however, these patients were unable to switch between operations.

³ Retroactive inhibition produced by the second group of words on the first was strong in patient L, showing that the traces of the last series of words were so weak that the patient could not revert to the first series. The number of perseverations during attempts to reproduce the first sequence after presentation of the second was at least twice as high in patients with frontal lobe lesions than in patients with lesions of the posterior brain areas. An example of this test involving series of two, three or more words followed by a second similar series (‘homogeneous interference’), as taken from the writings of Luria (1973), is given below.

	Two words	Three words	Phrases	Complex sentences
Series 1	house - forest	house - forest - cat	The girl drinks tea	Apple trees were growing in the garden beyond the high fence
Series 2	oak - table	night - needle - pie	The boy hit the dog	The hunter killed a wolf at the edge of the forest

confusion even among basic story elements. Still, characteristically, he continued to correctly assess his own errors.

We report the results of psychophysiological investigations of patient L. (E.D. Homskaya).

EEG showed slow waves in all brain regions of the frequency 1.5–3.5 per second (delta waves), and 4–7 per second (theta waves); alpha waves were harder to detect, and were registered only in central parietal regions (especially in the right hemisphere). Slow waves were prevalent in the anterior regions of the cerebral hemispheres.

As shown in the Fig. 3A, in the anterior central regions of the frontal lobe of the right (III) and the left (IV) hemisphere, slow oscillations (delta and theta waves) were significantly more prevalent compared to the central parietal regions (I and II). Alpha and beta frequency groups were not significantly present.

The presence of the slow waves should be regarded as an expression of the activity of the pathological processes in the deep structures of the brainstem.

The presentation of sound stimuli (intermittent sound intensity of 70 dB) did not lead to any changes in the EEG spectrum. The inhibition of the alpha-rhythm, as a feature of the orienting reflex, did not arise in response to the sound stimulus. Medium frequencies were also not influenced by sound stimuli.

Readings of vascular fluctuations in both the head and the hand indicated a clear dissociation in the form of clear prevalence of respiratory waves in the plethysmograph of the head (see Fig. 4A). Vascular reactions in response to direct unconditioned stimuli (e.g., taking a breath, application of a cold stimulus) were very notable. Sounds of low and medium intensity (up to 70 dB) did not result in a vascular orienting reflex in either the hand or the head. In response to a loud sound (100 dB) there was no orienting reflex, but there was an intense and painful reaction in the form of vascular constriction in the hand and the head (see Fig. 4B), after which, medium intensity sound evoked an orienting reflex seemed to recover (constriction of the peripheral blood vessels of the hand and dilatation of the vessels

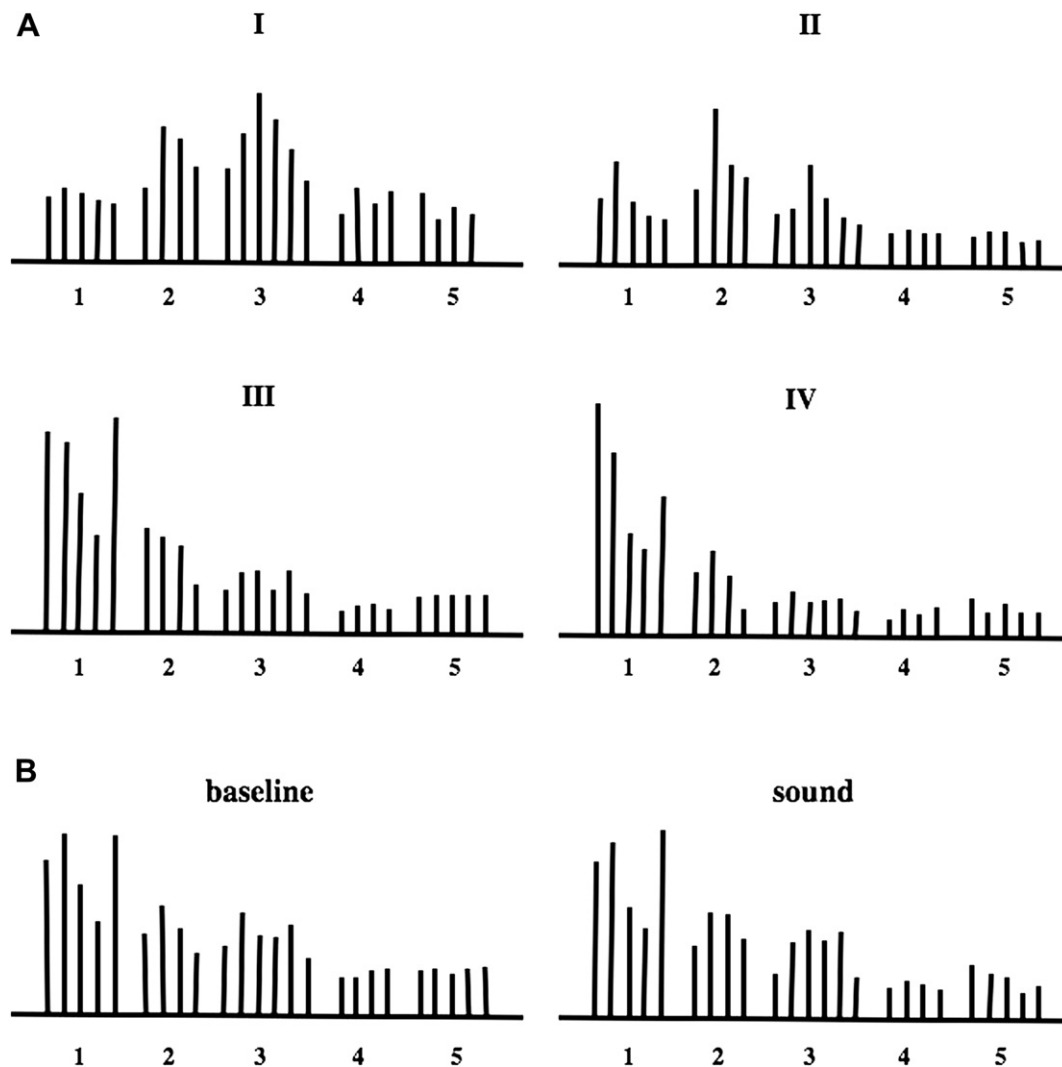


Fig. 3 – (A) EEG frequency spectrum of the patient at rest. Central parietal regions right (I) and left (II) and central regions of the frontal lobe right (III) and left (IV). Mean values for 100 sec. (B) EEG frequency spectrum (right central parietal regions) before and during the presentation of the intermittent sound stimulus of 70 dB (10 sec long). Bellow: mean values for 100 sec just before the presentation of the stimulus. Sound: mean values during the action of the sound stimulus. (1) Delta waves (1,5–3,5/sec). (2) Theta waves (4–7/sec). (3) Alpha waves (8–13/sec). (4) Slow beta waves (14–18/sec). (5) Fast beta waves (20–30/sec).

of the head).⁴ However, the appearance of the orienting reflex caused the patient to be considerably depressed. After an intense sound stimulus, the functional cortical state changed and it was possible to get a vascular orienting reflex to a medium intensity sound stimulus. The plethysmograph of the hand periodically showed respiratory waves of high amplitude, indicating significant oscillations of the cortical functional state, which disappeared after conversing with the patient or after presenting a series of acoustic signals.

The introduction of spoken instruction, “the sound will be followed by a prick”, caused a significant change in a fundamental aspect of plethysmograph readings of the hand (changes in respiration rate) and a dramatic “expectancy reaction” of the prick preceding the application of electrocutaneous stimulation (1 mA, see Fig. 4C). These effects took place against a background of extinction of the vascular orienting reflex to sounds of medium intensity. After relaxation of this “expectancy reaction” to the prick, the orienting reflex could be restored by a subsequent repetition of the spoken instruction.⁵

The neurophysiological examination therefore suggested that a tumour could be localized in the anterior (or medial) regions of the corpus callosum and frontal lobes, as well as in the posterior fossa.

Angiography proved uninformative. For that reason the patient underwent ventriculography that showed very enlarged lateral ventricles bilaterally, and a lack of air in the fourth ventricle, which pointed to the location of the tumour being in the posterior fossa.

During surgery (I.N. Vinogradova, 6-IV-'64) a tumour of the cerebellar vermis was found which surrounded both cerebellar hemispheres. The tumour was removed only partially. After the operation, the patient's condition remained serious,

⁴ The most elementary form of attention, attracted by the most powerful and significant stimuli observed even during the first few months of child's development, Bekhterev called the ‘concentration reaction’ and Pavlov the ‘orienting reflex’. Besides turning of the eyes and head to the stimulus, an ‘orienting reflex’ also includes autonomic responses studied in great detail by Luria, Homskaya, Sokolov, Vinogradova and others. Autonomic features include a psychogalvanic reflex, changes in respiration rate, constriction of the peripheral blood vessels (e.g., in the hand of patient L) while the blood vessels in the head are dilated; electrophysiological phenomena such as inhibition of the alpha-rhythm (or “desynchronization”), or strengthening of the evoked potentials in response to presentation of the corresponding stimulus. Luria and his collaborators developed a differential diagnostic aid tool based on the behaviour of the autonomic features of the orienting reflex evoked by spoken instruction that mobilizes attention (e.g., warning instruction in the case of patient L).

⁵ Under normal conditions the repeated presentation of a stimulus leads to extinction of the vascular orienting reflex; however, as soon as the subject is instructed to direct his attention (spoken instruction) the vascular components of the orienting reflex are immediately restored, as seen in the patient L. However, according to Luria, this does not happen in patients with large lesions of the frontal lobes, where there is a loss of activation as a result of spoken instruction, which would under normal conditions mobilize patients' attention (plethysmographic curve remains unchanged, unlike the case of patient L). It does happen however in patients with lesions in the posterior brain areas, superior parts of the brainstem, and limbic system.

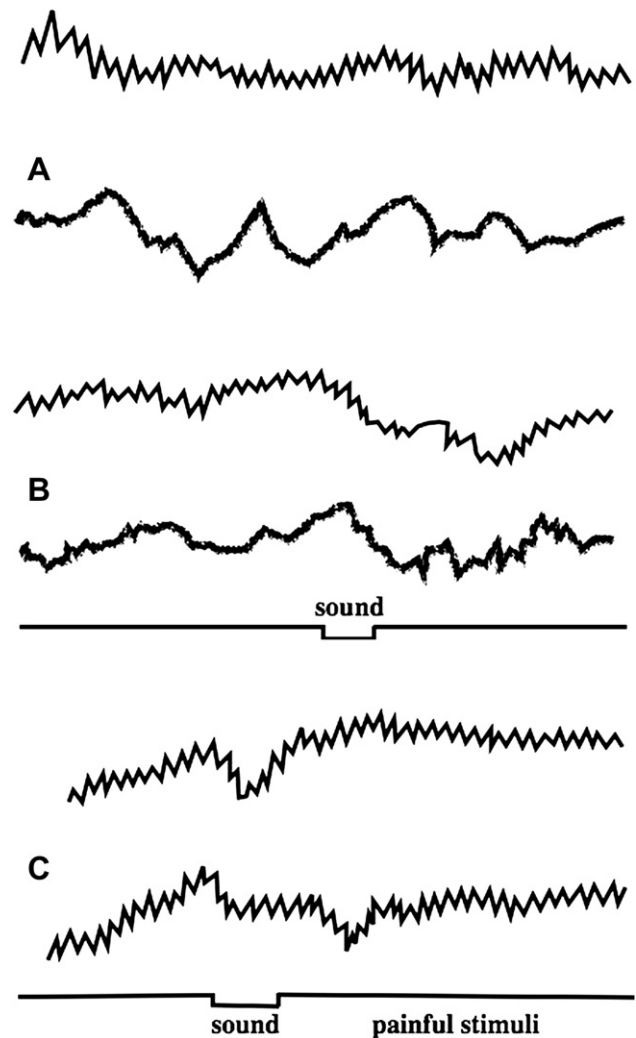


Fig. 4 – Plethysmograph of the head (upper pattern) and of the hand (lower pattern). (A) Plethysmograph of the head shows significant prevalence of the respiratory waves. (B) Painful reaction to the sound stimulus (100 dB): vascular constriction. (C) Manifestation of autonomic components of the orienting reflex to the medium intensity sound (60 dB) in response to a spoken instruction: “The sound will be followed by a prick” leading to vascular constriction in the hand and vascular dilatation in the head.

and three days later he died. Post-mortem brain autopsy confirmed a 3 × 3.5 cm arachnoendothelioma growing from the cerebellar midline, oedema of the brain substance, and herniated cerebellar tonsils compressing the bulbar regions.

The rapid onset of psychological disturbances was the hallmark of the clinical picture. Their presence along with extrapyramidal motor deficits, right pyramidal weakness, central paresis of the 7th and the 12th cranial nerves, oral reflexes, left hypostmia and the EEG findings, all pointed to a possible lesion in the anterior brain regions or in the corpus callosum. However, further careful analysis of this clinical case showed that these ‘frontal’ symptoms were secondary in character. This was in particular revealed by symptoms such as nystagmus, walking deficits, and signs of involvement of the superior colliculus.

Alterations of mental processes observed in this patient could be easily mistaken for a “frontal syndrome” based on the fact that deterioration was so strongly marked. Constant disturbance of orientation, severe abulia, significant difficulties with motor and cognitive tasks with constant intrusions and interferences, could be interpreted as disturbances originating from frontal lobe lesions. However, careful analysis of collected observations counters this hypothesis.

Firstly, the emotional reactions of the patient were appropriate as well as his awareness of his own deficits (which is typically lacking following massive frontal damage). Secondly, the patient performed certain complex tasks flawlessly without manifesting a tendency to replace them with inert stereotypes.⁶ In general, sporadic motor perseverations did not affect the overall performance to specific tasks.

All this differs considerably from the picture of disturbances of mental processes (disturbance of the motor execution, self regulation and self control) usually found in patients with massive lesions in the frontal lobes.

Lastly, and possibly most characteristic of this case, is that his performance changed dramatically from task to task, going from success to failure. This alternate performance, in our experience, is usually found in brainstem lesions or in cases of grave hydrocephalus and is due to severed relationship between the reticular formation and the cortex.

An index of the psychophysiological secondary nature of mental disturbances manifested by the patient was represented by the presence of sharp fluctuations of the functional states of cortical neurodynamics, and above all, by the possibility of evoking orienting reflex through the introduction of verbal instruction. This observation shows the possibility that cortico-reticular pathway for excitatory transmission is preserved, which is contrary to what is observed in many patients with primary lesion in the frontal lobes, in which the similar appearance of the orienting reflex in response to any verbal instruction is completely missing or is significantly weakened (Homskaja, 1960, 1961, 1964; Luria and Homskaja, 1963).

This report shows that an integrated approach combining neurological, neuropsychological and psychophysiological methods can substantially differentiate between frontal and pseudo-frontal syndrome diagnosis, which is often one of the most challenging tasks in clinical work involving focal brain lesions.

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⁶ Inert stereotypes or ‘inertia of motor stereotypes’ reflects disturbances of skilled movements, which are no longer performed smoothly. In Luria’s neuropsychological tests these deficits are manifested in patients’ inability to perform a complex movement or switch from one component of the movement to the next when asked to tap out a complex rhythm (e.g., –...–...). Patients with premotor lesions cannot switch smoothly from two beats to three, or from loud beat to weak, unlike patient L who seems to perform this task correctly.

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