Luria’s frontal lobe syndrome: psychological and anatomical considerations

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SUMMARY Luria has described a syndrome of disinhibited and impulsive behaviour, in which the patient is unable to follow sequential instructions. This he attributes to localised frontal lobe damage. However, group studies of patients with focal lesions of the frontal lobes fail to reveal such a syndrome. A patient is described who displayed a form of Luria’s syndrome temporarily. Psychometric and post mortem evidence indicate that the syndrome arises only under conditions of more global cerebral dysfunction.

Luria, Pribram and Homskaya described the frontal-lobe syndrome thus: “The destruction of the frontal lobes has relatively little effect on the course of simple and well-consolidated motor stereotypes. Yet, the patient with frontal lobe destruction is often unable to fulfil instructions, is unable to inhibit impulsive reactions or to hold back the tendency towards fixed repetition of movement.” In particular, “ . . . dissociation of verbal and motor reactions is typical”. These conclusions are based upon bedside observations of the patient “Zav”, a 43-year-old female office worker with a left frontal arachnoidal endothelioma (meningioma). This patient was able to reproduce single movements with ease, but was unable to copy sequences of movements, or to produce sequences of movements to a single command. She was also unable to repeat a series of rhythmical beats made by the experimenter on a piece of wood with his pencil: the patient’s own tapping quickly deteriorated into a perseverative continuous pattern. These behaviours were not due to defective appreciation of instructions; the patient was able to repeat instructions verbally even while producing erroneous performances. In such circumstances she was unable to say whether or not her performance was correct.

This syndrome has only rarely been reported in detail. Another case is that of IS, a 69-year-old housewife suffering from Hodgkin’s disease, with metastasis to the left frontal lobe. This patient was examined four weeks after subtotal removal of the infiltrating tumour. As with the first patient she was able to follow simple commands, but unable to perform serial tasks. In a figure-drawing exercise she displayed gross perseveration: at one point she drew the letter “A” in response to almost any command. Unlike the first patient she indicated that she knew she was making an error. Nevertheless this did not affect her performance.

Group studies of patients with frontal lobe lesions have failed to reveal a syndrome as consistent as that described by Luria. Anecdotal evidence obtained during maze-learning supports Luria’s claim, but more recent work employing the same task found no such evidence. A task designed specifically to examine the relation between verbal performance and motor performance failed to find the predicted dissociation. Thus it has been concluded that such a dissociation is not a general feature of the performances of patients with frontal lobe lesions.

Nonetheless, the existence of even a small number of patients exhibiting this syndrome is of theoretical interest. The precise localisation of the pathology underlying the syndrome would be of value. The patient described by Luria et al was operated upon within days of the examination, but died of heart failure about 24 hours after the operation. Histological examination revealed relatively widespread
pathology of the left hemisphere, as well as limited pathology of the right. The anterior edge of the tumour remnant was 4.5 cm from the frontal pole, the diameter of the defect being 7 cm in the anterior-posterior, 4 cm in the transverse and 2.8 cm in the dorsoventral direction. The whole hemisphere was markedly oedematous, the cingulate gyrus and the corpus callosum were distorted, and the head of the caudate nucleus was compressed. In addition both hippocampal gyri were pushed medially, cut by a deep furrow with traces of pressure at the edge of the tentorium cerebelli, and the posterior horn of the right lateral ventricle was dilated. Despite these massive lesions Luria speculated “. . . that the more generalised brain damage brings out in relief . . . the essence of a disturbance produced by a local lesion”1 although he did not specify any particular location within frontal cortex. The case IS did not shed any further light upon this issue, since the results of a post mortem examination have not yet been reported. Other cases described by Luria,23-8 are similarly lacking in detailed pathology, the usual description amounting to little more than the statement “A patient with a massive frontal lobe tumour”. A recent case of our own, JH, has been reported briefly elsewhere,9 and since post mortem evidence is now available a fuller description is warranted.

Case history
JH was a 64-year-old retired dance teacher, with a 6 month history of numbness of the left leg below the knee. The numbness had first been noticed after she had been found unconscious on the floor. She had been sick but not incontinent. Previous to this she had been witnessed to suffer an absence attack, though without shaking or falling. The numbness progressed and she was said to become “forgetful and irritable”. Upon admission she was orientated in time and place, but unable to remember verbal material over short periods. There was papilloedema in both eyes and anosmia on the left. There was slight weakness of the left leg with impairment of the joint position sense of the left foot. A CT scan and bilateral carotid angiography revealed a very large posterior frontal (midline) falx meningioma invading the superior sagittal sinus. The tumour, measuring 7 cm in diameter, was subsequently removed in its entirety. Psychological investigations commenced on the thirteenth postoperative day and continued for a month, when the patient was discharged. She was subsequently seen as an outpatient several times, until the point sixteen months after surgery when she suffered a myocardial infarction and died.

Description of the syndrome
On the thirteenth postoperative day JH displayed all of the symptoms characteristic of the frontal lobe syndrome described above: there was marked motor perseveration both in spontaneous behaviour (for instance, she displayed a tendency to tap or rub parts of her body continuously), and also while copying drawings (for instance, after completing a straight line or a square she would then retrace each line vigorously until it was about half a centimetre thick). When asked to copy a continuous but castellated line she would produce several lines of completed squares. This motor perseveration occurred relatively independently of verbal control. If asked to stop during the course of tapping or drawing, she would say “yes” but continue regardless. If she herself said “I will stop now” the behaviour also continued unchecked. However, if she was asked to start a different activity (for example placing her hands by her side) she was able to do so and thereby terminate the previous activity. At this time, when asked to fill in the numbers on an empty clock-face, she would draw as many numbers as could be fitted on the face, starting with a “1” in the appropriate place and counting upwards until returning to the “1” position. This often resulted in fourteen or fifteen numbers, although she was quite clear upon questioning that there could not be a “fourteen o’clock” or “fifteen o’clock” time.

Apart from these informal tests, JH also underwent standardised psychometric examination. These procedures have not previously been employed with patients displaying the frontal lobe syndrome.12 6-8 On the thirteenth postoperative day JH was found to have a WAIS verbal IQ of 63, and a performance IQ of 615 (see table for individual subtest scores). Since JH could be presumed to have been of average premorbid intelligence, this is evidence of generalised deterioration, which is not normally seen following circumscribed frontal lobe lesioning.4 6 A short form of the Minnesota Dysphasia Battery11 revealed some minor verbal deficits. JH did well at tests of reading and spelling, but displayed difficulty in naming pictures (although only 2/20 errors) and in identifying items serially (2/6 errors). This deficit was not purely expressive dysphasia, since a picture vocabulary test12 which requires only pointing as a response yielded an IQ of 56, which is close to those obtained from the WAIS. Verbal memory was also impaired. Immediate recall of two stories13 was 3/23 and 0/23 items respectively. She also did poorly when asked to remember ten series each of ten words.14 Her average was one item per list compared to the average for the normal population, which is about 6-5 items per list. Delayed recall of nonverbal material15 was also impaired. Thus it would appear that psychometric investigation of the frontal lobe syndrome described by Luria reveals not so much a pattern of specific impairments but rather a complex generalised intellectual disruption.

<table>
<thead>
<tr>
<th>Subtest</th>
<th>First occasion</th>
<th>Second occasion</th>
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<tr>
<td>Comprehension</td>
<td>3</td>
<td>10</td>
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<tr>
<td>Similarities</td>
<td>2</td>
<td>10</td>
</tr>
<tr>
<td>Vocabulary</td>
<td>3</td>
<td>10</td>
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<tr>
<td>Digit span</td>
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<td>8</td>
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<tr>
<td>Block design</td>
<td>0</td>
<td>10</td>
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<tr>
<td>Object assembly</td>
<td>5</td>
<td>11</td>
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Table Individual WAIS age-scaled subtest scores at the time of initial assessment and one month later
Course of the syndrome

The motor perseveration observed on the thirteenth postoperative day proved to be a relatively transient phenomenon. By the seventeenth postoperative day JH was showing signs of improvement in her ability to copy drawings, and on the eighteenth postoperative day and subsequently she was able to perform such tasks without difficulty. Psychometric assessment was repeated one month after the initial examination, and this time JH scored mainly in the normal range (for example verbal IQ 94, performance IQ 107—see table for individual subtest scores). It was still possible to detect perseveration on a card sorting test, as is common in patients with circumscribed frontal lobe lesions, but the gross perseveration characteristic of the syndrome described by Luria had by this time completely disappeared. Fluency tests also elicited impairments, as would be expected of a patient with bilateral frontal lobe damage. In short, the syndrome described by Luria appeared only in the context of widespread intellectual dysfunction in the immediate postoperative period. Within weeks this syndrome had disappeared, leaving in its wake only the more subtle deficits known to be associated with frontal lobe damage. None of the cases described by Luria and his colleagues have been administered standardised tests of the kind described here, and thus it might be that widespread intellectual dysfunction is a prerequisite for the syndrome to appear. In the present case it must be assumed that the immediate postoperative syndrome arose through the temporary effects of oedema or disrupted circulation.

Postmortem evidence

Postmortem examination revealed the extent of brain damage responsible for the more permanent but more subtle deficits. This evidence also shed light upon the extent of brain damage necessary to produce the syndrome described by Luria.

The fresh brain weighed 1060 g. When fixed and after

Fig. Nine coronal slices through the cerebral hemispheres numbered A to I from in front. Arrows in G and H indicate the central fissures (of Rolando). Arrowhead in E points to the scar in the right thalamus. Note a small hole in the left globus pallidus in D. The loss of cingulate cortex is seen on the left in C and on the right in D. The anterior part of the corpus callosum, but not the genu, is abnormally thin. The extent of the tumour was from B to E.
slicing, it weighed 996 g. There was a ragged defect 5 cm long and about 4-5 cm wide in the mid frontal parasagittal region, roughly corresponding to the site of the meningioma that had been removed previously. The borders of the defect included grey fibrous meningeal tissue and orange-yellow old iron pigment. Inspection of the under surface of the brain showed patchy atheroma with calcification and narrowing of the lumen of the larger arteries. There was bilateral uncinate growing some 3 mm from the medial border, a little deeper on the left where there was also some grooving in the parahippocampal gyrus. Coronal slicing of the cerebral hemispheres (fig) showed a patchy loss of cortex and scarring in the white matter in the defect that was seen from the outside. Anteriorly, scars in the convexities, about 1-5 cm from the midline, extended to within 2 cm of the frontal poles. The white matter between the outer surface of the top of the hemispheres and the ventricles felt soft and looked less white than normal. This abnormality extended posteriorly to the central fissure, more so on the right. There was a separate small cavitated cortical scar in the posterior upper border of the right frontal convexity. The cingulate gyri were preserved except for some cortical loss deep to the defect and softening of the white matter on both sides at that level. The corpus callosum was thinner than normal for some 3 cm posterior to the genu. The lateral ventricles were very much dilated with a stretch of white matter separating the caudate nuclei from the corpus callosum. There were two other old scars: one cavitated about 4 mm in diameter in the medial border of the left globus pallidus at the level of the genu of the internal capsule, and one narrow grey streak 3 mm long in the anterior part of the right lateral thalamus deep to the lateral ventricle. There were no lesions in the brain stem or cerebellum or spinal cord.

Discussion

At necropsy there was widespread cortical and subcortical pathology. It is likely that the preoperative motor and sensory clinical abnormalities were related to the old lesions in the right convexity or thalamus. The lesion in the left globus pallidus may not have given rise to any symptoms or signs. It is a little anterior to the old target area for stereotactic lesions in Parkinsonism. The lesions in the brain in the bed of the tumour may well have been affected by surgery; these and the cortical and white matter abnormalities more anterior and posterior probably relate to a disturbance of the circulation through the superior sagittal sinus caused by the tumour. The anterior cerebral arteries appeared normal and there was no evidence to suggest the presence of arterial infarcts. The location and extent of the abnormalities match the dearth of neurological signs; they are consistent with a relatively good level of intellectual performance before, and later after the operation. The reference to forgetfulness some time before her admission, and to poor short-term memory on admission, may hold implications for a disturbance of the limbic circuit at the level of the abnormalities in the cingulate gyri, in the absence of any other visible lesions. Histological examination has not been carried out because it seemed very unlikely that it could help with either the anatomy or the pathogenesis of the obvious abnormalities.

The widespread frontal lesions described here can account for the long term subtle psychological disturbances seen in this patient. However, even the gross damage noted above was not sufficient to sustain the "frontal lobe syndrome" described by Luria. This casts some doubt upon Luria's assertion that the syndrome reflects focal frontal dysfunction, especially since group studies of patients with circumscribed frontal lobe lesions fail to reveal such a syndrome. Nevertheless when the syndrome is reported it is associated with frontal lesions and not as a rule with occipital, temporal or indeed globally dementicing conditions. The evidence presented here would suggest that the syndrome is not produced by a frontal lesion alone, but by a frontal lesion in a setting of more global cerebral dysfunction from displacement, distortion or oedema.

References

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