Transtentorial Herniation with Posterior Cerebral Artery Territory Infarction

A New Mechanism of the Syndrome of Alexia Without Agraphia

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SUMMARY  A 39-year-old male developed a right homonymous hemianopsia and alexia without agraphia following emergency surgery for hemorrhage into a left frontal tumor. A computerized tomographic (CT) scan demonstrated low density areas in the left frontal region and in the territory of the left posterior cerebral artery. The alexia without agraphia syndrome appeared to result from compression of the left posterior cerebral artery by a transtentorial pressure cone, a mechanism not previously reported in this syndrome. The behavioral investigation confirmed the diagnosis and replicated recent findings related to the syndrome of alexia without agraphia.

Case Report

This 39 year old, right-handed male consulted an ophthalmologist for headaches and blurred vision. On examination, he was found to have papilledema, but his neurological examination was otherwise normal. Radionuclide brain scan and left carotid arteriography disclosed a large left frontal tumor. Prior to elective surgery, the patient was found comatose with fixed dilated pupils. Emergency craniotomy and resection, and intracranial trauma, surgical resection, and intracranial hemorrhage. We report a case in which alexia without agraphia appeared to follow transtentorial herniation and posterior cerebral artery compression secondary to hemorrhage into a frontal tumor.

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tated word name, a printed word name, and a picture
were compared as facilitators of the patient's ability to
identify a word from a list. Letter naming was mildly
impaired (12/144 errors), but latencies were much
shorter than for words. Latencies increased with
length in both the word naming and lexical decision
tasks indicating that the patient was reading by a
letter-by-letter decoding process. In the cueing test,
the dictated words were more effective than either the
printed word name or picture, indicating the preserva­
tion of auditory language processing. These results
were similar to previous findings in alexia without
agraphia.12

Discussion
The patient's reading disorder appeared typical of
alexia without agraphia in bedside testing, in the
BDAE, and in the experimental test battery. Alexia
with preserved writing, difficulty in naming colors,
short term memory loss, and right homonymous
hemiagnosia are all characteristic of the syndrome.
Letter-by-letter reading and ability to interpret dic­
tated spelled words have also been emphasized.3, 12, 14
The hallmark of alexia without agraphia, however, is
the remarkable ability of the patient to write sentences
to dictation, yet not to read these same sentences.
The pathogenesis of the syndrome in our patient
appeared to involve hemorrhage into the left frontal
tumor, with creation of a transtentorial pressure cone,
compression of the posterior cerebral artery, and in­
farction of the left occipital lobe.
Compression of the posterior cerebral artery and
resultant temporo-occipital infarction has been well
documented in cases of transtentorial herniation.17-19
While survival after transtentorial herniation is un­
usual, cases with both cortical blindness and hemi­
anopsia have been reported.17, 18, 20 Kertesz21 reported
a case of alexia and visual agnosia secondary to severe
head trauma with prolonged coma, but no evidence
for transtentorial herniation or posterior cerebral
artery compression was presented. The behavioral
syndrome of alexia without agraphia has not
previously been described as a sequel of transtentorial
herniation.
The experimental findings of preservation of letter
reading and letter-by-letter decoding of words confirm
previous findings in alexia without agraphia.12, 22, 23
These findings are not easily explained by the tradi­
tional model of alexia without agraphia, which postu­
lates a disconnection of the intact right visual cortex
from left hemisphere language centers, and especially
from the left angular gyrus, which is thought to be a
center for intermodality associations such as the visual
to auditory transcoding needed for reading.3, 24 The
direct site of this disconnection is thought to be the
splenium of the corpus callosum which, along with oc­
cipital lobe, is supplied by the posterior cerebral
artery. Most instances of alexia without agraphia in
The literature have been caused by stroke in the territory of the left posterior cerebral artery.\textsuperscript{1-5} Reports of alexia without agraphia in other pathological conditions affecting the left occipital lobe — including tumors,\textsuperscript{6-9} surgical resection,\textsuperscript{10} trauma,\textsuperscript{10-12} and intracerebral hemorrhage,\textsuperscript{3} all of which should not affect the corpus callosum — suggest that anatomical disconnection may not fully account for the syndrome. Others have suggested that an occipital white matter lesion alone may be sufficient to disconnect the right visual cortex,\textsuperscript{7} or even both visual cortical areas,\textsuperscript{26} from the angular gyrus. The experimental and clinical findings in alexia without agraphia can also be explained by a disturbance of a more fundamental visual process such as the ability to perceive more than one language stimulus simultaneously\textsuperscript{3} or the ability to recognize a "visual word form".\textsuperscript{23} The status of these competing theories regarding the mechanisms of alexia without agraphia will have to await the verdicts of future research.

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