A CASE OF 'PURE' WORD BLINDNESS

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The concept of 'pure' word blindness is still a matter of controversy. Some authors maintain that it is a selective asymbolia (Holmes, 1950; Symonds, 1953; Brain, 1955) while others (Critchley, 1953) consider it to be an aphasic disorder. Beringer and Stein (1930) attribute its mechanism to some pathological functional lability of the visual system. Martin (1954) assumes that it is due to visual disorientation, and Warrington and Zangwill (1957) ascribe it to disorders of ocular motility.

Alexic disorders have been named either according to semiotic criteria, for example, pure word blindness, pure optic alexia, or to pathological criteria related to the site of the lesion, for example, subcortical word blindness (Wernicke, 1886) or occipital blindness.

At the same time, a review of the literature on this subject illustrates the great variety of techniques used for clinical examination, and the lack of a standard clinical examination of alexia has made a synthesis of this problem as well as drawing general conclusions very difficult (Leischner, 1957).

We consider this report of a case of 'pure' word blindness to be of some interest: it was studied by means of various classical tests and also by a standard examination for alexia worked out at the Institute of Neurology, Bucharest, by Weigl and Fradis (1960).

Case Report

V.M., aged 62, right-handed, was admitted on September 27, 1960, complaining of difficulty in reading and of paraesthesia in the right upper extremity.

The patient had suffered from lues in 1916 for which he had been incompletely treated. For the past two or three years he had been found to be hypertensive, with maximal values of 20 to 21 mm. Hg.

The patient had only an elementary education. Since his youth and until his admission to hospital he had worked as a commercial employee in various concerns (cereals, textiles, metallurgy). His heredo-collateral antecedents were not significant.

The patient complained of paraesthesia in the right upper extremity, for four or five weeks. Subsequently, he suddenly experienced difficulty in reading words and letters and, at the same time, a narrowing of the visual fields. (He was no longer able to see clearly to the right.)

His writing had not changed, but in current speech he sometimes had difficulty in finding the right word. Despite this symptom he was still able to make himself understood to those around him, but on account of the visual disturbances and difficulty in reading he had to give up his work. He was admitted to a provincial hospital where a Wassermann test in the blood was found to be positive.

On examination there was a right homonymous hemianopia but macular vision was preserved (visual acuity 1). Examination of the fundus oculi showed a normal disc and second-degree arteriosclerosis of the retinal arteries. There was slight anisocoria, the right pupil being larger than the left, but both pupils reacted promptly to light and convergence. The eyeballs moved normally. Nystagmiform movements were found on looking to the right. There was right central facial paresis without any other cranial nerve disturbances. Power, tone, and coordination as well as reflexes were normal. No superficial or deep sensibility disorders were noticed.

The patient was well orientated in time and space, as well as concerning his own person and his environment. He had a lively and practical intelligence, promptly answering the examiner's questions and displaying a normal capacity for abstraction. His ideas were coherently and logically associated without interpretative tendencies. Attention was good and prompt, all tests being correctly performed. Fixation and recall memory for recent events were slightly impaired. His memory for remote events was particularly good. His affect was unchanged. No pathological disorders were observed in the volitional sphere.

His blood pressure was 17/9 mm. Hg but there were no other pathological visceral changes. The Bordet-Wassermann and cytochol tests were strongly positive in the blood. Cerebrospinal fluid contained 4.6 cells/c.mm., albumin 0.38 g. per 1,000 ml., Pandy and Nonne-Appelt tests positive, Wassermann reaction negative. Glycaemia was 1.50 g. per 1,000 ml. (three repeated determinations), the E.S.R. 25 mm./hr., 55 mm./two hours. A routine urine examination failed to disclose glycosuria. The blood urea was 0.20 g. Radiographs of the lungs were normal. A simple cranial radiograph showed marked diploic vascularization, perisellar ligamentous calcification, a normally calcified and located pineal gland, and partial calcification of the falx cerebri. An E.E.G. disclosed a flat tracing which was desynchronized in all leads. Hyperpnoea did not alter the tracing. There were no signs of a focal lesion.

Language Examination

Spoken Speech.—Spontaneous, narrative, and dialogue speech was fluent and coherent, the patient being only very rarely at a loss for a word. Morphological and syntactical constructions were adequate and no paraphasia was found. Repeated speech was well preserved; the patient repeated all the words he was given without making a single mistake. To test automatic speech, the days of the week and the number of days in a year were given correctly. He showed no deficit in understanding spoken language. He performed both the simple commands and the complex ones, composed of two, three, or even four commands, including the Pierre-Marie test and Head's tests, without error. In Kleist's tests the patient readily identified all the words which were pronounced correctly and distinguished all words which made sense from those which did not. When naming objects and animated things, he displayed a slightly increased latency in his answers but considerably shorter than in aphasia.

Standard Examination for Aphasia.—The patient named correctly the 20 pictures of objects and animated things contained in the standard test (Kreindler and Fradis, 1957). Only by a very painstaking test, when another 28 pictures of animated things and of objects were used, was there a slight anomia; for instance, the patient was unable to name an ashtray, a pad, a brow, a fox, as well as certain colours (see colour gnosis).

Written Language.—The patient's reading of individual letters, both silently and orally, displayed important disturbances. Printed or handwritten letters were only identified with difficulty and he made many mistakes. His mistakes in recognizing the 25 letters of the alphabet varied also in relation to the type of letter (Table I).

TABLE I
READING OF LETTERS

Type of Letter	No. of Letters Recognized out of 25 Shown
Printed (capital) letters	14 (56%)
Printed (small) letters	13 (52%)
Handwritten (capital) letters	10 (40%)
Handwritten (small) letters	15 (60%)

Consequently, the most important disorder in this group of tests was detected in reading handwritten capital letters, probably due to the fact that these letters are not much used in current writing.

To test his ability to recognize the categories of written symbols the patient was given printed and handwritten letters mixed with numbers and punctuation marks and was asked to sort them (Leischner's test, 1957). His sorting according to categories was faultless.

Standard tests of alexia according to Weigl and Fradis (1960) were also given. (1) The patient was given a model of a letter and told to find the identical letter in a group containing numerous written letters. He grouped the identical letters without making a single mistake. (2) He was given a model of a printed letter and told to find the corresponding handwritten one. In only seven out of 17 instances was the patient able to pass this test correctly.

(3) A letter was read to the patient and he was asked to find it in a group of written letters. He found 14 out of 17 letters, *i.e.*, he scored a better percentage rate (82%) compared with simple reading of letters (40-60%). This auditory facility operated also in reading words.

There was complete verbal alexia (word blindness) affecting both oral and silent reading. The patient was not able to read a single word, no matter how simple. Attempts were made to unblock alexia by the unblocking tests worked out by Weigl and Kreindler (1960).

Visual Unblocking of Concrete Words.—(1) The patient was given a written word which represented the name of an object commonly used or of a phenomenon of nature. At the same time he was shown several pictures. Then he was requested to point to the pictures which the word conveyed. He wrongly read 10 out of 20 words. (2) The patient was shown a picture and asked to designate the adequate word out of several written words. He correctly read 16 out of 20 words.

Auditory Unblocking.—(1) The patient was told a word and instructed to read it from a group containing several written words. He carried out this test faultlessly, making no mistake either as regards concrete or abstract words. (2) A written word was shown to the patient and he was asked to identify it out of several words heard by him. He correctly named 18 out of 20 words in the category of concrete words and all abstract words.

Reading of Sentences proved to be impossible. Facilitating (unblocking) auditory tests were carried out according to the model described for verbal alexia. The patient was told a sentence and instructed to read it out of several written sentences. He was able to read eight out of the nine concrete sentences and four out of the five abstract sentences. (2) He was then shown several written sentence and asked to identify it amongst several sentences heard by him. He recognized all the nine concrete sentences which were given to him to read. Visual and auditory unblocking have a transient effect on lexia, which is strictly limited to their period of action. Reading of punctuation marks was good.

Tactile reading (dermoalexia) was also good. The patient was able to read all the letters and words which were written on his skin.

Proprioceptive reading was unimpaired. The patient was able to identify by means of muscular proprioception all letters given to him. He could also correctly read the letters and words written by passive movements, the examiner leading the patient's hand.

Reading of figures and arabic numbers was less impaired than that of letters or words. He correctly read the numbers from 1 to 10, but misread numbers formed of several figures; there were more mistakes in the reading of complex numbers (Table II). The reading of arithmetic signs was correct. Reading of figures and Roman numbers unfamiliar to the patient was inconclusive.

The patient was able to tell the time correctly without making a single mistake.

Function of Numbers.—The patient correctly understood the number series from 2 to 2, 3 to 3, 4 to 4, 5 to 5, and was able to count faultlessly from 1 to 100. When

TABLE II
READING OF ARABIC NUMERALS

Presented Number	Numerals Read
25	25
76	75
64	64
125	105, 125
304	504, 304
601	201
156	156
1,256	1,436
6,290	5,260, 5,290
14,352	14,525
26,340	23,604

counting backwards, from 100 to 1, he skipped the numbers from 30 to 18, but read the rest correctly.

To test his classification of numbers with regard to their value, the patient was given a couple of numbers having near values and instructed to say which was the bigger and which the smaller. He performed this test correctly.

He gave correctly the number of days in the year, in a week, the number of weeks in a month, in a year, and the number of months in a year, making but a single mistake, *i.e.*, he stated the number of days in a year to be 330 instead of 365.

In giving the number of members of his family and of objects in the room he made no mistake at all.

Mental arithmetic was performed quite correctly, very few mistakes being recorded in elementary addition, subtraction, multiplication, or division (out of 30 tests only four were wrong.)

In solving simple written arithmetical problems involving the same tests, he made 12 mistakes (out of 30), which could be readily accounted for by the reading difficulty for figures.

Writing.—The patient was asked to write a letter to his wife. He wrote well, intelligibly and comparatively

fast but with a few mistakes consisting in the omission of some letters at the end of the words, and of some punctuation marks. In a letter consisting of 42 words he omitted four letters and five punctuation marks.

The patient wrote all the letters of the alphabet to dictation without a mistake. He correctly wrote the words dictated to him, excepting some small errors of the type mentioned for spontaneous writing. He was unable to read what he had written spontaneously or to dictation.

Copied writing was greatly altered, even unintelligible. The patient obediently copied in an '*écriture servile*' the letters and words given him as a model. To copy a word took about three times as long as to write the same word to dictation. A comparison between writing to dictation and copying, the same words being used for both, clearly showed the maintenance of his ability to write to dictation and the severe impairment as regards copying (Fig. 1). Writing of punctuation marks showed no pathological changes. Writing of arabic figures, spontaneously and to dictation, was faultless. Copying arabic figures showed far less disturbance than copying letters or words. The patient could copy eight out of 10 figures. The patient's written arithmetic proved satisfactory. Left-hand writing revealed nothing abnormal.

Writing with closed eyes was possible and showed no qualitative differences compared with writing with open eyes.

Rhythm Tests.—The patient was able faultlessly to perform various simple rhythm tests such as *///*, */// //*, */// //*, */// //*, */// //*, */// //*, */// //*, */// //*, */// //*, */// //*.

Music.—The patient could not read or write musical notes. For this reason, investigation as regards music was very summary. He could distinguish between folk music, classical, and modern music, and was able to recognize different folk tunes as well as some operatic airs.

FIG. 1.—A. Writing to dictation. B. Copied writing (the same words as in writing to dictation).

He would not sing either spontaneously or when requested, stating that he was unable to do so. He unsuccessfully whistled a familiar tune.

Praxis.—The tests for ideatory, ideomotor, facio-bucco-lingual praxis and dressing praxis were performed normally.

Constructional Praxis.—The patient correctly drew a square and a circle but failed to draw a rectangle correctly. He initially outlined a triangle, then drew a rectangle only with the assistance of the examiner. He could not draw geometrical forms in perspective, *i.e.*, instead of a cube he would draw a square. This deficit concerning perspective drawing was again encountered in other drawings (house). He erroneously drew a star with five corners, a bicycle, a man (Fig. 2), a car, but correctly drew a cross and a zigzag line.

The blindfolded patient was instructed to touch various geometrical objects (rectangle, circle, etc.) and then to draw them (Halstead's test). He correctly performed this test. Copied drawing revealed the same deficit in reproducing the element of perspective. Copied drawing was better than that done to command (excepting the sketch of a man, Fig. 2).

The patient was unable to build a cube by means of matches, either to command or if given a model. Nor could he assemble a single puzzle, either with or without a model (Koh's test).

Gnosis.—Gnosis of objects and pictures was very good. The patient was able to recognize the component elements of some complex pictures, some of which had symbolic value: a young man leading an assault of a fortress with flying colours; a sinking ship; a preaching priest; Jesus Christ's descent from the cross.

Right-left differentiation was performed perfectly, both as to his own person and to that of the examiner. Digital gnosis was correct as well as autotopognosis and stereognosis.

Gnosis and Naming of Colours.—On the colour chart he correctly indicated the white, black, green, blue, and

red colours. He mistook yellow and also violet. The other intermediary colours were indicated correctly. The naming of 'concrete' colours (blue like the sky, green like the grass, red like blood) was imperfect.

From a series of samples he was told to select the main colours (Holmgren's test), and chose them at random, in an unsystematic way. He made no mistake in selecting the samples but only chose some of the most vivid colours, refusing to indicate the remaining colours or being unable to find them. He thus assembled only two or four samples each of the main colours. Spontaneous grouping was very unsystematic, the patient grouping heaps of colours that clashed, for instance, green and pink. When grouping was done by the examiner according to basic colours, the patient firmly refused this way of grouping, saying 'different colours have been mixed up, they are not the same, that won't do' but then agreed, saying, 'still, according to the colours it is all right'.

Colours were named correctly only for white and black. The other colours were named erroneously, for instance, green was called light brown, yellow—grey, red—yellow, light blue—grey.

Spatial Gnosis.—The patient had a good orientation as regards immediate visual space and the usual visual space (house, hospital); here he made no mistakes, but he was unable to draw a map of the country, nor could he correctly locate the main towns on the map done by the examiner. He stated he had never been able to do this. He drew a rudimentary and incomplete plan of the room in which he was examined. Distances were estimated correctly (the size of the room, of the house, and the distance between various objects). The patient did not display signs of unilateral spatial inattention (he correctly counted beads uniformly scattered, halved a line correctly, and adequately bisected an angle). He had no difficulty in orientation and in fixing his gaze on a moving object.

Clinical Course

The patient was given antiluetic treatment and anticoagulant drugs (heparin) associated with a hypotensive

FIG. 2.—A. Drawing to command: sketch of a man (notice that both arms were placed to the left according to the unimpaired visual field). B. Copied drawing of a man was infinitely worse than the same drawing done to command.

drug (Serpasil). A month later, he had improved in reading letters (he was able to read 18 letters) and words (he could read some simple words), the disturbances as regards colours had improved and the slight anomia had disappeared.

Discussion

As has been stated by many authors, the terms 'pure' word blindness and 'pure' alexia are inadequate. In all cases reported in the literature as well as in our own case, alexia is associated with other symptoms, such as, for instance, right homonymous hemianopsia, colour agnosia, spatial agnosia, constructional apraxia, anomia, dysgraphia (affecting copying of script), dyscalculia. This association is attributable to the fact that the area subserving reading is common to that subserving other higher functions (colour gnosis, spatial gnosis). Yet, it is no less true that alexia, as in this case, may dominate the clinical picture with a minimal involvement of spoken speech and hence makes it possible to separate alexia from aphasia (Casey and Ettlinger, 1960).

Most authors consider 'pure' word blindness to be the result of a brain injury. Pathological findings are indicative of this. Dejerine (1892) found lesions located at the base of the cuneus and involving the lingual and fusiform gyri. In a case of 'pure' word blindness, Hinshelwood, Macphail, and Ferguson (1904) found an extensive lesion of the left occipital lobe expanding down to the occipital pole. Ross (quoted by Leischner, 1957) carried out an anatomico-clinical study in a case with word blindness and right homonymous hemianopsia in which he found a softening of the grey matter in the left occipital lobe; the gyrus angularis was intact. Pötzl (1919) likewise found left occipital injuries (gyrus lingualis) and lesions of the splenium corporis callosi in two cases of 'pure' word blindness. The same author (1927) reported another case of an 82-year-old alcoholic man, who presented with two episodes of right hemiparesis, right homonymous hemianopsia, and verbal alexia. His patient was able to read letters. Pathological findings disclosed two softenings on the internal face of the left occipital lobe, one of which involved a major part of the gyrus lingualis and penetrated into the lateral ventricle. The other lesion involved a small part of the left calcarine fissure (the portion corresponding to the cuneus).

In a synthetical survey on alexia, De Massary (1932) ascertained that in 'pure' word blindness the lesion prevalently affects the gyrus lingualis and the gyrus fusiformis. Lesions of the fibres of the corpus callosum were also reported in some of the cases examined at necropsy (Foix and Hillemand, 1925;

Kleist, 1934; Gloning, Gloning, Seitelberger, and Tschabitscher, 1955).

In our case there are several arguments in favour of a prevalently left occipital location of the lesion; namely the absence of disturbances of receptive speech; an association with agnosia as regards colours; conservation of spontaneous writing and of writing to dictation, which, as pointed out by Pötzl (1927) and by Ajuriaguerra and Hécaen (1960), are always impaired in parietal alexia. The major clinical symptoms exhibited by our patient suggest a preponderance of the lesion in the peri- and left parastriate areas (areas 18 and 19 of Brodmann). In this sense, the pathological findings reported by Sittig (1921) and by Kleist (1934) are worth mentioning. In their cases of colour agnosia lesions were found in area 19 of Brodmann. Yet, the presence of constructional apraxia and of a discrete anomia points to the parietal involvement of the adjoining zone (Brodmann's areas 40 and 39). The absence of the great syndromes (receptive aphasia, agraphia) generated by lesions of these left parietal areas leads us to suppose that in our case parietal involvement was but slight, the main lesion being the left occipital one.

The comparatively sudden onset, the regressive course, the vascular antecedents suggest a vascular aetiology. The vascular lesions were confined to the posterior cerebral artery. By taking into account the limited nature of the lesion, the existence of a thrombosis of a cortical branch of the posterior cerebral artery seems most likely, perhaps thrombosis of the left lingual artery as asserted by Kroll (quoted by Leischner, 1957).

Ophthalmological findings (Dr. Florentina Lascu) in our patient showed that fixation and scanning on the median line were good. A slight fixation deficit on the hemianopsic side, as expressed by nystagmic movements to the right, was present. The fixation deficit on the hemianopsic side was first reported by Holmes (1921) in brain injuries of the occipital lobe. He accounted for this symptom by the interruption of the corticotectal fibres as demonstrated pathologically. The fact that there were no differences between reading in the right visual field (blind) and the left visual field (unimpaired) and that right homonymous hemianopsia was compensated, the patient displaying correction movements of the head and eyeballs for right homonymous hemianopsia, has led us to attach no pathological importance to the fixation deficit in the right hemianopsic fields for alexia. In this connexion, it must be mentioned that Warrington and Zangwill (1957) attached no importance to fixation disturbances of the eyeballs, except in cases of uncompensated right homonymous hemianopsia, arguing that in this condition, too,

fixation disorders may possibly be secondary to alexia when ocular fixation is difficult owing to the patient's inability to understand the sense of the words he reads. No concentric constriction of sight was observed to exist, which might justify the assertion that there exists a functional lability of the visual system (Beringer and Stein, 1930). At the same time no disturbances of visual orientation were found to be present (Martin, 1954).

In our patient, alexia is the consequence of visual agnosia, displaying a more complex nature and comprising also agnosia for colours and for geometrical forms. It is significant that the graphic-motor picture of the written symbol of language was intact as the patient could write but could not read what he had written. He had the mental image of the graphic sign and could express it by an adequate motor action (graphically) but when seeing the graphic sign could not form the pertinent mental picture. Interior language was very well preserved. The patient was not really aphasic but agnostic for certain symbols and for colours. Only the optic symbol of the letters was altered in our patient, for the symbols conveyed by other pathways, dermolexia, feeling of letters, succeeded in releasing the mechanism enabling him to recognize the symbol preserved as such.

In our case particular importance in understanding the pathophysiological mechanism of alexia must be attached to the tests of visual and auditory unblocking. They demonstrate the possibility of reading words and even sentences with the aid of visual and auditory cues (other than those of the written symbols of speech) when direct perception or understanding of the words or sentences was impossible. These facts suggest, on the other hand, that the pictures of the symbols of language were not destroyed but merely could not be evoked visually. Reading, as emphasized by Leischner (1957), appears to be the result of an association of various functions, i.e., optic, gnostic, optocokinetic, acoustic, verbal, motor, and ideatory functions. Among these functions, the optic and acoustic components are predominant. The total absence of receptive language disturbances afforded us the opportunity efficiently to use the methods of auditory unblocking whereby a high reading score was attained (80-100%) as compared with a direct reading score of 0.

Another convincing test regarding the preservation of the pictures of written language symbols is the

quasi-normal performance of spontaneous writing and writing to dictation.

The fact that visual and auditory facilitation (unblocking) as well as other tactile, proprioceptive facilitations exert a transitory effect on lexia strictly limited to the period of their action, is in favour of the interpretation of 'pure' alexia as a disruption of the functional connexions between the various anatomical structures and pathways upon which normal reading depends.

Summary

A right-handed patient is reported, whose past history included lues and arterial hypertension, and in whom word blindness set in suddenly, associated with right homonymous hemianopsia, dysgraphia in copying, colour agnosia, constructional apraxia, slight anomia but no disturbance of receptive speech.

The use of facilitating (unblocking) auditory and visual tests made possible the reading of certain words and sentences which the patient had been unable to read when presented in isolation.

The role of the various sensory inputs in the mechanism of reading and of the sites of lesions which may evoke 'pure alexia' are discussed.

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