CASE STUDY

ACTA Vol. 17, No. 4, 2019, 469-485
NEUROPSYCHOLOGICA

Received: 11.02.2019
Accepted: 08.12.2019

A – Study Design
B – Data Collection
C – Statistical Analysis
D – Data Interpretation
E – Manuscript Preparation
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NEUROLINGUISTIC AND ACOUSTIC STUDY OF LOGOPENIC PRIMARY PROGRESSIVE APHASIA IN ARABIC

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SUMMARY

Background: The primary progressive aphasia (PPA) or Mesulam syndrome is an isolated and progressive deterioration of language, usually due to progressive focal atrophy of the left perisylvian regions. Given that very little data on PPA is available in non-Western languages in the literature, we describe the first case of logopenic PPA in Arabic.

Material/ Methods: Neuropsychological, neuroimaging and linguistic protocol have been administered to the patient. The Neurolinguistic assessment was carried out with the Moroccan version of the Montreal-Toulouse linguistic exploration protocol, the apraxia of speech protocol, the Moroccan version of MLSE (Mini-Linguistic Status Examination); some subtests of the BDAE (Boston Diagnostic Aphasia Examination) while the computerized acoustic analysis was performed with Vocalab4 ™.

Results: The acoustic analysis showed mainly instability in pitch and amplitude. However articulatory disruptions are very mild in our case. There is a parallelism between spoken language which is marked by phonological paraphasias with a „pseudo-stuttering „and written language disorder which displays a phonological alexia, a severe acalculia and an agraphia. Our patient presents L-PPA subtype 1 on the logopenic spectrum.

Conclusions: These results are consistent with the neuropsychological hypothesis of a dysfunction in phonological buffer reflecting the features of logopenic PPA. Furthermore, our case displayed atypical neurolinguistic patterns in comparison with other cases described in the European languages due to the Arabic specific linguistic structure.

Key words: neurolinguistic, primary progressive aphasia, atypical AD, logopenic, acoustic markers, Arabic

INTRODUCTION

Definition of PPA

The concept of primary progressive aphasia or Mesulam syndrome (Mesulam, 1982, 2001) is an isolated and progressive deterioration of language, usually due to progressive focal atrophy of the left perisylvian (dominant hemisphere) regions. Patients with PPA do not strictly have the same patterns as in Broca or Wernicke aphasia vascular types. Probably this is explained by the nature of in PPA which is multifocal, partial and progressive. PPA syndrome can be fluent or non-fluent and may or may not alter phonology, syntax, or verbal semantics (meaning comprehension of words). However, there is a heterologous semiology and different clinical pictures that were first grouped under the same term of primary progressive aphasia and were then better identified and described. A classification has been established (Gorno-Tempini et al., 2011, Harris et al., 2013, Mesulam et al., 2014, Dubois et al, 2014) to allow standardization of clinical diagnoses, the precision of the clinical criteria and the identification of the underlying disorders within PPA phenotypes. Morphological brain imaging (CT scan and MRI) confirms the absence of a specific cause of aphasia (stroke or tumor) and may show atrophy, confirming the degenerative process. Focused hypometabolism, (PET scan) seen by functional imaging, precedes the atrophy identified in MRI.

According to Gorno-Tempini et al. (2011), the different clinical phenotypes of language disorders include three subtypes: 1 • PPA agrammatic/ non-fluent subtype; 2 • PPA semantic subtype; 3 • PPA logopenic subtype. This classification of clinical phenotypes in the form of progressive aphasia is based on the identification of the correlation between clinical features and localization of atrophy regions.

Although, this classification into three distinct PPA subtypes does not match with all the observed clinical phenotypes. Some patients were unclassified according to the Gorno-Tempini guidelines, while others corresponded to two subtypes simultaneously. Furthermore, revisions to the criteria for logopenic subtype of PPA have been proposed to address these challenges and findings. Indeed, mixed forms are observed and it is more interesting to consider a new classification of PPA as continuums (Mesulam et al., 2014). Thus, the continuum of PPA continuum presented four subtypes: 1-PPA agrammatic subtype; 2 • PPA semantic subtype; 3 • APP logopenic subtype (with or without repetition disorder of sentences); 4 • Mixed PPA subtype (new form which results of the association of an agrammatic non-fluent PPA and a deficit of comprehension of the words). This new approach incorporated overlaps of PPA which often make difficult to differentiate the boundaries between the clinical phenotype of agrammatic non-fluent and the logopenic PPA subtype. (Mesulam et al., 2014).

Logopenic PPA is the most variant recently described in the literature. The deficit in finding words and repeating sentences is the main feature of this variant. Spontaneous language is disturbed by a slow flow and frequent breaks due mainly to the lack of the word, without any obvious sign of agrammatism (the sentences

are short, but syntactically correct). These deficits of language production are distinguished from those observed in the agrammatic variant (hesitation but with dysprosodic production marked by phonetic paraphasias and agrammatism) and are characterized by phonemic paraphasias in spontaneous language and / or confrontational naming. The deficits observed in this logopenic variant would be mainly underpinned by a phonological short-term memory deficit which would explain the preservation of the repetition of isolated words while the repetition of sentences is impaired. Therefore, it's frequent to observe a deficit of the repetition of sentences in the agrammatic variant, but in the logopenic subtype it would be due to the syntactic complexity rather than to the length of the sentence which explains the grammatical simplifications.

MATERIALS AND METHODS
CASE STUDY

Case history

Patient ES 53 years old, right-handed, married, mother of two children, had a bachelor degree in law. Since 2015, E.S has been having difficulties with the language that hindered communication with others. A speech and language assessment confirmed an isolated language disorder with a lack of the word. On the basis of the initial symptoms reported by the patient's husband, the language disorders began in March 2015.

The patient had a progressive cognitive decline, markedly pronounced in her speech and language, of more than 18 months, with notable difficulties in finding words. Some short-term memory deficits were also notable. There was no apparent change in the overall level of functioning with respect to the use of electronic devices, driving, maintaining personal hygiene and activities of daily living. E.S is still working and she is very passionate about violin and cooking. The language disorders have worsened since 2017.According to E.S: „I experienced frequently interruptions when I talk and this bothers me a lot, I am very embarrassed to speak and cannot express my thoughts coherently". ES is not informative given the lack of the word. Her language complaint is more or less detailed, with some anecdotes to illustrate her forgetfulness and lack of the word in French and Arabic. The speech was interrupted with latency and many redundancies occurred with hypophonia in the presence of „spasmodic dysphonia", „a laryngeal syncinesia" or „pseudo-stuttering." and a rapid extinction of the voice are often observed. E.S reported that she had difficulty managing multitasking with attentional errors. According to her, the search for words becomes more and more laborious.

Following this preliminary investigation, a more detailed neurological, neurolinguistic and acoustic exploration is carried out in order to make an accurate diagnosis given the heterogeneity of deficits presented by E.S.

Neuropsychological, neuroimaging and linguistic protocol

Neuropsychological, neuroimaging and linguistic protocol has been administered to E.S who presented a logopenic PPA at the Alzheimer center in Rabat. The Neurolinguistic assessment was carried out with the Moroccan version of the Montreal-Toulouse linguistic exploration protocol (M. El Alaoui Faris et al., 1994), the apraxia of speech protocol (Taiebine and El Alaoui Faris, 2017), the Moroccan initial version of MLSE (Catricalà et al, 2013); some subtests of the BDAE (Diouny 2010) while the computerized acoustic analysis was performed with Vocalab4 ™ (Menin-Sicard, A., & Sicard, E, 2009).

Neurological examination

In March 2018 the patient was seen by the neurologist who confirmed the language disorders. The neurological examination does not reveal Parkinsonian signs, oculomotor disorders, or posture disorders. Cardiovascular examination is normal. MRI revealed a parietal and occipital cortical atrophy with normal volume of both hippocampi. There were no cerebrovascular or ischemic signs. (see figure 1, 2 and 3).

The biological analysis is normal in terms of TSH at 0.81 and B12at 244 pg / ml. CSF biomarker analysis was positive and showed a decrease in Aβ1-42 amyloid protein, while Tau and phospho-Tau protein were increased. Therefore, E.S displays an atypical AD which has been started with a progressive disorganization of language.

Neuropsychological assessment

E. S showed an overall cognitive decline with dominant language impairment (MOCA: 11/30 vs MMSE: 14/30). The neuropsychological exploration shows nosognosia and a significant deterioration in the executive functions affecting all

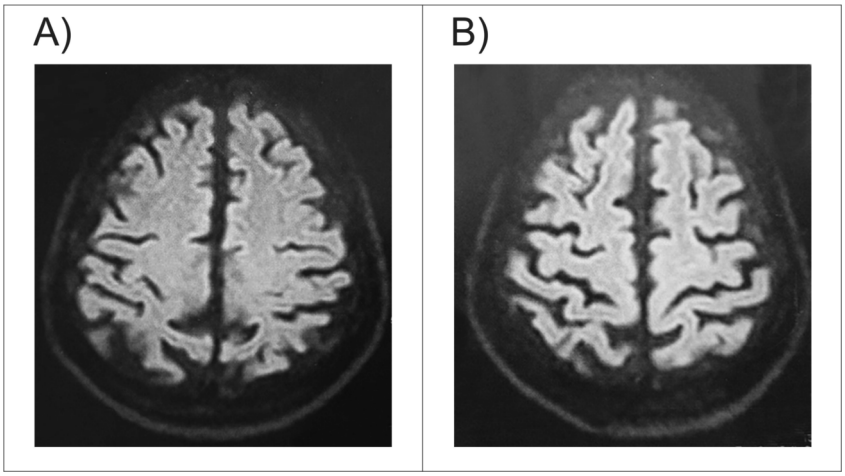

Fig. 1. Axial CT brain MRI showing posterior cortical atrophy

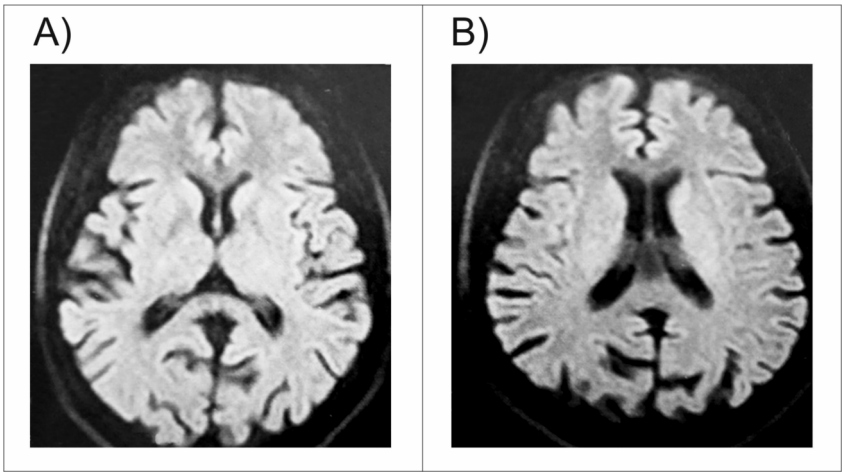

Fig. 2. Axial brain MRI showing parietal atrophy

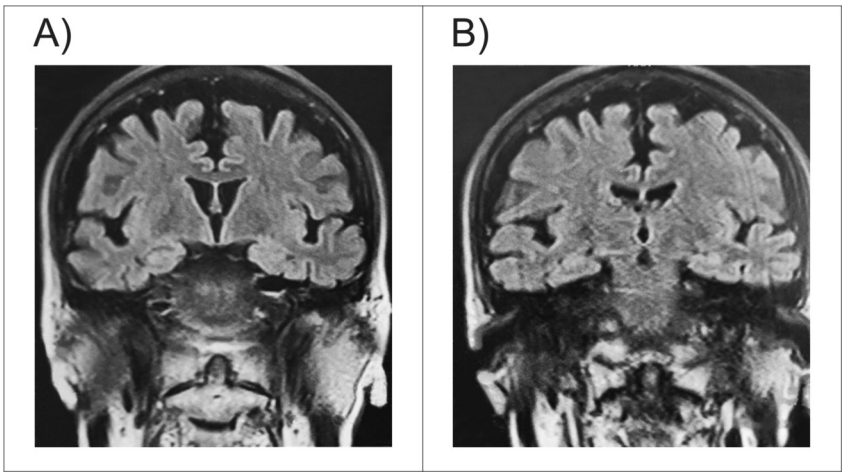

Fig. 3. Cerebral MRI coronal cut not objectifying hippocampal atrophy

the processes of the central administrator (flexibility, inhibition and multitasking). This executive disruption affects intellectual abilities, the implementation of episodic memory recovery strategies, as well as visuo-constructive functions. The latter has been severely affected. Semantic memory is also impaired. In addition, there are parietal-like disorders that dominate the clinical picture. It should be noted the presence of mild ideomotor apraxia, a mild aperceptive agnosia in the absence of elements of asemantic agnosia or hemineglect. E.S presented a severe agraphia and acalculia in the absence of signs of right and left indistinction as well as digital agnosia. (see table 1)

On the other hand, the visuo-gnosic functions are marked by an aperceptive visual agnosia in the absence of asemantic or associative agnosia. At this evolutionary stage of the disease, ideomotor praxis were still possible (symbolic ges-

tures and pantomines). We didn't observe an oral-linguo-facial apraxia, while the visuo-constructive praxias still be deteriorated. Behaviorally, an uninhibited character with a familiarity is present discreetly confirming the prefrontal dysfunction.

Tab.1. Results of the neuropsychological examination of E.S

Tests	Subtests	
MOCA- Montréal Cognitive assessment	Total score: Visuospatial and executive functions Naming Attention Language (repetition) Abstraction Memory Orientation	11/30 0/5 3/3 2/5 1/3 2/2 2/5 1/6
MMSE- Mini Mental Status Examination	Total score: Orientation Registration Attention et calculation Recall Language Praxis	14/30 3/10 3/3 1/5 0/3 6/7 0/1
LAST- Language Assessment Screening Test	Expression index Score: Receptive index score: Total score :	6/8 7/7 13/15
TMT A and B – Trail Making Test	TMT A : 3 errors TMT B : failed	(Time : 4 min 33 sec) (closing-in" symptom)
Clock Drawing test	3/8 spontaneous drawing 5/8 copy	
Isaacs Set Test (IST)	Letter Fluency: Semantic Category Fluency: animals fruits	1 (3 intrusions) (7) (5) 1 intrusion
Digit span task	Digit span forwards: Digit span backwards:	2 2
Corsi Block test	Visual memory span :	2
Free and Cued Selective Reminding Test (FCSR)	Immediate recall : Total recall 1: Total recall 2: Total recall 3: Delayed Total Recall: Recognition: Perseverations:	6/16 6/16 3/16 5/16 3/16 16/16 11 / 17 intrus
Rey Complex Figure Copy test	Time: Score:	5 min 32 seconds 5/36
Battery For Limb Apraxia Assessment	Symbolic gestures: Pantomine : Non-sense / abstract gestures :	5/5 6/10 6/8
Visual Object and Space Perception Battery (VOSP)	Silhouettes Object decision Dot counting Position discrimination Number location Cube analysis	(T=14/20) (T=11/20) (T=8/10) (T=12/20) (T =10/10) (T=5/10)
PEGV subtests – Protocole Montréal Toulouse d'évaluation des gnosies visuelles	Functional Association: Categorical Associations:	9/10 9/10

Tab.1(cont.). Results of the neuropsychological examination of E.S

The Stroop Color and Word Test (SCWT)	- Color reading: Self-corrected errors: - color naming: Self-corrected errors/ Non-Self-corrected errors: 3 -Interference task:	Temps 85 sec 7 / Non-Self-corrected errors: 4 93 sec failed
Subtests FAB and Mattis	Similarities (conceptualization): Go/ no Go: Conflicting instructions: Luria task « frises »: closing-in" symptom	1/3 1/3 1/3
Edinburgh Handedness Inventory	100% right-handed	
Instrumental Activities Of Daily Living	Total score:	8/8
MADRS - Montgomery-Asberg Depression Rating Scale	Total score:	10/60
Mc Near Self-assessment of cognitive deficits scale	Total score:	26/45

Neurolinguistic assessment

The initial assessment of linguistic disorders is carried out in august 2017 with the Moroccan version of the Montreal-Toulouse linguistic protocol (M. El Alaoui Faris et al., 1994). The results confirmed that the spontaneous speech of E.S is slightly reduced; the lack of the word is mild but there was a decrease in vocal pitch and amplitude during the interview. Pseudo-stuttering is predominant especially on the last syllable of the words that are missing. Phonemic paraphasias and neologisms are observed while the grammar is respected. Spoken comprehension is perfectly preserved for simple words and sentences, however difficulties in understanding appeared when it comes to syntactically complex sentences. Repetition is preserved for both words and sentences. The written language is characterized by a moderate dysorthographia and alexia.

The follow-up assessment is carried out later between April and June 2018. Spontaneous speech worsened with dysfluency and many phonemic distortions. The patient is uninformative because of the low-pitched voice. The induced speech is slightly unintelligible or even agrammatic. During naming task, E.S presented a severe lack of the word with many phonemic paraphasias. Repetition task is better than naming with slight phonetic distortions. The written language is marked by severe agraphia, alexia and acalculia (see figure 4). On the other

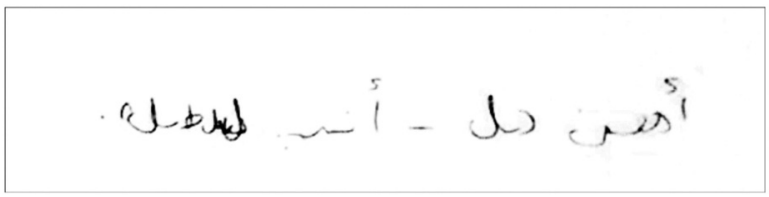

Fig. 4. Sample of spontaneous writing sentence in Arabic by E.S

hand, the assessment of apraxia of speech showed spared repetition of distinctive phonetic pairs for both consonants and vowels. However, the task of repetition is slightly impaired for: words 3 times in a row; sentences; and diadocokinesis.

On the other hand, on the MLSE subtests, the repetition is more or less preserved for the words but impaired for the non-words and the sentences (deficit in phonological buffer). The naming of objects is impaired (lack of the word with trans-linguistic or cross-linguistic errors). Object naming difficulties can be attributed, at least in part, to semantic interferences caused by loud lexical coding, so that the appropriate name cannot be distinguished from semantically related distractors. There is a dissociation between the comprehension of isolated words which is partially impaired and the comprehension of sentences which is very deficient. As for the writing, it deteriorated towards jargonagraphy and a severe apraxic agraphia. At the syntactic level, agrammatism have been objectified in spontaneous and induced speech. We note the presence of a phonological / deep alexia (dissociation between reading words preserved vs impairment of reading non-words with many lexicalizations and semantic paralexias) associated with a severe acalculia. (see table 2,3,4, 5, 6 and 7)

Tab. 2. Results of the neurolinguistic examination of E.S with MT86 (positive responses)

MT86 Moroccan version	- Listening comprehension	9/ 11
	- Reading comprehension	8/ 11
	- Reading aloud	11/ 13
	- Copywriting	2/ 4
	- dictation writing	1/ 7
	- Repetition	10/ 13
	- Naming	11 / 16
	- Oral-facial Praxis	5/ 6

Tab. 3. Results of the confrontation naming subtest with MLSE

TARGET WORD	PATIENT'S ERRORS	TYPE OF ERROR
مرساة /mirsātun/ (anchor)	Hameçon	Cross-linguistic error (Arabic to French) –Visual paraphasia
أكورديون /akordiyoun/ (accordion)	Instrument	Semantic paraphasia
خيمة /χaymatun/ (igloo)	فرن /fornun/ (oven)	Visual paraphasia
طاووس /ṭāwous/ (peacock)	جميل / ملك الطيور/ طاووس / beau/roidesoiseaux/paon	Phonemic and lexical approach
فقمة /foqmatun/ (seal)	Phoque	Cross-linguistic error

Tab. 4. Results of the sentence comprehension subtest with MLSE

TARGET WORD	PATIENT'S ERRORS	TYPE OF ERROR
اَلْكَلْبُ مَتْبُوعٌ بِالْوَلَد والبَنْت (The dog is followed by the children)	اَلْكَلَابْ مَتْبُوعِينْ بِالْوَلَد (The dogs are followed by the child)	Visual and morphological paraphasia(the other dog is identified as a shadow by the patient)
الرِّجَالْ مُنَغَّتَة فِيهُم المْرَاة (The boys are indicated by the woman)	المْرَاة مُنَغَّتِينْ فيها الرِّجَالْ (The woman is indicated by the boys)	Thematic error

Tab. 5. Results of the words and nonwords subtest with MLSE

TARGET WORD	PATIENT'S ERRORS	TYPE OF ERROR
عنامة /ʿanamatun/	علامة /ʿlamatun/ (sign)	Lexicalization (Substitution of phonemes)
فثاقة /faθāqatun/	ساقة /saqatun/	Pseudo-lexicalization

Tab. 6. Results of sentence repetition subtest with MLSE

TARGET SENTENCE	PATIENT'S ERRORS	TYPE OF ERROR
القِطَارْ غَادِي فُوقْ السِّكَّة (The train runs on the tracks)	القطار الصغير علّى السكة (the little train on the track)	Omission of the verbs and addition of word
اللاَّعِبْ اِللِّي دَارْ المْخَالْفَة اعْطَاهْ الحَكَمْ إنْذَارْ. (The footballer, who made a foul, was booked by referee).	ملاعب (stadiums)	Derivational and syntactic errors
مْنِينْ دْخَلْنَا لِلبِيتْ المَظْلَمْ، قَلْبْنَا عْلَى السَّارُوتْ، بَاشْ نْشَعْلُوا الضُّو. (Just as we entered in the dark room, we looked for the switch to turn on the light)	دخلت البيت لمظلم كن نقلبو على الساروت باش نشعل ضّو (I entered in the dark room to look for the switch to turn on the light)	Derivational and syntactic errors

Tab. 7. Results of the words and nonwords reading subtest with MLSE

TARGET WORD	PATIENT'S ERRORS	TYPE OF ERROR
شرجة /ṣaraʒatun/	شجرة /ṣaʒaratun/ (tree)	Lexicalization
حرسة /ḥarasatun/	حسرة /ḥasaratun/ (dissapointment)	Lexicalization

Acoustic analysis

Since our patient had a hypophonia associated with a pseudo-stuttering, we attempted to explore via objective and computerized voice measurements these patterns. The spectrographic analysis of temporal and acoustic characteristics was performed using the VOCALAB® 4 software. We analyzed the acoustic indicators of pauses, pitch, amplitude, endurance and noise to harmonics ratio.

Vocal sample of E.S (sustained vowel /a/) showed mainly instability in vocal pitch and amplitude. The vocal onset is quite unstable. Generally speaking, the

Arabic vowel system is straightforward: three different vowel qualities, each with a short and long variant. The difference in vowel length in Arabic is not a difference in vowel quality, but in duration. This is similar to the difference in duration of musical notes, where a half-note, for example, is held twice as long as a quarter note, and so on. The harmonic content is present at the beginning of the vocal sample and then decreased to one or two harmonics (harmonic poverty). In our patient, the word retrieval impairment is probably due to a disruption of post-lexical and phono-motor processes (Caramazza, 1997, Deacon et al, 2000). However, the articulation disorders are very discrete at the time of the evaluation and the progression of acoustic time course remains hypothetical towards a non-fluent PPA in the presence of syntactic disorders. It should be mentionned that our patient could possibly develop a mixed variant (non-fluent PPA and logopenic PPA) which has been described by Mesulam et al (2014). Hence the longitudinal monitoring of these acoustic parameters is recommended in order to predict the outcome of neurolinguistic patterns in PPA and may distinguish its different subtypes.

DISCUSSION

Our patient presents an atypical AD having started with a logopenic PPA. The diagnosis of AD was confirmed by biomarker analysis in the CSF.

A recent classification has been established (Gorno-Tempini et al., 2011, Harris et al., 2013, Mesulam et al., 2014) to allow a standardization of the clinical diagnosis of PPA. Since logopenic PPA (L-PPA) is often associated with Alzheimer's disease (AD), activities of daily living may be deteriorated more rapidly in L-PPA than in non-fluent PPA (Leyton et al., 2013; 2014). Patients with L-PPA have typical linguistic deficits, a form of cerebral atrophy and perhaps genetic susceptibility, which justifies considering this variant as a discrete endophenotype of AD. More specific clinical, neuroradiological and anatomic markers can reinforce the consistency of this syndrome. (Leyton and Hodges, 2013).

In typical AD forms, the majority of ADs that occur in young people have the usual characteristics of the disease with an initial and predominant decline in episodic memory. Since the new NINCDS-ADRDA criteria (McKhann G et al, 2011) specified that the age of onset should be between 40 and 90 years, this could delay the diagnosis. However, even in the typical forms with inaugural memory disorders, instrumental cognitive deficits are earlier and more severe, and amnesia is less present at the prodromal stage than in late forms (Koss E et al, 1996, Licht EA et al., 2007). Similarly, frontal and executive functions can be particularly impaired (Reid W et al, 1996; Macoir et al, 2017). In addition, there is a reversed correlation between anosognosia and the age of onset of the disease, with younger patients having a better awareness of their disorders than older patients (Kashiwa Y et al, 2005), which may contribute to divert the diagnosis.

Teichmann et al. (2013) predicted that the use of cerebrospinal fluid (CSF) markers with neurolinguistic markers would allow division of the entire logopenic PPA group into a subgroup of patients with probable underlying Alzheimer's dis-

ease (Logopenic PPA +) and another subgroup of patients who are unlikely to have Alzheimer's disease (Logopenic APP-). In order to individualize the clinical bio-markers predictive of a probable underlying pathology of Alzheimer's disease, the authors compared the tow subtypes of PPA by correlating them to neurolinguistic batteries.

On the other hand, neuroimaging studies of logopenic PPA revealed different abnormalities observed mainly in the left temporoparietal junction and the posterior, supramarginal and angular temporal gyri. The most common underlying pathology is Alzheimer's disease (Gorno-Tempini et al., 2011). In the perisylvian region, the atrophy is more marked in the temporoparietal junction in the logopenic subtype, however it's located in the inferior frontal gyrus in the agrammatic subtype (Gorno-Tempini et al, 2004, Hodges et al, 1992; Mummery et al, 2000, Adlam et al, 2006). The distinction between logopenic PPA and non-fluent PPA is relevant for clinical practice; logopenic PPA is associated with the Alzheimer's disease while non-fluent PPA and particularly the apraxia of speech, is strongly related to tau-positive pathology (Josephs et al, 2006; 2012). Therefore, studies using the Pittsburgh Compound B scans as a suspected biomarker of Alzheimer's disease have demonstrated high retention of Pittsburgh compound B in almost all cases with logopenic PPA while its retention is low or normal in non-fluent PPA (Rabinovici et al, 2008). Unfortunately, the Pittsburgh Compound B scan is not widely available in current practice. According to Mesulam et al (2014), the progression of logopenic PPA is the most variable of PPA subtypes: in some patients, the logopenic PPA is the prodromal stage of non-fluent or semantic PPA; in other patients, the progression of atrophy towards the medial temporal lobe leads to an episodic memory deficit and lately to the PPA + syndrome ; while some patients have an isolated logopenic PPA for several years which is most commonly associated with an Alzheimer's disease.

While the definition of PPA is precise and the distinction between its forms: non-fluent, logopenic and fluent are accepted by most authors, the clinical presentation remains nonetheless heterogeneous. PPA patients sometimes resist to the classical classification found in the subtypes used in the aphasiological literature (Gorno-Tempini, 2011). ES illustrated this pattern with the presence of a linguistic atypia manifested by the coexistence of phonological, agrammatic and acoustic disorders. On the other hand, our case is not compatible with the diagnostic criteria of progressive apraxia of speech (PAOS) or dysarthria. Unlike the conventional conduction aphasia, the primary logopenic progressive aphasia does not present difficulty of repetition of words, relatively less phonemic paraphasias than anomia and successive autocorrections. According to Mesulam et al (2014), the progression of logopenic PPA is the most variable of PPA subtypes. Therefore, the progression of atrophy towards the medial temporal lobe leads to an episodic memory deficit and lately to the PPA + syndrome which is currently the case for our patient.

Arabic generally has an idiosyncratic morphology whose main feature is its nonlinear or nonconcatenative structure. Unlike English or Italian, Arabic mor-

phemes are typically interwoven with each other in a way that makes borders difficult to distinguish between them. (Boudelaa & Marslen-Wilson, 2000). When the phonological decoding skills are not fully mastered and the morphology of the specific language is essential in word recognition (Abu-Rabia et al 2003), usually heavy reliance on the morphology may occur (Beland & Mimouni, 2001). In addition to the consonant and vowel phoneme inventory of Arabic, there are also two phonemic processes, (1) vowel lengthening (as in kataba ' he wrote' → kaataba 'he corresponded') and (2) gemination, or doubling (tashdiid), as in darasa 'he studied' → darrasa 'he taught). Each of these processes contributes to the derivation of words from a lexical root and forms a key component of the derivational system of Arabic (Ryding K.C, 2014). This fact explains the prevalence of morphological and visual errors in our patient, when the phonological decoding process is partially functioning, in Arabic orthography words that are visually and phonologically similar have a high potential of being related to the same root (root's effect). Sometimes morpheme boundaries are "fuzzy," that is, they fuse into each other in Arabic in synthetic type of language, indicating various kinds of grammatical and lexical information. A former knowledge of Arabic root types and their variants as well as a knowledge of morphological processes need to be combined in order to deal with the complexities of Arabic word formation (Ryding K.C, 2014). However, this pattern is distorted in our patient as the visual processing seems to be disrupted in terms of lexical retrieval during confrontation naming. The semantic errors were present on average across all tasks due to its heavy reliance on visual orthography, which resulted in phonemic and visual paraphasias.

From the above results, we conclude that E.S displayed a speech which is generally semi-telegraphic with simplified syntactic structures but showed a tendency to use overgeneralization in the naming task. Object naming difficulties can be attributed, at least in part, to semantic interferences caused by loud lexical coding, so that the appropriate name cannot be retrieved directly from her semantic lexicon. Lexicalization errors in reading/repetition, where E.S produced a word form instead of the non-word. According to the dual mechanism account, over-lexicalization errors are a result of applying regular inflection to non-words. This error type reflects an over-reliance on orthography pathway to compensate for the deficiency of the phonological route.

In Arabic (as in many other languages) it is often the case that morphology overlaps or interfaces with phonology – e.g., through application of phonological rules – (morphophonology) and also with syntax, through application of syntactic requirements such as government or agreement rules (morphosyntax). The explanation of morphological errors in Arabic is given by the presence of the root effect of the word. When words are linked visually, phonetically and semantically, they have systematic morphological links. The predominance of morphological errors in Arabic is due to the semantico-morphological relations of words more present in Arabic than in other Western languages (Béland and Mimouni 2001). On the other hand, Khwaileh et al, (2017) attempted to investigate the morpho-

syntactic processing of regular (sound) and irregular (broken) Arabic plurals in patients with morpho-syntactic impairment. Error analysis revealed different patterns of morpho-syntactic errors depending on the type of pluralization (sound vs broken). Omissions formed the vast majority of errors in sound plurals, while substitution was the only error mechanism that occurred in broken plurals. The dissociation was statistically significant for retrieval of morpho-syntactic information (vocalic pattern) but not for lexical meaning (consonantal root), suggesting that the participants' selective impairment was an effect of the morpho-syntax of plurals.

In this context, E.S fail to integrate perfectly into these subtypes, or can move from one to another as a continuum or within the same disorder in the form of a spectrum. In the latter, the atypical neurolinguistic pattern in ES is characterized by the presence of a type 1 logopenic PPA (Giannini et al, 2017) which evolves within a continuum while taking signs of agrammatic PPA at the level of syntactic encoding disorder. This evolution is compromised by the cooccurrence of acoustic disturbances. On the basis of these elements, we suggest that L-PPA may present itself as an initial symptom of AD in atypical cases. The presence of a significant impairment of episodic memory indicates a progression of atrophy towards the medial temporal lobe and thus the evolution towards a multi-domain APP + (Mesulam et al, 2014). This clinical manifestation may occur due to the fact that the language mechanisms rely on working memory and episodic memory, in addition to the neuroanatomical overlap that may occur between clinical presentations of typical AD and L-PPA they share the same neuro-pathological findings (Magnin et al, 2013).

Although many contemporary clinical case studies suggest that the logopenic variant of PPA is essentially equivalent to a language variant of AD. In addition, other clinical phenotypes of PPA may be associated with an AD pathology. (Grossman M et al, 2010. Etcheverry et al, 2012; Magnin et al, 2013 ; Mi Jin Oh et al, 2018).

In terms of experimental studies, Meilán JJ et al (2014), have examined spontaneous speech in free conversations recorded in patients with AD to evaluate their speech. In mild AD, phonological, articulatory and phonological paraphasia were found as well as speech disorders; the ratio of perseverative errors was significantly lower, more empty language and shorter conversions, as well as higher hesitation rates and more phonological errors. This finding didn't match with the features displayed by E.S.

On the other side, Meilán JJ et al (2012), have attempted to specifically determine the acoustic parameters of speech that are sensitive to the onset of the disease, and their association with the neurolinguistic deficit of AD. With regard to dysprosodia, Tosto et al. (2011) found a prosodic alteration in AD: presence of emphasis on certain syllables, changes in rhythm or timing, and differences in pitch and intonation.

Ultimately, our case shows the difficulty of determining a clinical diagnosis that has been split between L-PPA and nf-PPA at certain stage of the AD disease. On the basis of the initial symptoms reported by the patient's husband, language was clearly the first area affected, resulting in a diagnosis of PPA. However, the

dementia was moderate at the time of the initial neurological evaluation and its condition changes severely with the presentation not only of the symptoms of the language, but also of the nonverbal domains. The presence of a significant impairment of episodic memory indicates a progression of atrophy towards the medial temporal lobe and thus the evolution towards APP + syndrome. On the other hand, our ES case is not compatible with the diagnostic criteria of progressive speech apraxia or dysarthria. Nevertheless, our case presents some „symptomatic similarities" with speech apraxia and conduction aphasia as phonological impairment is more pronounced at the beginning or the onset of syllables and words than its coda. In terms of differential diagnosis, the fact of having a severe apraxic agraphia with jargonagraphia excludes E.S from a similarity with conduction aphasia, especially that she didn't proceed by way of phonemic approaches and self-corrections during oral or written production.

CONCLUSION

E.S presented an atypical AD that started with language disorders, which is confirmed by CSF biomarker analysis. Neurolinguistic analysis classified the disorder as L-PPA. This analysis showed that linguistic semiology is very complex in PPA. We have seen how the studies published regarding this matter still concern a small number of patients and use batteries which are sometimes different.

Finally, the contribution of acoustic evaluation in neurodegenerative diseases, objectify significant alterations of voice and speech and which differ qualitatively from those caused by normal aging or other pathologies. These vocal deficiencies can be considered as objective, subjective, subtle symptoms for early diagnosis that could coexist or even differentiate and predict the outcome of the neurolinguistic profile. This finding could allow the creation of a diagnostic test for AD through the analysis of acoustic parameters of speech that are very low in terms of time and resources (Meilán et al, 2012, 2014). Although different current studies tried to decipher the PPA at the early stages by exploring and screening multiple markers (neuropsychological, neurological, neurobiological, neuroradiological, neurolinguistic, acoustic and computational), research studies investigating intervention outcomes and trials have been very limited due to the progressive nature of the disorder. In this context, many researchers and clinicians are using different therapeutic approaches ranging from proactive management by Rogers (2000) to a successful use of Augmentative and Alternative Communication (AAC) in Polish (Góral-Półrola et al, 2016). Further research is needed to replicate them in individuals with PPA cross-linguistically in non-European languages.

DISCLOSURE STATEMENT

No sponsorship or funding arrangements relating to this article and no conflicts of interest exist.

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