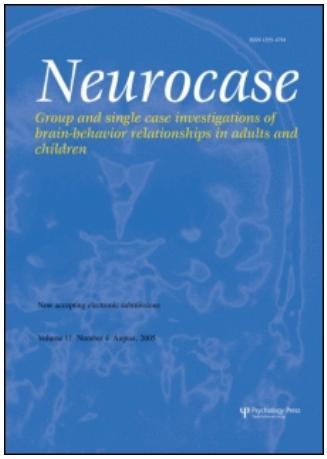

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Relative preservation of 'animate' knowledge in an atypical presentation of herpes simplex virus encephalitis

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A comprehensive battery of neuropsychological tests designed to assess primary cognitive functions, including language and semantic memory, was given to MG, a patient with confirmed herpes simplex virus encephalitis. MG's initial jargon aphasia resolved over time to leave her with a mild phonological impairment. She had a very mild amnesia that was worse for verbal material and a category-specific impairment of semantic memory. This latter impairment resulted in a significant anomia that was worse for manmade/artefact items than for animate kinds. Her naming difficulties were associated with a mild impairment in comprehension that was not specific to category or feature type. MRI revealed a strongly asymmetric and atypical distribution of pathology in MG with the disease affecting the left medial temporal lobe, temporal pole, left frontotemporal and temporoparietal regions.

Introduction

Neuropathological findings in acute herpes simplex virus encephalitis (HSVE) are variable but the characteristic picture of the disease is associated with focal necrosis in the medial temporal lobes, hippocampus, the cingulate, and insular cortex and often involves more lateral temporal lobe regions as well as the orbital surfaces of the frontal lobes (Hierons *et al.,* 1978; Kennedy and Chaudhuri, 2002). Limited damage may also occur, but more rarely so, in the parietal and occipital lobes (Hierons *et al.,* 1978; Kapur *et al.*, 1994). Although HSVE cases with unilateral damage have been noted before (Tranel *et al.*, 1997) previous reviews have highlighted bilateral temporal lobe involvement as the pattern that is strongly associated with this disease. The distribution of pathology is thought to result from entry of the virus via the olfactory pathway or the trigeminal ganglia to the temporal lobes (Kennedy and Chaudhuri, 2002). A study of 42 patients (McGrath *et al*., 1997) showed that the most common presenting symptoms in HSVE were: headache, confusion, nausea, fever, seizures and drowsiness. Since the 1980s, anti-viral treatment has altered the prognosis of HSVE. If treatment is given within 4 days of the onset of the symptoms the brain lesions are smaller and less widespread, leading to an increased variability in the cognitive profiles of survivors (McGrath *et al*., 1997; Whitley *et al*., 1987).

In this study we present the profile of patient MG, whose encephalitis was atypical in two important aspects: (1) like a small number of other patients (Tranel *et al.,*1997) MG's damage was limited to the left hemisphere; and (2), although she did have the typical medial temporal lobe involvement on the left, the distribution of pathology extended from the temporal pole to include left frontoparietal and perisylvian regions. Her atypical distribution of damage was matched by a unique and similarly atypical neuropsychological profile. She presented in the acute phase with jargon aphasia and following a period of recovery, this resolved to severe naming difficulties and a mild comprehension deficit consistent with a Wernicke's type aphasia plus a very mild amnesia restricted to verbal material. While this neuropsychological/aphasiological picture is consistent with the distribution of her pathology, it is atypical for the profile of HSVE as presented in various review articles (Kapur *et al*., 1994; Kennedy and Chaudhuri, 2002). The most common cognitive impairment is anterograde amnesia of varying severity (Warrington and Shallice, 1984; Gordon *et al*., 1990; Kapur *et al*., 1994; Hokkanen *et al.,* 1996; McGrath *et al*., 1997; Utley *et al.*, 1997). Some patients have been described with a dense retrograde amnesia (Warrington and McCarthy, 1983; Kopelman *et al.,* 1999) but loss of memories is usually confined to a short period prior to the disease onset (Parkin, 1984). Semantic memory deficits, anomia and acquired surface dyslexia are also commonly reported in HSVE patients, again with considerable variability in severity (Warrington and McCarthy, 1984; Pietrini *et al*., 1988; Stewart *et al.,* 1992). HSVE patients whose damage has affected the orbitofrontal cortex may exhibit behavioral disturbances, including impulsivity, perseveration of action and emotional lability or rigidity (Hokkanen and Launes, 2000).

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In the cognitive neuropsychological literature, HSVE patients have been studied largely because of the category-specific semantic memory impairment that can occur in these patients. In 1984, Warrington and Shallice described four HSVE patients all of whom showed gross deficits when required to identify living things and foods but were less impaired in the identification of man-made artefacts. Following the publication of this seminal paper, there have been a considerable number of reports of HSVE patients who show a similar discrepancy in their knowledge favouring man-made/inanimate over living/animate things (Pietrini *et al*., 1988; Sartori and Job, 1988; Sartori *et al*., 1993; Capitani *et al*., 1994; DeRenzi and Lucchelli, 1994; Barbarotto *et al.,* 1996; Gainotti and Silveri, 1996; Laiacona *et al.,* 1997; Borgo and Shallice, 2001). As far as we are aware, patient MG reported here is the first HSVE patient to have been comprehensively described with a relative sparing of knowledge and naming of animate categories.

This picture of category-specific impairment for artefacts with relative preservation of living things is comparatively rare in all patient groups (around 50–60 patients impaired on living categories compared to10-20 for non-living; see Gainotti, 2000; Capitani *et al*., 2003 for reviews of the clinical evidence for category-specificity). The demonstration of the double dissociation is important because it rules out a simple interpretation of category-specific disorders in terms of differential difficulty and, where the same stimuli have been used across patients, an explanation based on confounding variables such as familiarity, frequency, and so on (though it might follow if patients' performance is affected by two different variables such as age-of-acquisition and familiarity (Lambon Ralph *et al*., 1998). The majority of the patients who show this type of category specificity have suffered CVA (Warrington and McCarthy, 1983; Warrington and McCarthy, 1987; Hillis *et al*., 1990; Hillis and Caramazza, 1991; Sacchett and Humphreys, 1992; Cappa *et al*., 1998). The literature also contains two cases with semantic dementia (Lambon Ralph *et al*., 1998; Silveri *et al*., 1997) and there have been some reports in dementia of Alzheimer's type. (Gonnerman *et al*., 1997; Garrard *et al*., 1998; Laiacona *et al*., 1998;). The deficits affecting non-living items in these studies have been linked with lesions to fronto-parietal regions resulting from ischaemia in the area of the middle cerebral artery, in most cases, or pathology focused on the parietal region (Warrington and McCarthy, 1983; Hillis *et al*., 1990; Sacchett and Humphreys, 1992; Garrard *et al*., 1998). Other patients had some temporal lobe involvement in addition to fronto-parietal damage (Silveri *et al*, 1997; Warrington and McCarthy, 1994) while a few cases had solely left temporal lesions. (Hillis and Caramazza, 1991; Cappa *et al*., 1998; Lambon Ralph *et al*., 1998).

In this article we present a single case study of an HSVE patient, MG, who had a profound, jargon aphasia on admittance to hospital. Early language assessment revealed severe phonological disturbance on expressive tasks. Subsequent performance on two naming tasks (that have been used to demonstrate a naming impairment for living things in other patients) showed a naming deficit that was significantly greater for man-made items. We discuss this unusual presentation in relation to MG's atypical HSVE neuropathological profile and other neuropsychological results.

Case report

Background: Neurological

MG, a 57 year-old, right-handed secretary with 11 years education, was admitted to Queen's Medical Centre, University Hospital, Nottingham with fever, confusion and jargon aphasia. Viral serology analysis of CSF and a CT scan confirmed a diagnosis of HSVE. Anti-viral treatment was administered. A CT scan was performed 3 weeks after the patient was admitted to hospital and showed change in the left temporal and perisylvian region. In March 1999, 21 months post-onset, an MRI scan revealed widespread high-signal intensity changes in the fronto-temporal and temporo-parietal regions on the left side. No cortical changes were noted in the right hemisphere (see Figures 1 and 2—Coronal and axial sections showing unilateral damage to the left hemisphere).

Background: Neuropsychological

MG presented with a primary, atypical symptom of jargon aphasia. Her speech was rapid, fluent but incomprehensible and it was this speech pattern that had alerted MG's husband to the severity of her otherwise influenza-like symptoms.

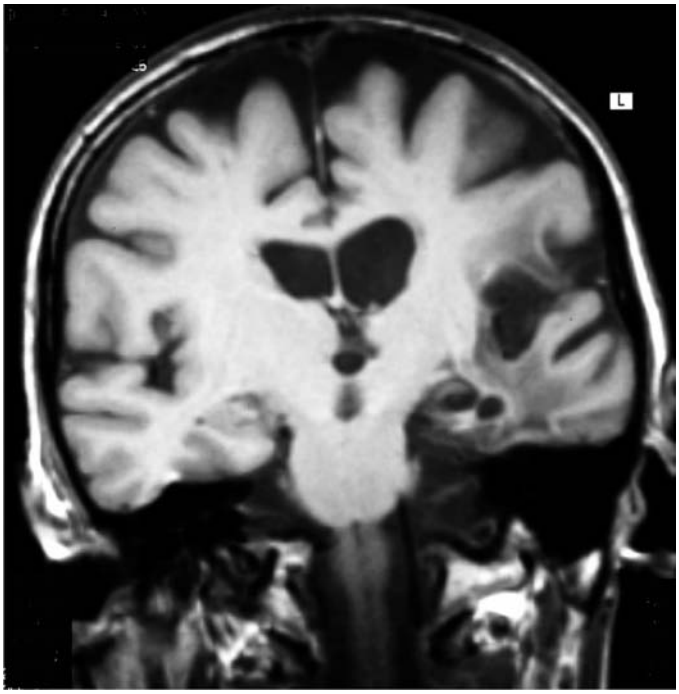

Fig. 1. Coronal section showing left medial temporal and temporal lobe atrophy.

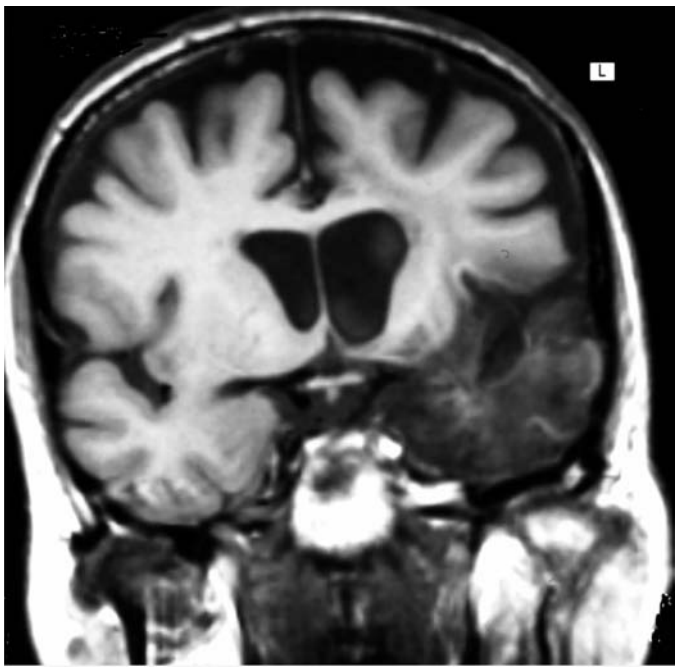

Fig. 2. Coronal section showing atrophy in the left temporal pole.

This aphasia was still present, although reduced in severity, when MG was assessed by a clinical psychologist four months post hospital admittance This assessment revealed that MG was oriented for time and place and could give details of her remote personal history, although this was limited by her aphasia. She co-operated fully with assessment and showed no signs of behavioral disturbance. Her performance on the vocabulary subtest of the WAIS (Wechsler, 1998) revealed a mild, generalized loss of vocabulary. She had severe naming difficulties; she failed to name any of the pictures from the Graded Naming Test (McKenna & Warrington, 1983) and her word fluency was very low. In contrast her performance on the Trail Making Test (Reitan & Wolfson, 1993) and Raven's Colour Matrices (Raven, 1962)was within normal limits suggesting good executive skills. It was difficult to measure MG's verbal recall performance because of her aphasia but there was evidence of a retention-impairment. Her performance on the Rey Complex Figure (Osterreith, 1944) was impaired but there was evidence of retention. Her verbal recognition memory was severely impaired (29/50, <5th percentile) but her face recognition was normal (47/50, 95th percentile; Recognition Memory Test; Warrington, 1984). MG's resolving aphasia was tracked over a period of 18 months with tests taken from the Psycholinguistic Assessments of Language Processing in Aphasia (PALPA; Kay *et al.,*1992). At the end of this period, MG's phonological disturbance was mild enough to reveal a category-specific naming impairment and, accordingly, a more detailed assessment of her semantic memory was carried out.

MG's atypical neuropsychological presentation

Changing language profile during the acute phase

Phonological tasks. MG's performance on a range of language assessments drawn from the PALPA battery was measured between 2 and 3 months post admittance (shown in Table 1). As well as producing phonological errors in her spontaneous speech, MG made similar errors on a number of other simple, single word assessments, consistent with a generalized phonological impairment. Although her reading accuracy was unaffected by imageability or frequency, her

Table 1. MG's language performance during the acute phase

Test	Performance	Control performance
Reading and spelling		
PALPA 4 - phonological discrimination (picture)	35/40*	39/40 (SD = 1.7)
PALPA 5 - phonological discrimination (word)	66/80*	76/80 (SD = 4.3)
PALPA 28 – written homophones	56/60	55/60 (SD = 2.77)
PALPA 31 – Reading aloud	74/80*	79/80 (SD = 0.13)
PALPA 36 - non-word reading	20/24	23/24 (SD = 2.9)
PALPA 40 – spelling	25/40*	36/40 (SD = 4.8)
Repetition and phonological manipulation		
PALPA 8 – non-word repetition	30/30	not available
PALPA 9 – word repetition	80/80	79/80 (SD = 2.7)
Naming		
PALPA 53 – picture naming		not available
Time 1 (1 month post-illness)	0/20*	
Time 2 (3 months post-illness)	0/13*	
Time 3 (5 months post-illness)	18/40*	
PALPA 14 – rhyme judgements from picture stimuli	1/5* (abandoned)	not available

*denotes impaired performance

performance on word reading was slightly weak and contained a number of phonologically-related errors (e.g., SQUIRREL → "quarrel", SQUINT → "quint", PUMP → "bumb"). Her reading of nonwords fell into the normal range and she was able to make homophone judgements about written words. MG's phonological impairment also led to a spelling deficit and her accuracy in this domain was modulated by word frequency (PALPA 40). MG's phonological impairment was sufficiently mild at this late stage to allow normal repetition of simple words and nonwords (PALPA 8 & 9) as well as good performance on phonemic and segmentation tasks (PALPA 16 & 17). On phoneme discriminations to picture or word stimuli (PALPA 4 & 5), however, her accuracy fell below the normal range.

Naming. The first set of naming tests was administered one month after her admittance to hospital. MG's initial picture naming was at floor (PALPA 53) and her errors were a combination of omissions and phonologically related responses. One month following this initial assessment, MG's naming was still at floor but when tested on the same stimuli five months post admittance her performance had improved dramatically, though remaining impaired. At this stage MG's phonological impairment had resolved sufficiently that 15/18 responses were correct on her first attempt, with the three remaining responses requiring an incremental build-up of phonology (conduit d'approche: e.g., ELEPHANT → "tellybird…..tellyman….telefan….elephant"; GLOVE → "gl…glds…glove" and COW → "kezt….kau…then c..o..w.") Of the 12 incorrect responses, two were omissions and all the others were characterized by a phonological build-up to unrelated names (e.g. SCREW → "port-thing…/po:n"; BREAD → "f/../f/../fr/../freezer/"; and WATCH → "/ma:/ /m/….matthew"). MG's performance on the Boston Naming Test (Kaplan *et al.,* 1976) was at a similar level of impairment (10/49) and her errors were primarily phonological. MG was unable to make rhyme judgements from pictures (PALPA 14) because of her inability to access the names of the stimuli.

Comprehension. MG's semantic impairment in the first few months of her recovery was highlighted by synonym judgement and semantic association tests (PALPA 49 & 51). Her performance was particularly weak for abstract concepts. On the Pyramids and Palm Trees Test of associative semantics (Howard and Patterson, 1992) MG showed a mild impairment for picture stimuli but was within the normal range for word stimuli. At three months post-onset MG's spoken and written word-to-picture matching (PALPA 47 &48) was severely impaired, with omission errors predominating. However, her performance on these tests also improved over time such that after five months her accuracy was within the normal range on spoken word-to-picture matching and slightly impaired on the written form. (See Table 2 for a summary of MG's scores on these tests).

Stable performance in the chronic phase (18 months post onset)

Visuo-perceptual performance. MG's performance on the Birmingham Object Recognition Battery (BORB; Riddoch and Humphreys, 1993) fell within, but at the lower end, of the normal range (54/62 easy and difficult subtests). She performed at ceiling for all but the object decision subtest (17/20) of the Visual Object and Space Perception Battery (VOSP; Warrington and James, 1991); the silhouette subtests were not administered).

Category-specific semantic impairment

Naming. A summary of MG's naming performance, including types of error, is shown in Table 3. Initial assessments of

Table 2. Comprehension in the acute stage

Test	Performance	Control performance
PALPA 49 – spoken synonym judgement	High-imageability 27/30 Low-imageability 21/30	not available
PALPA 51 – written semantic relatedness	High-imageability 12/15* Low-imageability 8/15*	13/15 (SD =1.26) 12/15 (SD = 1.82)
Pyramids and Palm Trees		
Pictures	47/52*	51/52 (SD = 1.1)
Words	50/52	51/52 (SD = 1.4)
PALPA 47 – spoken word – picture matching		
Time 1 (1 month post-illness)	25/40*	39/40 (SD = 1.07)
Time 2 (3 months post-illness)	38/40	39/40 (SD = 1.01)
PALPA 48 – written word – picture matching		
Time 1 (1 month post-illness)	25/40*	
Time 2 (3 months post-illness)	33/40*	

*denotes performance outside normal range

Table 3. MG's naming performance: Chronic phase (after 18 months recovery)

		Error types		
Test	Accuracy	Semantic associate	Gesture / verbal description	Omission
'*Matched' set*				
Animals	24/32 (75%)	8	0	0
Artefacts	13/32 (41%)	5	5	7
	$\chi^2 = 7.75$, p = .005			
100 Object Naming				
Animals	31/37 (84%)	2	2	2
Fruit, vegetables & body parts	13/13 (100%)	0	0	0
Living total	44/50 (88%)			
Man-made total	31/50 (62%)	5	7	7
	$\chi^2 = 7.68$, p = .005			
PALPA – Picture Naming				
Animal	6	2	2	0
Fruit & vegetables	4	1	1	0
Living total	10/16 (63%)			
Man-made total	9/24 (38%)	6	4	5
	$\chi^2 = 3.4$, p = .06			
Snodgrass & Vanderwart				
Living:				
Animals	38/54 (70%)	7	2	7
Fruit	8/11 (73%)	2	0	1
Vegetables	6/12 (50%)	1	4	1
Body parts	10/11 (91%)	1	0	0
Total	62/88 (70%)			
Non-living:				
Tools	1/10 (10%)	0	4	5
Musical instruments	5/9 (56%)	0	2	2
Vehicles	6/9 (67%)	1	0	2
Buildings	2/4 (50%)	1	0	1
Clothing	19/20 (95%)	0	1	0
Household/furniture	38/56 (68%)	9	5	4
General	25/39 (64%)	4	3	7
Total	96/147 (65%)			
	χ^2 n.s.			
Gainotti & Silveri Naming Test				
Animals	8/11 (73%)	Na.	Na.	Na.
Fruit & vegetables	13/19 (68%)			
Living total	21/30 (70%)			
Non-living total	19/30 (63%)			
	χ^2 n.s.			
Category-Specific Names Test				
Animals	12(10^{th} percentile)	4	2	12
Fruit & vegetables	5($<5^{th}$ percentile)	4	3	18
Non-praxic artefacts	4($<5^{th}$ percentile)	0	10	16
Praxic artifacts	3($<5^{th}$ percentile)	0	18	7
Total living:	16/60 (27%)			
Total non-living:	7/60 (12%)			

MG's comprehension and naming had hinted at a greater impairment for artefacts. We conducted, therefore, a more detailed analysis of her naming at 18 months post onset. MG showed a significant category-specific deficit in favor of animals and animate categories on a number of naming assessments. On a test of 64 items (32 animals and 32 manmade items, matched for concept familiarity, imageability and word frequency taken from Lambon Ralph *et al*. [(1998)], MG

showed a significant effect of semantic category with better performance on the animal items. The same pattern was seen in MG's performance on a second naming task, the 100-item naming test (Kaplan *et al.*, 1976) that includes fruit, vegetables and body parts in the living category. Of the 100 pictures, objective ratings for animacy, age of acquisition, familiarity, frequency, visual complexity and imageability were available for 78 of the items. A simultaneous logistic regression analysis showed that animacy and frequency predicted MG's naming accuracy (Wald = 8.21, p = 0.004 and Wald = 6.07, p = 0.014, respectively). When MG was given the PALPA picture naming task for the third time, her performance was relatively better for the living things (10/16: 63%) than for manmade categories (9/24: 38%), a difference that approached significance ($\chi^2 = 3.3$, p = 0.06). Her performance did not differ for the living and nonliving categories on the Gainotti and Silveri (1996) picture naming task. Because her category effect was not apparent across all these naming tests, we asked her to name the much larger number of Snodgrass and Vanderwart (1980) stimuli. On this set, MG named 64% of animate and 49% of inanimate items correctly. Objective ratings for age of acquisition, familiarity, frequency, imageability, visual complexity and animacy were available for 219 of the 260 Snodgrass and Vanderwart pictures. Results from a logistic regression analysis showed that animacy (animals vs. others: Wald = 8.2, p = 0.004) and frequency (Wald = 6.1, p = 0.01) predicted her naming accuracy. MG was also asked to complete the very stringent, psychometrically-graded Category-Specific Names Test (McKenna, 1997). Like most anomic patients on this hard test, her scores on the fruit and vegetables, praxic and non-praxic categories fell below the 5th percentile. Her animal naming was relatively better, falling in the 10th percentile. This naming test is unusual in that norms for discrepancy scores are published alongside control performance for each category. The difference between MG's scores for living and nonliving items (17/60 vs. 7/60) was abnormal: a discrepancy score of 10 or more in this direction occurs in less than 1% of the control population (McKenna and Parry, 1994). In summary, these naming tests show that her performance was relatively better for animate concepts than for fruit and vegetables or manmade items.

Comprehension. MG's results on a variety of comprehension tests are summarized in Table 4. These tests were administered, along with the naming battery, following the 18-month recovery period. Her performance on the PALPA spoken and written word to picture matching tests, that had been impaired when first tested, reached normal levels. MG's performance on sentence-to-picture matching (PALPA 55) was also normal at this time. At this stage MG's comprehension was also tested on a more thorough word-to-picture matching assessment because anecdotal evidence from her husband suggested that she had a mild semantic impairment. (Lambon Ralph *et al*., 1998) The task uses the same 100 concepts as for the naming test described above. Each target picture is presented with four close semantic foils. Subjects are required to pick which picture matches the spoken name. MG's score fell below the range for a group of elderly controls (Lambon Ralph *et al*., 1998).

According to the sensory-functional theory (SFT; Caramazza and Shelton, 1998), MG's poorer naming performance for manmade artefacts should be coupled with poorer comprehension of functional/associative features. Differential knowledge of sensory and functional features has rarely been probed in the few patients who have a relative preservation for animate concepts, although the SFT clearly predicts that these cases should demonstrate relatively poor functional knowledge. In order to test this hypothesis a definition-to-word matching test (Lambon Ralph *et al*., 1998) was used. In this task two definitions are presented (both in spoken and written form) for each of the 64 items used for picture naming (see above), one that emphasizes sensory information and another that contains functional-associative attributes. The patient has to pick one of five semantically related names that matches the definition. Although her overall score was abnormal, there was no difference between MG's performance

Table 4. Tests showing MG's comprehension performance (after 18 months recovery)

Test	Performance	Control performance
PALPA 47 – spoken word – picture matching	39/40	39/40 (SD =1.07)
PALPA 48 – written word – picture matching	39/40	39/40 (SD =1.01)
100 Picture – spoken word – picture matching	93/100*	96-100
PALPA 55 – sentence - picture matching	58/60	57/60 (SD = 4.01)
Definition – word matching		
Animal –perceptual	11/16	not available
Animal – associative	10/16	
Manmade – perceptual	9/12	
Manmade – associative	10/12	
Definition – naming (total)	40/56*	54-56

*

when matching by associative-functional or sensory-perceptual definitions.

Discussion

We have presented data collected from a patient with herpes simplex virus encephalitis who was atypical both in her neuropathological and neuropsychological profiles. MG's distribution of unilateral, left fronto-temporal, temporo-parietal pathology and primary presentation of jargon aphasia is relatively rare in this disease (Tranel *et al.* [1997] reported 5 HSE patients with damage restricted to one hemisphere but no further details about these patients are presented). As far as we are aware, she is the first HSVE patient to be described with a category-specific impairment favoring animate concepts over other categories. This study makes two important contributions; it has implications for the diagnosis of HSVE and for the interpretation of category-specific impairments in terms of brain lesion site and neuropsychological profile.

Implications for diagnosis

Previous reports highlighting atypical HSVE have been at the post-presentation phase and have described cases of atypical MRI imaging (Harrison *et al*., 2003) or atypical virology and neuropathological findings (Klapper *et al.*, 1984; Rose *et al.*, 1992; Fodor *et al.*, 1998). Reports of atypical symptom presentation are equally vital for the accurate recognition, diagnosis and treatment of HSVE. This study is important in highlighting an atypical clinical presentation (jargon aphasia) along with atypical MRI imaging. Recognition of an aphasic presentation as a possible consequence of HSVE is important for early and accurate diagnosis, and for minimizing the delay before anti-viral treatment commences.

Category-specificity

MG showed a category-specific deficit with a relative, but by no means absolute, preservation of animate items. This pattern of impairment has not been previously described in the context of HSVE. Where category-specific impairments have been documented in HSVE, they have always favored manmade items over living categories (Pietrini *et al.*, 1988; Sartori and Job, 1988; Sartori *et al*., 1993, Capitani *et al.*, 1994; DeRenzi and Lucchelli, 1994; Barbarotto *et al.,* 1996; Gainotti and Silveri, 1996; Laiacona, *et al.*, 1997; Borgo and Shallice, 2001). It has become clear in the literature on semantic memory that the exact location of damage is very important in determining the direction of any category-specific impairment (Garrard *et al*., 1998; Lambon Ralph *et al*., 2003). For example, a recent review (Capitani *et al*., 2003) highlighted the fact that cases who show category-specific impairments for living things tend to have bilateral but asymmetric temporal lobe damage, usually greater on the left. In contrast, the majority of patients who show an advantage for living things tend to have damage to the left temporal lobe that extends into parietal and fronto-parietal areas (Garrard *et al*., 1998; Lambon Ralph *et al*., 2003; Lu *et al*., 2002; Perani *et al*., 1995; Tranel *et al.,* 1997). So although MG's neuropsychology and distribution of pathology are relatively atypical for HSVE, the combination of a preservation of animate items and left front-temporal and temporo-parietal damage is consistent with other patients in the literature with this type of category-specificity.

Disparity between MG's anomia and comprehension

Although MG did have a clear yet mild semantic impairment, her anomia across categories was considerably more marked, probably reflecting the combined effect of semantic and phonological impairments (Lambon Ralph, Sage and Roberts, 2000). MG's naming performance improved over time largely in line with the improvement in phonological processing and we believe that the residual deficit following this change reflected the underlying semantic impairment. In addition, MG's category-specific pattern was most marked in her naming performance. Category-specific naming impairments have been reported in other patients who appear to show no deficits in receptive/comprehension tasks (Hart *et al.,* 1985; Farah and Wallace, 1992; Cappa *et al*., 1998) and this has led some authors to conclude that there may be category-specific dissociations in the lexical system itself (Damasio *et al.,*1996; Silveri *et al*., 1997). This interpretation is plausible if it is assumed that naming and comprehension tasks are equally sensitive to a mild semantic impairment. However, it has been argued that demands of speech production are such that it is very sensitive to mild semantic impairment. (Gainotti *et al*., 1995; Lambon Ralph *et al*., 2001) Likewise, Laiacona and Capitani (2001) argued that evidence from patients who appear to have category-specific lexical access impairments should be viewed with caution because of a possibility of the "presence of subtle semantic defects not detected during examination" (page 62).

Evidence from other patient studies lends support to the interpretation of MG's performance as resulting from a deficit in a semantic system rather than at a lexical level. Gainotti (2000) suggested that the fronto-parietal region of the left hemisphere is crucial in contributing to the semantic representation of man-made objects. Tranel, Damasio and Damasio (1997) reported that unilaterally brain damaged patients with abnormal retrieval of concepts/names for tools had lesion overlap that was maximal in the left-occipital-temporal-parietal junction. These results are entirely consistent with MG's lesion and category-specific performance.

The possibility that MG's category-specific pattern to her naming performance is underpinned by a semantic impairment is also supported by a longitudinal and cross-sectional study of semantic dementia. This study was able to demonstrate that the degree of anomia was related not only to the degree of damage to the semantic system but also to the distribution of

that damage. Damage to left hemisphere, semantically related areas produced a much greater degree of anomia than corresponding right hemisphere damage (Lambon Ralph *et al*., 2001). It is entirely possible, therefore, that a mild, category-specific semantic impairment underpinned MG's performance which was readily detected in her naming performance due to the sensitivity of this system and the left-lateralised distribution of her pathology.

Alternative accounts of category-specificity

The sensory-functional theory (SFT) of category-specificity is based on the notion that categories vary in reliance on different types of semantic feature. Animate concepts are assumed to require differentiation by sensory information while artefacts are separated according to function (Warrington and Shallice, 1984). This predicts that poor performance for animate concepts should be associated with loss of sensory information while deficits for artefacts should be coupled with poor knowledge of function. Some early studies of patients with HSVE and category-specific deficits for animate concepts did find evidence of this predicted association (e.g., Gainotti and Silveri, 1996). In the last few years, however, there has been an increasing number of cases reported who do not fit with the SFT. At least seven studies have reported patients with a living things deficit without the predicted differential impairment to sensory knowledge. (Farah & McClelland, 1991; Laiacona *et al.,* 1993; Sheridan and Humphreys, 1993; Funnell and De Mornay Davies, 1996; Lambon Ralph *et al*., 1998; Moss *et al*., 1998; Samson e*t al.*, 1998). Another problem for the SFT is that patients with semantic dementia (SD) who suffer from a progressive deterioration of knowledge about the meanings of words, concepts and objects (Snowden *et al.*, 1989; Hodges *et al*., 1992) do show poor knowledge of perceptual features but very rarely show a category-specific deficit for living things (Parkin, 1993; Cardebat *et al*., 1996; Lambon Ralph *et al*., 1999; Lambon Ralph *et al.*, 2003). MG's performance on the feature knowledge task can add to this debate because it is rare (certainly largely unreported) for patients with an advantage for living things to be thoroughly tested on their feature knowledge. The definition-to-word matching tasks used in this study found that MG's feature knowledge was impaired overall but did not show the predicted difference favouring sensory knowledge.

An earlier account of category-specific impairments was based on the influence of variables such as concept familiarity, word frequency, age of acquisition and visual complexity on task performance (Funnel and Sheridan, 1992; Stewart *et al.,* 1992; Howard *et al*., 1995). It was suggested that apparent category-specific effects could be removed if one or more of these variables were controlled. This account cannot be a complete explanation of category-specificity because there are now a number of reports of category-specific patients in which these variables have been carefully controlled (see Capitani *et al*., 2003). However, these counter cases are almost exclusively focused on patients with deficits for living things and the potential role of frequency, familiarity and visual complexity. Very few studies have addressed the possibility that poor knowledge of artefacts may be associated with the impact of other variables (Howard *et al*., 1995; Lambon Ralph *et al*., 1998). While frequency, familiarity and visual complexity favor artefacts over animate concepts, other variables, including age of acquisition and imageability have the opposite correlation. In addition, aphasic patients' (who often have left temporo-parietal lesions) language performance is often modulated by these same variables (Nickels and Howard, 1995; Ellis *et al.,* 1996). It is possible, therefore, that preservation of animate concepts actually reflects the influence of age of acquisition and imageability. The detailed analyses of MG's naming performance, which controlled for these and other variables, seem to rule out the possibility, at least in this specific patient. MG showed a category-specific deficit on the controlled-set naming task and animacy predicted performance on two larger naming tests even when various factors were included in the regression model.

Fig. 3. Axial Section showing left fronto-temporal, temporo-parietal atrophy.

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