

**Supplementary Table 1.** Pseudotumoral amyloid angiopathy cases reported in literature (Modified from<sup>10</sup>)

Publication/ case number	Age sex	Clinical presentation	T2/FLAIR	T1 Gd	T2 GRE	H-MRS	Histology	Drugs treatment	Outcome/ follow up
Mandybur 1992 <sup>40</sup>									
1	62F	Confusion	Isolated, Infiltrative HI	No CE	-	-	Biopsy: CAA-RI	Steroids+CP	Favorable
Osumi 1995 <sup>12</sup>									
2	59F	Headache, motor deficit	Isolated, Infiltrative HI (T)	No CE	-	-	Surgery: CAA	Steroids	Death/5 months
Ortiz 1996 <sup>41</sup>									
3	64F	Confusion, headache	Isolated, Infiltrative HI	No CE	-	-	Biopsy: CAA	Steroids	Favorable
Fountain 1996 <sup>42</sup>									
4	66M	Confusion, cognitive decline, aphasia	Multiple, Infiltrative HI	No CE	-	-	Biopsy: CAA-RI	Steroids, CP	Stable
Polivka 1999 <sup>13</sup>									
5	60M	Headache	Isolated, Infiltrative HI	No CE	-	-	Surgery: CAA-RI	None	Favorable/5 years
6	74F	Sensori-motor deficit	Isolated, Infiltrative HI	CE	-	-	Surgery: CAA-RI	None	NI
De Broucker 2000 <sup>14</sup>									
7	64F	Seizures	Isolated, Infiltrative HI	No CE			Biopsy: CAA	None	Favorable/2 years
8	69F	Aphasia, motor deficit	Isolated, Infiltrative HI	LCE	MB		Biopsy: CAA	None	Favorable/1 year
Caulo 2001 <sup>23</sup>									
9	41M	Headache, seizure	Bilateral infiltrative HI (FTPO)	No CE	-	-	Biopsy: CAA	None	NI/1 year
Oide 2002 <sup>15</sup>									
10	69M	Cognitive decline, seizure	Multiple, extensive HI	No	-	-	Biopsy: CAA-RI	None	Ongoing deterioration
Schwab 2003 <sup>43</sup>									
11	74M	Cognitive decline, seizure	Infiltrative HI	LCE	-	-	Biopsy: CAA-RI	Steroids	Favorable/1 year
12	70F	Seizure	Infiltrative HI	LCE	-	-	Biopsy: CAA-RI	Steroids+CP	Favorable/1 year
Tamargo 2003 <sup>44</sup>									
13	80F	Confusion, aphasia, hemineglect	Multiple, Infiltrative HI	Mild CE	-	N	Biopsy: CAA-RI	Steroids	Favorable/1 year
Vandermisse 2003 <sup>26</sup>									
14	46M	Cognitive decline	Isolated, Infiltrative HI	No CE	-		Biopsy: CAA	None	Stable/11 years
Safriel 2004 <sup>16</sup>									
15	49M	Seizure	Isolated, Infiltrative HI (TO)	No CE	-	N	Surgery: CAA-RI	Steroids	Favorable/9 months
16	71F	Cognitive deterioration	Isolated, Infiltrative HI (PO)	No CE	-	N	Biopsy: CAA-RI	None	Ongoing deterioration/ 11 years
Mulvey 2005 <sup>21</sup>									
17	53M	Hemiparesis Headaches Seizure	Bilateral extensive HI (FTPO)	No CE	-	-	Surgery: CAA	Steroids	NI
Scolding 2005 <sup>29</sup>									
18	70F	Confusion, hallucinations, reduced conscious level	Temporal lobe swelling Multifocal WM lesions in occipital lobe	CE			Biopsy: CAA-RI	Steroids+CP	Favorable
19	72M	Hemiparesis Confusion Dysphasia Dyspraxia	Isolated FP diffuse WM HI with mass effect	No CE			Surgery: CAA-RI	Steroids	Death
Andrade 2006 <sup>17</sup>									
20	45M	Seizure	Isolated, Infiltrative HI (F)	No CE	-	-	Surgery: CAA	None	Favorable/6 months
Mikolaenko 2006 <sup>25</sup>									
21	50M	Seizure	Well-circumscribed isolated (F)	Mild CE	-	-	Surgery: CAA-RI	None	Favorable
Wong 2006 <sup>39</sup>									
22	79F	Subtle cognitive dysfunction	Bilateral WM HI (FTP)	Mild CE	-	-	Biopsy: CAA-RI	Steroids+MTTX	Favorable (1 recidive)
McHugh 2007 <sup>27</sup>									
23	80F	Seizures, aphasia, hemianopia, hemiparesis	Bilateral diffuse HI WM (F)	No CE	MB		Biopsy: CAA-RI	Steroids	Favorable (2 recidives)/ 24 months
Kloppenborg 2010 <sup>45</sup>									
24	74M	Gait disorder, sleepiness seizure, confusion	Bilateral WM HI (F)	Mild CE			Biopsy: CAA-RI	Steroids	Favorable
Morishige 2010 <sup>22</sup>									
25	78F	Aphasia Cognitive decline	Unilateral WM HI ( F)	Mild CE	- NI	-	Biopsy: CAA-RI	Steroids	Favorable/ 24 months
Bekkelund 2011 <sup>20</sup>									
26	57M	Crural monoparesis	Unilateral hemispheric HI lesion (FTPO)	LCE	MB		Biopsy: CAA-RI	Steroids+CP	Favorable (1 recidive)/ 15 months
Karbowniczek 2012 <sup>18</sup>									
27	64F	Cognitive decline	Two well-delimited HI	No CE	-	-	Surgery: CAA	None	Cognitive deterioration
28	38M	Headache, hemiparesis, aphasia	Well-delimited HI	No CE	-	-	Surgery: CAA	None	Favorable
Kotsenas 2013 <sup>19</sup>									
29	63F	Confusion, cognitive decline, motor and visual deficit, seizure	Isolated, Infiltrative HI (TPO)	Mild LCE	MB		Biopsy: Vascular inflammation	Steroids in 3 patients	Favorable
30	62F	Sensitive deficit	Isolated, Infiltrative HI (P)	Avid LCE	MB SS	-	Biopsy: CAA		
31	77M	Confusion, cognitive decline, aphasia	Isolated, Infiltrative HI (FP)	No CE	MB SS	-	Biopsy: Vascular inflammation	None in 2 patients	Favorable
32	74M	Confusion, cognitive decline	Isolated, Infiltrative HI (PO)	Mild LCE	MB	-	Biopsy: CAA-RI		
33	71F	Confusion, cognitive decline, headaches	Isolated, Infiltrative HI lesion (TPO)	Avid LCE	MB	-	Biopsy: CAA-RI		Favorable
Danve 2014 <sup>9</sup>									
34	63F	Seizure	Multiple and bilateral HI lesions (FT)	Mild CE	MB within lesion	-	Biopsy: CAA-RI	Steroids+CP/ Micopheno late	Favorable/11 months
Franco-Macias 2016 <sup>11</sup>									
35	67F	Aphasia Hemiparesis Seizure	Unilateral extense HI lesion (FTP)	LCE	No MB. SS+Old he mato ma		Biopsy: CAA-RI	Steroids	Stable (persistent aphasia, cognitive decline)
Ronsin 2016 <sup>10</sup>									
36	70M	Confusion	Multiple , bilateral, asymmetric HI ST WM lesions (FTO)	LCE	MB, SS	-	Biopsy: CAA	Steroids	Death (ICH)/3 months
37	80M	Subacute cognitive decline	Multiple , bilateral, asymmetric HI ST WM lesions (FTO)	No CE	MB, SS	N	-	Steroids	Favorable /12 months
38	63M	Subacute cognitive decline and motor deficit	Bilateral, asymmetric HI ST WM lesions (PO)	No CE	MB	N	-	Steroids	Favorable /24 months
39	82M	Subacute cognitive decline and headache	Isolated HI ST WM lesion (PO)	LCE	MB, SS	N	-	Steroids	Favorable /18 months
40	63M	Subacute cognitive decline and headache	Extense confluent bilateral asymmetric HI ST WM lesions (FPO)	No CE	MB, SS	N	-	None	Favorable /12 months
Tolchin 2016 <sup>24</sup>									
41	52M	Confusion, paranoia. Non convulsive status epilepticus	Multiple, bilateral, asymmetric HI ST WM lesions (FTO)	No CE	MB	-	Biopsy: CAA	Steroids	Favorable/UK
Our patient									
42	54M	Acute aphasia, seizure	Isolated, Infiltrative HI (T)	No CE	MB inside the lesion	N	Surgery: CAA-RI	Steroids	Favorable/24 months

CAA-RI: Cerebral β amyloid angiopathy related angiitis, CAA: cerebral amyloid angiopathy, CE: contrast enhancement, CP: cyclophosphamide, DWI: diffusion weighted imaging, F: Frontal, HI: hyperintense, ICH (intra-cerebral hemorrhage), IT: infratentorial, LCE: leptomeningeal contrast enhancement, M: male, MB: microbleeds, H-MRS: MRI spectroscopy, MTTX: methotrexate, N: normal, ND: non diagnostic, O: Occipital, P: Parietal, SS: superficial siderosis, ST: supratentorial, T: Temporal, WM: white matter