A WOLF IN SHEEP’S CLOTHING: WHEN A STROKE IS NOT A STROKE, AND HOW COMPREHENSIVE SPEECH AND LANGUAGE THERAPY ASSESSMENT AND INTERVENTION CAN HELP TO SET THE ALARM

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Abstract: A 70-year-old male patient, who was diagnosed with human immunodeficiency virus (HIV) in 2001 and did not undergo antiretroviral therapy, was admitted two months after the onset of left cortical-subcortical ischemic stroke, with involvement of the corona radiata and the left thalamus. As a consequence of this vascular event, he suffered aphasia, severe dysarthria, dysphagia, and right hemiparesis.

The patient took part in a rehabilitation program at a rehabilitation center for neurological diseases with a specialized interdisciplinary rehabilitation team. He underwent speech therapy intervention with a frequency of 2-3 hours per day.

Despite the intensity of the program, there was a decline in his clinical and functional status during hospitalisation, with decreased capacity to swallow and communicate (either because of the exacerbation of the aphasia, or due to the worsening of dysarthria and apraxic features). Due to this global deterioration, additional imaging, as well as serological and aetiological examination was performed, which led to a diagnosis of progressive multifocal leukoencephalopathy in the context of HIV1 infection. Antiretroviral therapy was administered to reverse the symptoms.

This case illustrates the relevance of clinical disclosure for the establishment of appropriate functional prognoses and discharge planning. It also shows that the initial clinical features can be misleading and can lead to concealment of the real aetiology, as well as delays in appropriate treatment, especially in multipathology patients.

Keywords: case report, aphasia; dysphagia, rehabilitation, progressive multifocal leukoencephalopathy, speech therapy, HIV, stroke

INTRODUCTION

Human immunodeficiency virus (HIV) infection results in a wide variety of clinical manifestations, on a spectrum that ranges from asymptomatic infection to severe immunodeficiency associated with opportunistic infections (Sterling & Chaisson, 2020). In infected people, viral replication causes a gradual decrease in cell-mediated immunity until its eventual inhibition, leading to several manifestations of opportunistic diseases (Sterling & Chaisson, 2020).

The clinical spectrum of untreated HIV begins with the primary infection (acute retroviral syndrome), evolving to asymptomatic infection, infection with initial symptoms, advanced immunodeficiency with opportunistic complications, and, ultimately, death (Sterling & Chaisson, 2020).

Neurological diseases manifested by individuals infected with HIV can be divided into two types: those that result directly from the action of HIV (primary neurological diseases), which are harder to diagnose due to their insidious onset and because they mimic the side effects of medication; and those that are a direct consequence of immunosuppression (Kallail, Downs, & Scherz, 2008). The latter include opportunistic neurological infections, which are more often a consequence of...
the spread or worsening of a latent, persistent in-
fection, than a manifestation of a recently acquired
one. These infections are not common in individ-
uals undergoing antiretroviral therapy (Bensalem
& Berger, 2002; Clifford, 2017; Power, Boissé,
Rourke, & Gill, 2009). Among opportunistic neu-
rological infections, progressive multifocal leuko-
encephalopathy (PML) is a viral infection in the
central nervous system (CNS) caused by the John
Cunningham (JC) virus and it affects individuals
with a compromised immune system, whether
due to immunosuppression, immunomodulation,
or the presence of a pre-existing disease (Grebenciucova & Berger, 2018; Lima, Bernal-Cano, Clifford, Gandhi, & Koralnik, 2010). Not surpris-
ingly, about 82% of PML patients are individuals
infected with the HIV virus (Molloy & Calabrese,
2009). The JC virus is generally inactive and be-
comes a concern when it infects the host’s CNS
(Grebenciucova & Berger, 2018; Moreh, Israel,
Korem, & Meiner, 2017). In an immunocompro-
mised patient, the latent virus is reactivated and
its genetic code invades the CNS, infecting oligo-
dendrocytes (Bensalem & Berger, 2002; Chahin,
Weber, & Berger, 2016; Grebenciucova & Berger,
2018; Siddiqi, Agnihotri, & Koralnik, 2020) and
astrocytes (Chahin et al., 2016), causing focal
CNS demyelination (Chahin et al., 2016; Moreh
et al., 2017; Siddiqi et al., 2020).

CASE STUDY

The subject of this case report is a 70-year-old
male patient, a Portuguese native speaker (first
language) who completed 12th grade education,
is currently retired from a previous job as a sales-
man and living with his wife. He has a personal
history of arterial hypertension and obesity, and
he was diagnosed with HIV in 2001 and had nev-
er undergone antiretroviral therapy. On May 6th,
2021, he went to a health clinic due to a three-
day sudden onset of stroke, with progressive
decreased strength in the right lower limb, facial
paresis, and dysarthria. He was transferred to the
hospital emergency department in the hospital
and presented with a Glasgow Coma Scale value
of 15 (O4V5M6), right central facial paresis, and
centred tongue at admission. Despite the fact that
the patient had been infected with HIV for more
than 20 years, he omitted that information from
closest family members and hospital staff.

A CT scan was performed, according to which
“There are no unequivocal acute intracranial, in-
tra- or extra- axial pathological signs, specifically
macroangiopathic. Signs of mild chronic micro-
vascular leukoencephalopathy are observed in the
cerebral hemispheres. Frontal, bilateral, mildly
symmetrical cerebral cortical atrophy to integrate
into the cognitive/functional context. No hydro-
cephalus, midline deviations, or cerebellar tonsil
ectopia. No visible acute skull fractures”. Blood
biochemical analysis revealed no significant devi-
ations. In this context, the patient was transferred
to an internal medicine service with the diagno-
sis of ischemic stroke without imaging findings.
During hospitalisation, he showed improvement
in neurological deficits and was transferred to a
convalescence unit.

Three weeks later, after worsening of dysar-
thria, a new CT showed: “(…) discrete left corti-
co-subcortical hypodensity, which corresponds to
a subacute ischemic infarction. The hypodensity
continues inferiorly along the corona radiata up
to the left thalamus. No other parenchymal en-
cephalic anomalies are detected. (…)”.

On July 7th, 2021, the patient was transferred
to our rehabilitation centre to continue the reha-
bilitation process, where he underwent an inter-
disciplinary rehabilitation intervention focused on
functional independence; however, this case study
will focus on speech therapy assessment.

The language assessment tool used was the Lis-
bon Aphasia Assessment Battery (BAAL) (Cas-
тро-Caldaš, 1979; Damásio, 1973; Ferro, 1986), an
aphasia assessment test adapted and standardised
for European Portuguese that encompasses speech
fluency, object naming, verbal and non-verbal
comprehension, repetition, reading, writing, and
oral apraxia (Sociedade Portuguesa de Terapia da
Fala, 2020). It also uses taxonomic classification
criteria to determine the different types of apha-
sia and the aphasia quotient (AQ) (Ferro, 1986).
The patient showed non-fluent speech (occasional-
ly correct words), along with numerous anomic
pauses and paraphasias (especially literal), a short sentence length (2–3 words), faulty grammatical structure, marked articulation distortions, and apraxic behaviour. In the naming task, the patient managed to name 12/16 objects presented, revealing literal paraphasias. He identified all the objects presented correctly. In the assessment of language comprehension with simple commands, he managed to perform 7.5/8 correctly (4/4 directed to the body and 3.5/4 directed to objects). The results of the token test were below what was expected for his level of education (14/22). In the additional test of answering yes/no questions, he was consistent in the answers given (8/8), with the need for extra time to answer, especially due to difficulties in word retrieval and motor speech programming. In the repetition task, he had a performance of 10/30, showing literal paraphasias. In the reading assessment, he was able to correctly pair 16/16 written words to objects. He was able to read a text and respond appropriately to all related questions (6/6). In the writing assessment, he managed to write only his first name. Based on the performance on the language assessment, the patient presented with Broca’s aphasia (AQ = 60.02%, Grade 2; Table 1).

Clinical swallowing assessment revealed a reduction in lip strength with consequences for the oral phase of swallowing (difficulties capturing food and the presence of residue accumulation in the perioral region). Mastication was unilateral (on the left side), showed poor efficiency, and resulted in difficulties containing the bolus, increased oral transit time, and residue accumulation in the right lateral vestibule. The swallowing reflex was delayed. There was decreased elevation and support of the laryngopharyngeal complex, with occasional episodes of gurgling voice, which was cleared only after verbal instructions. The patient showed fatigue during the meal. He was able to swallow non-thickened liquid without clinical signs of penetration/aspiration. To illustrate the severity of dysphagia, the functional outcome, and dietary recommendations, we used the Dysphagia Outcome and Severity Scale (DOSS, O’Neil, Purdy, Falk, & Gallo, 1999) and the Functional Oral Intake Scale (FOIS, Crary, Mann, & Groher, 2005; Portuguese version: Queirós, Moreira, Silva, Costa, & Lains, 2013). DOSS provides a severity classification based on several parameters, including oral stage bolus transfer, pharyngeal stage retention, and airway protection (O’Neil et al., 1999). On the other hand, the FOIS scale documents the changes in functional oral intake of food and liquids (Crary et al., 2005). The functional outcome of the patient corresponded to level 4 on the DOSS, Mild-Moderate Oropharyngeal Dysphagia, and a level 5 on the FOIS: total oral intake of multiple consistencies requiring special preparation (Table 1).

During the inpatient stay, there was a decline in clinical and functional status, with severe repercussions in speech, language, and swallowing performance.

Language reassessment showed non-fluent speech, almost absent with behaviour compatible with apraxia of speech, namely increased groping for sound positions, giving up talking most of the time, errors in stress, and sound distortions. In the naming task, he could not name any stimuli. In the language comprehension assessment with simple commands, he managed to perform 4/8 correctly (3/4 directed to the body and 1/4 directed to objects). Token test was far below the acceptable level for his level of education (7.5/22). He also failed in the word repetition task (0/30) and showed increased difficulties in reading and was able to read only simple sentences. In the writing assessment, he managed to write only short words with copy. Based on his performance on the language assessment, the patient was diagnosed with Global aphasia (AQ = 12.5%, Grade 1; Table 1).

In addition, the patient’s swallowing performance also worsened. It was observed that he had increased difficulty in containing bolus in the oral cavity, with the need for lip external pressure to hold the bolus inside the mouth and initiate the swallowing reflex. He was unable to chew and there was a need to change the consistency of his food to purees (level 4 according to the classification of the International Dysphagia Diet Standardisation Initiative – IDSSI, Cichero et al., 2017). There was an increase in signs of fatigue during meals, a marked decrease upon elevation.
and support of the laryngopharyngeal complex, and a delay in triggering the swallowing reflex (> 5 seconds). There was a need to modify liquid viscosity to mildly thick (IDSSI level 2), and he began showing difficulties in managing saliva. Gurgling voice was frequently observed when the patient was outside and during mealtimes. His status decreased to level 3 on the DOSS, which corresponds to moderate dysphagia, and to level 4 on the FOIS, which corresponds to total oral intake of a single consistency (Table 1).

Table 1. Performance on language and swallowing assessments

<table>
<thead>
<tr>
<th>Time post onset</th>
<th>First assessment</th>
<th>Reassessment</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>3M7D</td>
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<tr>
<td><strong>Language assessment</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Lisbon Aphasia Assessment Battery</td>
<td>Non-fluent speech (occasionally correct words)</td>
<td>Non-fluent speech (absent)</td>
</tr>
<tr>
<td>Fluency</td>
<td>Broca’s aphasia, AQ = 60.02%</td>
<td>Global aphasia, AQ = 12.5%</td>
</tr>
<tr>
<td>Naming</td>
<td>12/16</td>
<td>0/16</td>
</tr>
<tr>
<td>Object identification</td>
<td>16/16</td>
<td>10/16</td>
</tr>
<tr>
<td>Verbal commands</td>
<td>7.5/8</td>
<td>4/8</td>
</tr>
<tr>
<td>Token test</td>
<td>14/22</td>
<td>7.5/22</td>
</tr>
<tr>
<td>Repetition</td>
<td>10/30</td>
<td>0/30</td>
</tr>
<tr>
<td>Reading comprehension</td>
<td>6/6</td>
<td>2/6</td>
</tr>
<tr>
<td>Writing</td>
<td>Only the first name spontaneously</td>
<td>Short words by copy</td>
</tr>
<tr>
<td><strong>Swallowing assessment</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Dysphagia Outcome and Severity Scale</td>
<td>Level 4: Mild-moderate dysphagia: Intermittent supervision/cueing, one or two consistencies restricted. Retention in pharynx, cleared with cue. Retention in the oral cavity that is cleared with cue.</td>
<td>Level 3: Moderate dysphagia: Total assist, supervision, or strategies, two or more diet consistencies restricted. Moderate retention in pharynx, cleared with cue. Moderate retention in oral cavity, cleared with cue. Airway penetration to the level of the vocal cords without cough with two or more consistencies.</td>
</tr>
<tr>
<td>Functional Oral Intake Scale</td>
<td>Level 5: Total oral intake of multiple consistencies requiring special preparation</td>
<td>Level 4: Total oral intake of a single consistency</td>
</tr>
</tbody>
</table>

Thus, a more specific neuroimaging examination was requested, namely brain magnetic resonance imaging (MRI) with contrast, in which a “diffusion restriction halo was detected in the periphery of a non-swelled area of T2/FLAIR hypersignal, which extends cortico-subcortical pyramidal and peri-pyramidal supra and infratentorial on the left and in the middle cerebellar peduncles - evidence of demyelination (progressive multifocal leukoencephalopathy or immune reconstitution syndrome). Symmetrical bilateral perifrontal and perisylvic cortical volume loss - frontal atrophy. (…)” (Figure 1).

Although the initial clinical presentation pointed to ischemic stroke, after a gradual worsening of neurological and functional abilities, additional aetiological imaging and serological examination was performed. The aetiological diagnosis was progressive multifocal leukoencephalopathy, in the context of the HIV1 Infection, which after this diagnosis would be classified as AIDS stage C3 based on the CDC classification (Castro et al., 1993). The patient then began receiving antiretroviral therapy to reverse the symptoms.
The case took an unexpected turn when the patient was diagnosed with progressive multifocal leukoencephalopathy instead of a stroke. PML lesions can occur in any area of the CNS (Adang & Berger, 2015; Moreh et al., 2017), therefore, the patient’s wide range of impairments was not surprising. The white matter, the frontal, parietal, and occipital lobes, as well as the cerebellum, internal capsule, and basal ganglia, are the most affected areas (Chahin et al., 2016; Moreh et al., 2017). Predictably, the most common symptoms presented were mono- or hemiparesis, alteration of the psychic state, limb ataxia, memory loss, apraxia, aphasia, and agnosia (Bensalem & Berger, 2002; Chahin et al., 2016; Grebenciucova & Berger, 2018; Le & Spudich, 2016; Moreh et al., 2017; Siddiqi et al., 2020). Limb weakness is a frequent presentation of PML in individuals with HIV (Bensalem & Berger, 2002; Berger, Pall, Lanska, & Whiteman, 1998; Grebenciucova & Berger, 2018). In this case study, right limb weakness was observed, as well as associated language impairments. Therefore, based on clinical observation, the first diagnosis was left hemisphere stroke. The diagnosis was apparently confirmed by the second CT scan, which showed lesions on the left hemisphere that could explain a large proportion of the deficits; however, even those types of lesions can be expected in PML. In the CT scans of individuals with PML, Chahin et al. (2016) mentioned hypodense lesions without the mass effect. The MRI is thus a more precise imaging method to diagnose PML (Chahin et al., 2016) and, in this case, the MRI made it possible to finally observe the nature and extent of brain damage, and hence find a more precise correlation between the lesions and their functional impact on our patient, as well as help define the prognosis. The images were clear, showing bilateral frontal lobe lesions that perfectly explained the motor aphasia and the speech apraxia. Supra- and infratentorial lesions were also observed on the left hemisphere. According to a study published in 2015, patients with infratentorial lesions face a worse functional prognosis and a higher risk of developing critical neurologic sequelae (Le & Spudich, 2016; Yamamoto, Watanabe, Kikuchi, Oka, & Gatanaga, 2015).

Miskin, Ngo, and Koralnik (2016) found that, similar to our patient, the most common initial diagnosis in individuals with PML was vascular diagnoses, which was attributed to about 33% of the participants in their study. A delay in attributing the diagnosis, in addition to being expensive for the national health system, causes anxiety in patients and their families and leads to delays in the appropriate treatment, thus, worsening the prognosis (Miskin et al., 2016).

In their study, Moreh and colleagues (Moreh et al., 2017) found a positive effect of rehabilitation on PML, and thus recommended it concomitantly with antiretroviral therapy. However, our patient didn’t seem to benefit from rehabilitation, which could be because of the delay in antiretroviral treatment. Besides, the HIV infection had a 20-year onset, and during that period, he never underwent antiretroviral therapy. Kamtchum-Tatue and colleagues (2016) reported the presence of neurological symptoms in their study participants up to three months after infection; however, due to the lack of antiretroviral treatment, the patient displayed cognitive and memory problems. Due to the patient’s wide range of impairments, the rehabilitation options were limited. However, the patient showed improvements in his cognitive function and memory loss. The case was presented to the patient’s family and friends, who were surprised by the patient’s improvement.
remission of the symptoms occurred after initiation of antiretroviral therapy (Kamtchum-Tatue, Wan Sulaiman, & Lekoubou, 2016). With a 20-year HIV onset, the damage to the CNS would probably be considerable in our patient.

According to the information from our case study, and according to previous research, the functional impact cannot be exclusively attributed to PML (Siddiqi et al., 2020). Hence, some of the impairments shown by our patient can be either related to PML or HAND (HIV-associated neurocognitive disorders) (Antinori et al., 2007; Siddiqi et al., 2020). A study conducted by Halvorsen, Moelleken and Kearney (2003) about swallowing dysfunction in HIV patients detected the presence of aspirations in patients with and without PML. The subjects of the above-mentioned study (Halvorsen et al., 2003) showed many symptoms that were similar to our patient, namely delayed pharyngeal swallow, decreased laryngeal elevation, pooling in the valleculae and pyriform sinus, and clinical signs of penetration.

After the diagnosis, the patient began antiretroviral therapy, which is the treatment postulated by many researchers and, according to them, this treatment can lead to an improvement in clinical and functional status (Grebenciucova & Berger, 2018; Koralnik, 2006; Lima et al., 2010). Previous studies have also shown that optimising antiretroviral therapy (Bensalem & Berger, 2002; Grebenciucova & Berger, 2018; Koralnik, 2006; Siddiqi et al., 2020) and avoiding immunosuppressants are currently the best available ways to reverse the effects of PML (Siddiqi et al., 2020). In spite of that, it is estimated that the average life expectancy after diagnosis is 3 months to 1 year (Bensalem & Berger, 2002; Koralnik, 2006; Lima, Andrade, Etchebehere, & Silva-Vergara, 1998). Our patient initiated clinical decline and ended up passing away a few months after discharge.

CONCLUSION

This case illustrates the relevance of clinical disclosure for the establishment of appropriate functional prognosis, as well as effective treatment, and discharge planning. It also shows that the initial clinical features can be misleading, leading to concealment of the real aetiology, as well as a delay in appropriate treatment, especially in multipathology patients.

The concealment of clinical information by patients and caregivers, as well as a lack of access to the patient’s clinical file between units (or at least some kind of alert), can result in ambiguous diagnoses.

It is important to promote clinical and scientific meetings to bring awareness to this type of misleading situation. This will help clinicians investigate the aetiology of the condition and administer treatment more efficiently. On the other hand, the family and patient’s expectations will be more appropriate given the functional prognosis, and discharge can be planned more realistically and carefully.
REFERENCES


