

Table 1. Summary of patient characteristics

Reference	Age,		Sex	DI	Eye	Diplopia	HA	Cerebellar	Spine	Muscle	Mental		Tumor	Other neurologic	Other	Sclerotic lesions	Staining		Birbeck granules
	Year	years									SZ	status					CD68	CD1a	
Adem et al. [6]	2005	26	M	-	-	-	-	-	-	-	+	-	-	-	-	+	-	-	-
Koziolek et al. [3]	2005	55	F	+	-	-	-	-	-	-	-	-	-	weakness	bp, e	Lytic	-	-	-
Rushing et al. [7,8]	2004	26	M	-	-	-	-	-	-	-	+	+	-	-	-	NR	+	-	-
Sheu et al. [9]	2004	48	M	+	-	-	-	-	-	-	-	-	-	rp, e	rp, e	NR	+	-	-
Boulos et al. [10]	2004	64	M	-	+	-	-	-	-	-	+	+	-	CN 6 palsy	L	NR	+	-	-
Andrade et al. [11]	2004	40	F	-	-	-	-	-	-	-	-	-	-	rp, B, m	rp, B, m	+	+	-	-
Black et al. [12]	2004	51	M	+	-	-	-	-	-	-	+	-	-	personality change	rp	+	-	-	-
De Abreu et al. [13]	2004	52	M	-	+	-	-	-	-	-	-	-	-	x, rp, bp	+	+	-	-	
Kovacs et al. [14]	2004	35	F	+	-	-	-	+	+	-	-	+	-	rp, e, L	NR	+	-	-	
Taillia et al. [15]	2004	26	M	+	-	-	-	-	-	-	+	+	-	-	+	+	-	-	
Simiele Narvaarte et al. [16]	2004	54	M	+	-	-	-	+	+	-	-	-	-	decrease libido	bp	+	+	-	
Johnson et al. [17]	2004/																		
and Pereira Neto et al. [18]	2004	34	M	-	+	-	-	+	+	+	-	+	-	Later rp, h	NR	+	-	+	
Allen et al. [19]	2004	60	M	-	+	-	-	-	-	-	-	-	-	L	+	+	-	-	
Lyders et al. [20]	2003/																		
and Tashjian et al. [21]	2004	27	F	+	-	-	-	+	+	-	+	-	-	MS	-	+	-	-	
Bisceglia et al.* [22]	2003	44	M	+	+	-	+	-	-	-	-	+	-	-	bp, SL	+	+	-	
		35	M	+	-	-	-	-	-	-	-	-	-	-	bp, SL	+	-	-	
Hoffmann et al. [23]	2004	61	M	-	+	-	-	-	-	-	-	-	-	rp, h	NR	+	-	-	
Thorns et al. [24]	2003	55	F	-	+	-	-	-	-	-	-	+	-	Vertigo	rp, L	+	+	-	
Ivan et al. [25]	2003	32	F	-	-	-	-	-	-	-	-	-	-	Liver	Lytic	+	-	-	
Weidauer et al. [26]	2003	44	M	-	-	-	-	+	-	-	-	-	-	bp, f, wt	NR	+	-	-	
Lenahan et al. [27]	2003	25	M	-	+	-	-	-	-	-	-	-	-	wt, rp	NR	+	-	-	
Curgunlu et al. [28]	2003/																		
and Albayram et al. [29]	2002	43	M	+	+	-	-	+	-	-	-	-	-	L, rp, H	NR	+	-	-	
Le Goff et al. [4]	2002	61	M	+	+	-	-	-	-	-	-	-	-	rp	NR	+	-	-	
Mascalchi et al. [30]	2002	52	M	+	-	-	-	+	-	-	-	-	-	-	+	+	-	-	
Perras et al. [31]	2002	47	M	-	-	-	+	-	-	-	-	-	-	hypomania, e, f, venous occlusion bp, L	+	+	-		

Reference	Age,			Sex	DI	Eye	Diplopia	HA	Cerebellar	Spine	Muscle	Mental status	Mental		Tumor	Other neurologic	Other	Sclerotic lesions	Staining		Birbeck granules		
	Year	years	Age										SZ	status					CD68	CD1a		S-100	
Oweity et al. [32]	2002	55	M	+	-	-	-	-	-	-	-	-	-	+	decrease libido	bp, f, rp, wt, e	+	+	-	-	-		
Reithmeier et al. [33]	2002	51	M	+	-	-	-	-	-	-	-	-	+	decrease libido	L	NR	NR	+	+	-	-	-	
Mahnel et al. [34]	2002	53	M	+	-	-	-	-	+	-	-	+	+	forgetfulness, libido	e	NR	NR	+	+	-	-	-	
Esmaeli et al. [35]	2001	55	M	+	+	-	-	-	-	-	-	-	-	+									
Wu et al. [36]	2001	43	F	-	+	-	-	-	-	-	-	-	-	+	h								
Wärermann et al. [5]	2001	46	F	-	+	-	-	-	-	-	-	-	-	+	SL, rp								+
Grothe et al. [2]	2001	50	Not mentioned	-	+	-	-	-	+	-	-	-	-	-	e, SL								+
Vasakova et al. [37]	2001	63	M	+	-	-	-	-	-	-	-	-	-	-	f, bp, L	NR	NR	NR	+	+	-	-	-
Rush et al.† [38]	2000	40	F	+	+	-	-	-	-	-	-	-	-	-	bp, L								+
Sheidow et al. [39]	2000	57	M	+	+	-	-	-	-	-	-	-	-	-	L, wt, x, h								+
Sistermann et al. [40]	2000	51	M	+	+	-	-	-	-	-	-	-	-	-	x, rp, h								+
Stojkovic et al. [41]	2000	18	F	-	+	-	-	-	-	-	-	-	-	-	rp, bp								+
Marsot-Dupuch et al. [1]	2000	19	M	-	+	-	-	-	-	-	-	-	-	+	L, bp								+
Yamamoto and Mizuno [42]	2000	66	M	-	-	-	-	-	-	-	-	-	-	-	wt								+
Pego-Reigosa et al. [43]	2000	50	F	-	+	-	-	-	-	-	-	-	-	-	x								+
Wright et al.‡ [44]	1999	42	F	+	+	-	-	-	-	-	-	-	-	-	rp, h								-
van der Lee et al. [45]	1999	55	F	+	+	-	-	-	-	-	-	-	-	-	bp								-
Amrith et al. [46]	1999	35	M	-	+	-	-	-	-	-	-	-	-	-	rp								+
Boralevi et al. [47]	1998	46	F	+	-	-	-	-	+	-	-	+	-	-	f, wt, L, rp, x								+
Pautas et al. [48]	1998	31	M	-	-	-	-	-	-	-	-	-	-	-	x, rp								-
Evidente et al. [49]	1998	69	M	+	-	-	-	-	+	-	-	-	-	-	Py								-
Bancroft and Berquist§ [50]	1998	75	M	+	+	-	-	-	-	-	-	-	-	-	L								-
		53	M	-	+	-	-	-	+	-	-	-	-	-	x								+
															wt, L								+
															f, wt								+
															rp								+

Reference	Year	Age, years	Sex	DI	Eye	Diplopia	HA	Cerebellar	Spine	Muscle	Mental		Tumor	Other neurologic	Other	Sclerotic lesions	Staining		Birbeck granules
											SZ	status					CD68	CD1a	
Tritos et al. [51]	1998	58	F	+	-	-	-	+	-	-	-	-	-	rp	+	+	-	-	-
de Palma et al. [52]	1998	61	M	-	+	-	-	-	-	-	-	-	-	rp, L, h	+	+	-	-	-
Kim and Romero [53]	1997	41	F	+	-	-	+	-	-	-	-	-	-	bp	+	+	-	-	-
Bohlega et al. [54]	1997	33	F	+	-	+	-	+	-	-	-	-	-	bp	+	+	-	-	-
Babu et al. [55]	1997	22	M	-	+	-	-	-	+	-	-	-	-	-	+	+	-	-	-
		65	F	-	+	-	-	-	-	-	-	-	-	Paraparesis	h	+	-	-	-
Adle-Bisasette et al. [56]	1997	48	F	+/-	-	-	-	+	-	-	-	-	-	SL, rp, L	+	+	-	-	+
		34	F	+	+	-	-	-	-	-	-	-	-	h, wt, rp	+	+	-	-	-
		55	F	-	+	-	-	-	-	+	+	-	-	rp, L, h	Lytic	+	-	-	-
Gomez et al. [57]	1996	50	F	+	-	-	-	-	-	-	-	-	-	L, bp, wt	+	+	-	-	-
		73	F	+	-	-	-	-	-	-	-	-	-	x	NR	+	-	-	-
Veyssier-Belot et al. # [58]	1996	32	M	+	-	-	-	-	-	-	-	-	-	x, L, bp	+	+	-	-	-
		62	F	+	-	-	-	-	-	-	-	-	-	bp	+	+	-	-	-
		39	F	+	-	-	-	-	-	-	-	-	-	Weakness	+	+	-	-	-
		54	M	-	-	-	-	-	-	-	-	-	-	Hemiplegia	x, L, bp	+	-	-	-
		40	M	-	-	-	-	-	-	-	-	-	-	Cerebral signs	+	+	-	-	-
		57	F	-	-	-	-	-	-	-	-	-	-	h	+	+	-	-	-
		70	M	-	+	-	-	-	-	-	-	-	-	x, rp, bp	+	+	-	-	-
		34	F	+	+	-	-	-	-	-	-	-	-	rp, L	+	+	-	-	-
		51	M	-	+	-	-	-	-	-	+	-	-	bp	+	+	-	-	-
		57	M	-	-	-	-	-	-	-	-	-	-	L	+	+	-	-	-
		70	M	-	+	-	-	-	-	-	-	-	-	rp	+	+	-	-	-
		48	F	-	+	-	-	-	-	-	-	-	-	f, wt, rp, bp	+	+	-	-	-
		31	M	+	-	-	-	-	-	-	-	-	-	+	+	+	-	-	-
		44	M	-	-	-	-	+	-	-	-	-	-	-	+	+	-	-	-
		54	M	-	-	+	-	+	-	-	-	-	-	-	+	+	-	-	-
		52	M	-	+	-	-	-	-	-	-	-	+	h	+	+	-	-	-
		30	M	+	-	-	-	-	-	-	-	-	-	bp	+	+	-	-	-
		76	M	+	+	-	-	-	-	-	-	-	-	bp	+	+	-	-	-
		7	M	+	-	-	-	-	-	+	-	-	-	GH def	+	+	-	-	-

Reference	Age,		Year	Sex	DI	Eye	Diplopia	HA	Cerebellar	Spine	Muscle	Mental		Tumor	Other neurologic	Other	Sclerotic lesions	Staining		Birbeck granules	
	years	+										status	SZ					CD68	CD1a		S-100
	53	M	+	-	-	-	-	-	+	-	-	-	-	-	-	L	+				
	17	F	-	+	-	-	-	-	-	-	-	-	-	-	-	bp	+				
	51	F	-	-	-	-	-	-	-	-	-	-	-	-	x, wt	+					
	47	M	+	-	-	-	-	-	-	-	-	-	-	-	bp	+					
	43	F	+	-	-	-	-	-	+	-	-	-	-	-		NR					
	(incomplete)																				
	42	F	+	-	-	-	-	-	-	-	-	-	-	-	-		+				
	(lack of x-ray or pathology)																				
	68	F	-	-	-	-	-	-	-	-	-	-	-	-	stroke	x, f	NR				
	36	F	-	+	-	-	-	-	-	-	-	-	-	-	H, rp	NR					
	35	F	+	-	-	-	-	-	-	-	-	-	-	-	bp	+					
	76	M	-	+	-	-	-	-	-	-	-	-	-	-	x	NR					
	34	M	-	+	-	-	-	-	-	-	-	-	-	-	x	+					
	74	F	+	+	-	-	-	-	+	-	-	+	-	-	x, bp	+					
	56	F	+	+	-	-	-	-	-	-	-	-	-	hp	x, f, bp, rp	+					
	50	F	+	+	-	-	-	-	-	-	-	+	-	Py	x, L, bp, rp	+					
	46	F	+	-	-	-	-	-	-	-	-	-	-	-	bp	+					
	57	M	-	+	-	-	-	-	-	-	-	-	-	-	rp, h, f	+					
	44	F	-	+	-	-	-	-	+	-	-	-	-	-	f, wt, rp, bp	+					
	70	F	+	-	-	-	-	-	-	-	-	-	-	-	f, wt, bp	+					
Martinez [59]	1995	66	F	-	+	-	-	-	+	-	-	-	-	+	paraparesis	x	+				
Tan et al. [60]	1995	59	F	-	-	-	-	-	-	-	+	-	-	-	B, h, L, rp	+					
Fukazawa et al. [61]	1995	59	F	+	-	-	-	-	+	-	-	-	-	-	f, rp, x	+					
Offret et al. [62]	1995	43	M	+	+	-	-	+	+	-	-	-	-	?	wt, rp, bp	+					
Farre et al. [63]	1995	57	F	+	-	-	-	-	-	-	-	-	-	+	SL	+					
	50	F	-	+	-	-	-	-	-	-	-	-	-	-	rp, L	Lytic					
Chollet et al. [64]	1994	47	M	-	+	-	-	+	-	-	-	-	-	-	rp, wt, h	-					
Smith et al. [65]	1993	58	M	-	-	+	-	-	+	-	-	-	-	-	MS	NR					

Reference	Age,		Sex	DI	Eye	Diplopia	HA	Cerebellar	Spine	Muscle	Mental status	SZ	Tumor	Other neurologic	Other	Sclerotic lesions	Staining		Birbeck granules	
	Year	years															CD68	CD1a		S-100
present study																				
case 1	56	F	-	-	+	-	+	-	-	-	-	-	-	wt, bp		+	+	-	-	-
case 2	75	M	-	-	-	-	+	+	-	-	+	+	-	bp		+	+	-	-	-

-, absent; +, present; B, breast; bp, bone pain; e, endocrine; f, fever; GH def, growth hormone deficiency; h, heart; HA, headache; hp, hemiparesis; L, lung; m, mouth; MS, multiple sclerosis; NR, not recorded; Py, pyramidal signs; rp, retroperitoneal; SL, skin lesions; SZ, seizures; wt, weight loss; x, xanthomas.

Heading "Tumor" excludes eye pseudotumor.

*Two of 4 patients had neurologic involvement.

†Two of 6 patients had neurologic involvement.

‡Two of 10 patients had neurologic involvement.

§Two of 5 patients had neurologic involvement.

//One of 2 patients had neurologic involvement.

*One patient was same as in Boralevi et al. [47] and therefore excluded.

#Thirty-six of 52 patients had neurologic involvement.