

Supplementary Materials

Age at Onset and Presenting Symptom of Neurofibromatosis Type 2 as Prognostic Factors for Clinical Course of Vestibular Schwannomas

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Table S1. Parameters for the 106 NF2 patients.

Patient ID	1	2	3	5	7	8	9	11
Sex	M	F	F	M	F	F	F	F
Family history	Negative	Negative	Negative	Negative	Positive (father)	Negative	Negative	Negative
NF2 mutation type	NA	FS	NS	FS	Large genome alteration	NS	NS	FS
Age at signs of symptoms	7	1	6	1	3	2	5	4
Age at time of diagnosis	8	10	6	1	3	3	7	13
Age at start of treatment (L/R)	13/9	13/17	11/14	11/13	10/11	14/14	11/12	14/19
Age at time of surgery (L/R)	13/9	13/17	11/14	11/13	10/11	14/14	11/12	14/16
Age at deafness (L/R)	-/-	-/-	-/-	19/-	-/-	16/-	-/-	14/25
BVZ in the following course	Yes	Yes	Yes	Yes	No	No	No	Yes
Radiation in the following course (L/R)	No/No	No/No	No/No	No/No	No/No	No/No	No/No	No/No
Presenting symptoms	Visual impairment, cutaneous plexiform schwannoma	Cutaneous plexiform schwannoma, seizure, thoracic and cerebellar dysplasia	Strabismus, cutaneous plexiform schwannoma	Visual impairment, strabismus	Cutaneous plexiform schwannoma, wasting of the right hand muscles, cataract	Cataract, seizure	Strabismus	Cutaneous plexiform schwannoma
Presenting feature/pathology								
Ophthalmological findings								
Cataract	-	-	-	-	+	+	-	-
Retinal/Maculopathy	+	-	-	+	-	-	-	-
Strabismus	-	-	Idiopathic	Idiopathic	-	-	Idiopathic	-
Others	-	-	-	-	-	-	-	-
Cutaneous features								

CAL spots	-	+	+	-	-	-	-	-
Cutaneous plexiform schwannoma	+	+	+	-	+	-	-	+
Cranial lesions								
VS bilateral	-	-	-	-	-	-	-	-
VS unilateral	-	-	-	-	-	-	-	-
Cranial nerves	-	-	-	-	-	-	-	-
Meningioma	-	-	-	-	-	Temporal	-	-
Astrocytoma	-	-	-	-	-		-	-
Cortex dysplasia	-	Cerebellar	-	-	-		-	-
Vascular	-	-	-	-	-	-	-	-
Others	-	Bifrontal angiomas	-	-	-	-	-	-
Spinal lesions								
Ependymoma	-	-	-	-	-	-	-	-
Schwannoma	-	-	-	-	-	-	-	-
Meningioma	-	-	-	-	-	-	-	-
Peripheral abnormalities								
Peripheral nerve schwannoma	-	-	-	-	-	-	-	-
Neuropathy	-	-	-	-	+	-	-	-
Others	-	Thoracic dysplasia	-	-	-	-	-	-
Incidental finding/asymptomatic	-	-	-	-	-	-	-	-
Symptom/Pathology leading to diagnosis (if different to presenting symptom)	-	-	-	-	-	-	Hearing loss (VS)	Thoracic hypesthesia, back pain (spinal extramedullary tumors)

L = left, R = right, Age = median age in years, NA = not available, CAL = café-au-lait spots, BVZ = bevacizumab; MOS = mosaic, NO = no mutation, NS = nonsense mutation, FS = frameshift mutation, MIS = missense mutation, SP = splicing mutation, DEL = deletion, VS = vestibular schwannoma, SAH = subarachnoid hemorrhage.

Patient No								
	14	15	16	17	18	19	20	21
Sex	M	F	F	F	F	F	M	M
Family history	Negative	Negative	Negative	Negative	Negative	Negative	Negative	Negative
NF2 mutation type	NS	SP	NS	NA	DEL	DEL	NA	SP
Age at signs of symptoms	1	6	11	11	5	16	11	0
Age at time of diagnosis	6	6	11	12	12	16	11	10
Age at start of treatment (L/R)	-/16	-/12	14/-	14/14	-/15	17/20	-/18	13/12

Age at time of surgery (L/R)	-/16	-/12	14/-	16/-	-/15	17/25	-/18	13/12
Age at deafness (L/R)	-/16	-/-	14/13	-/-	-/16	18/25	-/-	-/-
BVZ in the following course	No	No	No	Yes	No	Yes	No	No
Radiation in the following course (L/R)	No/No	No/No	No/No	No/No	No/No	No/No	No/No	No/No
Presenting symptoms	Cataract, epiretinal gliosis, paresis of the left arm	Strabismus, wasting of the right thigh muscles, cutaneous plexiform schwannoma	Facial palsy, hypacusis	Hypacusis	Peripheral nerve schwannoma, radiating pain	Back pain, gait disturbances, scoliosis	Cutaneous plexiform schwannoma, retinal hamartoma, cataract	Visual impairment
Presenting feature/pathology								
Ophthalmological findings								
Cataract	+	-	-	-	-	-	+	+
Retinal/Maculopathy	+	-	-	-	-	-	+	+
Strabismus	-	Tumor-associated	-	-	-	-	-	-
Others	-	-	-	-	-	-	-	-
Cutaneous features								
CAL spots	-	-	-	-	-	-	-	-
Cutaneous plexiform schwannoma	-	+	-	-	-	-	+	-
Cranial lesions								
VS bilateral	-	-	+	+	-	-	-	-
VS unilateral	-	-	-	-	-	-	-	-
Cranial nerves	-	III schwannoma	VII schwannoma	-	-	-	-	-
Meningioma	-	craniocervical	-	-	-	-	-	-
Cortex dysplasia	-	-	-	-	-	-	-	-
Vascular	-	-	-	-	-	-	-	-
Others	-	-	-	-	-	-	-	-
Spinal lesions								
Ependymoma	-	-	-	-	-	+	-	-
Schwannoma	-	-	-	-	-	-	-	-
Meningioma	-	Craniocervical	-	-	-	-	-	-
Peripheral abnormalities								
Peripheral nerve schwannoma	-	-	-	-	+	-	-	-
Neuropathy	+	+	-	-	-	-	-	-
Others	-	-	-	-	-	Scoliosis	-	-
Incidental finding/asymptomatic	-	-	-	-	-	-	-	-

Symptom/Pathology leading to diagnosis (if different to presenting symptom)	Cutaneous plexiform schwannoma, cortex dysplasia	-	-	-	Hypacusis (VS)	-	-	-
Patient No								
	22	23	24	25	29	30	35	36
Sex	F	F	M	M	F	F	F	M
Family history	Negative	Negative	Negative	Negative	Positive (mother)	Positive (father)	Negative	Negative
NF2 mutation type	NA	FS	SP	SP	MOS	NA	NA	FS
Age at signs of symptoms	14	5	1	1	17	14	13	17
Age at time of diagnosis	15	14	13	15	17	15	13	17
Age at start of treatment (L/R)	26/15	15/16	14/14	16/17	21/21	24/24	17/-	23/-
Age at time of surgery (L/R)	26/15	15/16	14/14	16/17	-/-	-/-	17/-	23/-
Age at deafness (L/R)	-/15	-/-	-/-	-/-	-/-	-/-	17/-	25/-
BVZ in the following course	No	Yes	No	No	Yes	Yes	No	No
Radiation in the following course (L/R)	No/No	No/No	No/No	No/No	No/No	No/No	No/No	No/No
Presenting symptoms	Strabismus, cutaneous plexiform schwannoma	Cutaneous plexiform schwannoma	Visual impairment, strabismus	Scoliosis	Scoliosis, cutaneous plexiform schwannoma, strabismus	Paresis of the right leg, cutaneous plexiform schwannoma	Cutaneous plexiform schwannoma, anisocoria	Hemihypesthesia, cognitive disorder, intracranial pressure symptoms
Presenting feature/pathology								
Ophthalmological findings								
Cataract	-	-	+	-	-	-	+	-
Retinal/Maculopathy	-	-	-	-	-	-	+	-
Strabismus	Idiopathic	-	Idiopathic	-	Idiopathic	-	-	-
Others	-	-	-	-	-	-	Idiopathic anisocoria	-
Cutaneous features								
CAL spots	-	-	-	-	-	-	-	-
Cutaneous plexiform schwannoma	+	+	-	-	+	+	+	-
Cranial lesions								

VS bilateral	-	-	-	-	-	-	-	-
VS unilateral	-	-	-	-	-	-	-	-
Cranial nerves	-	-	-	-	-	-	-	-
Meningioma	-	-	-	-	-	-	-	-
Cortex dysplasia	-	-	-	-	-	-	-	-
Vascular	-	-	-	-	-	-	-	SAH, pericallosal artery aneurysm
Others	-	-	-	-	-	-	-	-
Spinal lesions								
Ependymoma	-	-	-	-	-	-	-	-
Schwannoma	-	-	-	-	-	-	-	-
Meningioma	-	-	-	-	-	-	-	-
Peripheral abnormalities								
Peripheral nerve schwannoma	-	-	-	-	-	+	-	-
Neuropathy	-	-	-	-	-	-	-	-
Others	-	-	-	Scoliosis	Scoliosis	-	-	-
Incidental finding/asymptomatic								
Symptom/Pathology leading to diagnosis (if different to presenting symptom)	-	Dizziness, tinnitus (VS)	Dizziness, gait and taste disturbances, ataxia, dysarthria, facial palsy, hypacusis ischemic stroke left ponto-medullary transition and VS)	Dizziness, hypacusis (VS)	Positive family history	Positive family history	-	-

Patient No								
	37	38	40	42	45	48	49	51
Sex	M	M	F	F	M	F	M	M
Family history	Negative	Negative	Positive (father)	Negative	Negative	Negative	Negative	Negative
NF 2 mutation type	FS	NA	NO	NS	SP	SP	DEL	MOS
Age at signs of symptoms	18	5	11	Asymptomatic	7	1	1	17
Age at time of diagnosis	18	6	11	10	7	1	1	17
Age at start of treatment (L/R)	19/23	16 and 25/25	-/-	-/-	21/20	16/-	-/-	30/-
Age at time of surgery (L/R)	19/23	16 and 25/25	-/-	-/-	21/20	16/-	-/-	30/-
Age at deafness (L/R)	19/23	16/-	-/-	-/-	-/-	-/-	-/-	-/-

BVZ in the following course	Yes	Yes	No	No	No	No	No	No
Radiation in the following course (L/R)	No/No	No/No	No/No	No/No	No/No	No/No	No/No	No/No
Presenting symptoms	Facial palsy	Facial palsy, hypacusis, cavus foot, cutaneous plexiform schwannoma, amblyopia, epiretinal gliosis	Asymptomatic, positive family history	Asymptomatic, incidental finding (post-trauma)	Hemiparesis, dysarthria	Strabismus, papilledema	Visual impairment	Strabismus, visual loss, facial palsy
Presenting feature/pathology								
Ophthalmological findings								
Cataract	-	-	-	-	-	-	+	-
Retinal/Maculopathy	-	+	-	-	-	-	-	-
Strabismus	-	-	-	-	-	-	-	Idiopathic
Others	-	Amblyopia	-	-	-	Idiopathic papilledema	-	-
Cutaneous features								
CAL spots	-	-	-	-	-	-	-	-
Cutaneous plexiform schwannoma	-	+	-	-	-	-	-	-
Cranial lesions								
VS bilateral	+	+	-	-	-	-	-	-
VS unilateral	-	-	-	-	-	-	-	-
Cranial nerves	-	-	-	-	-	-	-	-
Meningioma	-	-	-	-	-	Frontal	-	-
Cortex dysplasia	-	-	-	-	-	-	-	-
Vascular	-	-	-	-	Ischemic stroke pons right	-	-	-
Others	-	-	-	-	-	-	-	-
Spinal lesions								
Ependymoma	-	-	-	-	-	-	-	-
Schwannoma	-	-	-	-	-	-	-	-
Meningioma	-	-	-	-	-	-	-	-
Peripheral abnormalities								
Peripheral nerve schwannoma	-	-	-	-	-	-	-	-
Neuropathy	-	-	-	-	-	-	-	-
Others	-	Cavus foot	-	-	--	-	-	-

Incidental finding/asymptomatic	-	-	-	Post trauma	-	-	-	-
Symptom/Pathology leading to diagnosis (if different to presenting symptom)	-	-	Positive family history	Post trauma (spinal tumors)	-	-	-	-
Patient No								
	53	60	62	63	67	68	94	106
Sex	M	M	F	F	F	M	M	F
Family history	Negative	Negative	Negative	Negative	Negative	Negative	Positive (father)	Negative
NF 2 mutation type	NA	NA	SP	NA	NA	NA	SP	NA
Age at signs of symptoms	8	Asymptomatic	14	10	2	4	8	15
Age at time of diagnosis	12	16	15	15	9	7	8	18
Age at start of treatment (L/R)	-/13	20,	15/16	16/15	28/28	-/-	25/13	20/22
Age at time of surgery (L/R)	-/13	20 and 23/16 and 18	15/-	16/15	28/28	-/-	25/13	20/22
Age at deafness (L/R)	-/-	23/16	16/-	-/31	-/17	-/-	-/-	20/-
BVZ in the following course	No	No	No	No	No	No	No	No
Radiation in the following course (L/R)	No/No	No/No	No/No	No/No	No/No	No/No	No/No	No/No
Presenting symptoms	Facial palsy	Incidental finding	Cutaneous plexiform schwannoma, strabismus, peripheral nerve schwannoma	Cutaneous plexiform schwannoma, positive family history	Cutaneous plexiform schwannoma, visual loss, amblyopia left	Cutaneous plexiform schwannoma, proptosis, visual loss	Cutaneous plexiform schwannoma, cataract	Hypacusis, tinnitus, dizziness, gait disturbances
Presenting feature/pathology								
Ophthalmological findings								
Cataract	-	-	-	-	-	-	+	-
Retinal/Maculopathy	-	-	-	-	-	-	-	-
Strabismus	-	-	Tumor-associated	-	-	-	-	-
Others	-	-	-	-	Amblyopia	-	-	-
Cutaneous features								
CAL spots	-	-	-	-	-	-	-	-

Cutaneous plexiform schwannoma	-	-	+	+	+	+	+	-
Cranial lesions								
VS lesions	+	-	-	-	-	-	-	+
VS unilateral	-	-	-	-	-	-	-	-
Cranial nerves	-	-	III schwannoma	-	-	-	-	-
Meningioma	-	-	-	-	-	Intraorbital	-	-
Cortex dysplasia	-	-	-	-	-	-	-	-
Vascular	-	-	-	-	-	-	-	-
Other	-	-	-	-	-	-	-	-
Spinal lesions								
Ependymoma	-	-	-	-	-	-	-	-
Schwannoma	-	-	-	-	-	-	-	-
Meningioma	-	-	-	-	-	-	-	-
Peripheral abnormalities								
Peripheral nerve schwannoma	-	-	+	-	-	-	-	-
Neuropathy	-	-	-	-	-	-	-	-
Others	-	-	-	-	-	-	-	-
Incidental finding/asymptomatic	-	+	-	-	-	-	-	-
Symptom/Pathology leading to diagnosis (if different to presenting symptom)	-	-	Hypacusis, gait disturbances, tinnitus (VS)	Positive family history	-	-	-	-

Patient No								
	109	111	117	120	122	129	130	136
Sex	M	M	M	F	M	F	F	M
Family history	Negative	Negative	Negative	Positive (mother, aunt, sister, cousin)	Positive (father)	Negative	Negative	Negative
NF2 mutation type	NA	NA	DEL	FS	FS	FS	No	NA
Age at signs of symptoms	16	17	15	14	10	10	7	16
Age at time of diagnosis	16	17	15	14	10	13	7	16
Age at start of treatment (L/R)	-/17	19/19	26/15	27/22 and 28	-/-	15/14	14/-	25/16
Age at time of surgery (L/R)	-/17	19/19	27/-	27/22 and 28	-/-	15/14	14/-	25/16
Age at deafness (L/R)	Unknown/unknown	-/23	-/16	-/-	-/-	-/-	-/-	-/-
BVZ in the following course	No	No	Yes	No	No	No	No	No

Radiation in the following course (L/R)	No/No	No/No	No/No	No/No	No/No	No/No	No/No	No/No
Presenting symptoms	Hypacusis	Hypacusis, hoarseness	Cutaneous plexiform schwannoma, strabismus, CAL	Hypacusis	Hypacusis	Cutaneous plexiform schwannoma, hypacusis	Paresis left arm	Hypacusis
Presenting feature/pathology								
Ophthalmological findings								
Cataract	-	-	-	-	-	-	-	-
Retinal/Maculopathy	-	-	-	-	-	-	-	-
Strabismus	-	-	Idiopathic	-	-	-	-	-
Others	-	-	-	-	-	-	-	-
Cutaneous features								
CAL spots	-	-	+	-	-	-	-	-
Cutaneous plexiform schwannoma	-	-	+	-	-	-	-	-
Cranial lesions								
VS bilateral	+	+	-	+	+	+	-	+
VS unilateral	-	-	-	-	-	-	-	-
Cranial nerves	-	-	-	-	-	-	-	-
Meningioma	-	-	-	-	-	-	-	-
Cortex dysplasia	-	-	-	-	-	-	-	-
Vascular	-	-	-	-	-	-	-	-
Others	-	-	-	-	-	-	-	-
Spinal lesions								
Ependymoma	-	-	-	-	-	-	-	-
Schwannoma	-	-	-	-	-	-	-	-
Meningioma	-	-	-	-	-	-	-	-
Peripheral abnormalities								
Peripheral nerve schwannoma	-	-	-	-	-	-	+	-
Neuropathy	-	-	-	-	-	-	-	-
Others	-	-	-	-	-	-	-	-
Incidental finding/asymptomatic								
Symptom/Pathology leading to diagnosis (if different to presenting symptom)	-	-	Facial palsy (VS)	-	-	-	-	-
Patient No								
	137	141	144	152	156	159	162	166
Sex	F	F	M	M	M	F	F	M
Family history	Negative	Negative	Positive (mother)	Positive (brother)	Negative	Negative	Negative	Positive (mother)
NF2 mutation type	NA	NA	NA	FS	DEL	FS	NS	FS

Age at signs of symptoms	4	3	4	11	13	17	14	0
Age at time of diagnosis	11	13	6	11	18	17	14	11
Age at start of treatment (L/R)	23/11	16/16	-/-	-/-	29/19	-/-	18/18	14/15
Age at time of surgery (L/R)	23/11	16/16	-/-	-/-	29/19	-/-	23/23	14/15
Age at deafness (L/R)	-/22	-/-	Unknown/ unknown	-/-	Unknown/ unknown	-/-	-/-	-/-
BVZ in the following course	No	No	No	No	Yes	No	Yes	Yes
Radiation in the following course (L/R)	No/No	No/No	No/No	No/No	Yes/No	No/No	No/No	No/No
Presenting symptoms	Strabismus	Cutaneous plexiform schwannoma, seizure	Cutaneous plexiform schwannoma, radiating pain	Facial palsy, strabismus, positive family history	Back pain	Proptosis	Cutaneous plexiform schwannoma, proptosis	Cutaneous plexiform schwannoma, strabismus, visual loss
Presenting feature/pathology								
Ophthalmological findings								
Cataract	-	-	-	-	-	-	-	-
Retinal/Maculopathy	-	-	-	-	-	-	-	+
Strabismus	Idiopathic	-	-	Tumor-associated	-	-	-	Idiopathic
Others	-	-	-	-	-	-	-	-
Cutaneous features								
CAL spots	-	-	-	-	-	-	-	-
Cutaneous plexiform schwannoma	-	+	+	-	-	-	+	+
Cranial lesions								
VS bilateral	-	+	-	-	-	-	-	-
VS unilateral	-	-	-	-	-	-	-	-
Cranial nerves	-	-	-	VII and III schwannomas	-	Periorbital schwannoma	Periorbital schwannoma	-
Meningioma	-	Temporal	-	-	-	-	-	-
Cortex dysplasia	-	-	-	-	-	-	-	-
Vascular	-	-	-	-	-	-	-	-
Others	-	-	-	-	-	-	-	-
Spinal lesions								
Ependymoma	-	-	-	-	-	-	-	-
Schwannoma	-	-	-	-	+	-	-	-
Meningioma	-	-	-	-	-	-	-	-
Peripheral abnormalities								
Peripheral nerve schwannoma	-	-	+	-	-	-	-	-
Neuropathy	-	-	-	-	-	-	-	-
Others	-	-	-	-	-	-	-	-

Incidental finding/asymptomatic	-	-	-	-	-	-	-	-
Symptom/Pathology leading to diagnosis (if different to presenting symptom)	Hypacusis, dizziness (VS)	-	-	-	-	-	-	-
Patient No								
	167	168	172	178	179			
Sex	M	M	M	F	M			
Family history	Negative	Positive (father)	Negative	Negative	Negative			
NF2 mutation type	NA	FS	FS	No	NS			
Age at signs of symptoms	16	16	11	9	0			
Age at time of diagnosis	16	16	11	9	5			
Age at start of treatment (L/R)	-/-	-/29	15/15	-/-	-/-			
Age at time of surgery (L/R)	-/-	-/29	19/20	-/-	-/-			
Age at deafness (L/R)	-/-	-/-	-/-	-/-	-/-			
BVZ in the following course	No	No	Yes	No	No			
Radiation in the following course (L/R)	No/No	No/No	No/No	No/No	No/No			
Presenting symptoms	Hypesthesia right leg	Cataract	Visual loss	Foot lift plegia right	Strabismus, visual loss			
Presenting feature/pathology								
Ophthalmological findings								
Cataract	-	+	-	-	-			
Retinal/Maculopathy	-	-	-	-	-			
Strabismus	-	-	-	-	-	Idiopathic		
Others	-	-	-	-	-			
Cutaneous features								
CAL spots	-	-	-	-	-			
Cultaneous plexiform schwannoma	-	-	-	-	-			
Cranial lesions								
VS bilateral	-	-		-	-			
VS unilateral	-	-	-	-	-			
Cranial nerves	-	-	-	-	-			
Meningioma	-	-	Optic nerve sheath	-	-	Optic nerve sheath		
Cortex dysplasia	-	-	-	-	-			
Vascular	-	-	-	-	-			
Others	-	-	-	-	-			
Spinal lesions								

Ependymoma	-	-	-	-	-
Schwannoma	-	-	-	-	-
Meningioma	Thoracic	-	-	-	-
Peripheral abnormalities					
Peripheral nerve schwannoma	-	-	-	-	-
Neuropathy	-	-	-	+	-
Others	-	-	-	-	-
Incidental finding/asymptomatic					
Symptom/Pathology leading to diagnosis (if different to presenting symptom)	-	-	-	-	-

Patient No								
	10	12	13	26	27	28	31	33
Sex	F	F	M	F	F	F	F	F
Family history	Negative	Negative	Negative	Negative	Negative	Positive (grandfather)	Positive (father)	Negative
NF2 mutation type	No	SP	MOS	SP	SP	MOS	NA	MOS
Age at signs of symptoms	21	22	14	20	31	22	36	10
Age at time of diagnosis	22	22	26	21	36	22	36	29
Age at start of treatment (L/R)	22/23	24/23	30/30	-/26	-/36	-/23	58/58	31/31
Age at time of surgery (L/R)	22/23	24/23	30/30	-/26	-/36	-/23	-/-	34/-
Age at deafness (L/R)	-/25	-/-	-/-	-/-	-/39	27/26	-/-	-/-
BVZ in the following course	Yes	Yes	Yes	no	Yes	No	Yes	Yes
Radiation in the following course (L/R)	No/No	No/No	No/No	No/No	No/No	No/No	No/No	No/No
Presenting symptoms	Cutaneous plexiform schwannoma, CAL	Hemiparesis, dizziness, facial palsy, dysarthria, fine motor skill disorders	Wasting of the left hand muscles, cutaneous plexiform schwannoma	Seizure	Dizziness, sudden hearing loss	Headache, dizziness	Asymptomatic	Cutaneous plexiform schwannoma
Presenting feature/pathology								
Ophthalmological findings								
Cataract	-	-	-	-	-	-	-	-
Retinal/Maculopathy	-	-	-	-	-	-	-	-
Strabismus	-	-	-	-	-	-	-	-
Others	-	-	-	-	-	-	-	-
Cutaneous features								
CAL spots	+	-	-	-	-	-	-	-
Cultaneous plexiform schwannoma	+	-	+	-	-	-	-	+
Cranial lesions								

VS bilateral	-	-	-	-	+	-	-	-
VS unilateral	-	-	-	-	-	-	-	-
Cranial nerves	-	-	-	-	-	-	-	-
Meningioma	-	-	-	Frontal	-	Confluens sinuum	-	-
Cortex dysplasia	-	-	-	-	-	-	-	-
Vascular	-	Ischemic stroke left cerebellar peduncle	-	-	-	-	-	-
Others	-	-	-	-	-	-	-	-
Spinal lesions								
Ependymoma	-	-	-	-	-	-	-	-
Schwannoma	-	-	-	-	-	-	-	-
Meningioma	-	-	-	-	-	-	-	-
Peripheral abnormalities								
Peripheral nerve schwannoma	-	-	-	-	-	-	-	-
Neuropathy	-	-	-	-	-	-	-	-
Others	-	-	-	-	-	-	-	-
Incidental finding/asymptomatic								
Symptom/Pathology leading to diagnosis (if different to presenting symptom)	Hypacusis, tinnitus (VS)	-	Tinnitus, balance disorders (VS)	-	-	-	Positive family history	Cauda equina syndrome (ependymoma), tinnitus (VS)

Patient No								
	34	39	41	43	44	56	64	66
Sex	M	F	M	F	F	M	F	F
Family history	Negative	Positive (father)	Positive (daughter)	Negative	Negative	Negative	Negative	Negative
NF2 mutation type	NA	NS	MOS	MIS	MOS	MOS	NA	NA
Age at signs of symptoms	31	20	27	32	20	24	NA	23
Age at time of diagnosis	31	20	29	32	20	28	34	44
Age at start of treatment (L/R)	-/40	23/23	34/30	33/43	40/-	32/-	-/-	44/32
Age at time of surgery (L/R)	-/40	23/23	34/30	33/-	-/-	32/-	-/-	44/32
Age at deafness (L/R)	-/-	24/-	-/-	-/-	-/-	-/-	41/-	-/-
BVZ in the following course	No	Yes	No	Yes	No	No	No	No
Radiation in the following course (L/R)	No/No	No/No	No/No	No/No	Yes/No	No/No	No/No	No/No

Presenting symptoms	Hypacusis, sudden hearing loss, dizziness	Peripheral nerve schwannoma, hypacusis	Sudden hearing loss, hypacusis, tinnitus	Sudden hearing loss, hypacusis, tinnitus	Multiple subcutaneous peripheral nerve schwannomas lower extremity	Multiple subcutaneous peripheral nerve schwannomas	Incidental finding (unspecific neck pain)	Incidental finding
Presenting feature/pathology								
Ophthalmological findings								
Cataract	-	-	-	-	-	-	-	-
Retinal/Maculopathy	-	-	-	-	-	-	-	-
Strabismus	-	-	-	-	-	-	-	-
Others	-	-	-	-	-	-	-	-
Cutaneous features								
CAL spots	-	-	-	-	-	-	-	-
Cultaneous plexiform schwannoma	-	-	-	-	-	-	-	-
Cranial lesions								
VS bilateral	+	+	+	+	-	-	-	-
VS unilateral	-	-	-	-	-	-	-	-
Cranial nerves	-	-	-	-	-	-	-	-
Meningioma	-	-	-	-	-	-	-	-
Cortex dysplasia	-	-	-	-	-	-	-	-
Vascular	-	-	-	-	-	-	-	-
Others	-	-	-	-	-	-	-	-
Spinal lesions								
Ependymoma	-	-	-	-	-	-	-	-
Schwannoma	-	-	-	-	-	-	-	-
Meningioma	-	-	-	-	-	-	-	-
Peripheral abnormalities								
Peripheral nerve schwannoma	-	+	-	-	+	+	-	-
Neuropathy	-	-	-	-	-	-	-	-
Others	-	-	-	-	-	-	-	-
Incidental finding/asymptomatic								
	-	-	-	-	-	-	+	+
Symptom/Pathology leading to diagnosis (if different to presenting symptom)								
	-	-	-	-	-	-	-	-
Patient No								
	70	79	86	87	88	95	97	107
Sex	F	M	F	F	M	M	F	M

Family history	Negative	Positive (mother, sister)	Negative	Negative	Negative	Negative	Positive (daughter)	Negative
NF2 mutation type	NA	MOS	NA	NA	NA	NA	NA	MOS
Age at signs of symptoms	1	18	22	21	28	14	46	29
Age at time of diagnosis	39	21	24	22	32	20	25	29
Age at start of treatment (L/R)	-/-	Unknown/40	29/-	23/-	32/33	39/21	47/NA	-/40
Age at time of surgery (L/R)	-/-	-/40	29/-	23/-	32/33	39/21	47/NA	-/40
Age at deafness (L/R)	-/-	Unknown/34	-/-	23/unknown	-/-	-/30	-/-	Unknown/-
BVZ in the following course	No	No	No	No	Yes	No	No	No
Radiation in the following course (L/R)	No/No	Yes/No	No/No	No/No	No/No	No/No	No/No	No/No
Presenting symptoms	Cutaneous plexiform schwannoma, multiple subcutaneous peripheral nerve schwannoma	Headache, wasting and weakness of the left arm	Cutaneous plexiform schwannoma	Headache, incidental finding	Headache, hypacusis	Incidental finding (family history for seizures)	Breathlessness	Tinnitus, hypacusis, CAL
Presenting feature/pathology								
Ophthalmological findings								
Cataract	-	-	-	-	-	-	-	-
Retinal/Maculopathy	-	-	-	-	-	-	-	-
Strabismus	-	-	-	-	-	-	-	-
Others	-	-	-	-	-	-	-	-
Cutaneous features								
CAL spots	-	-	-	-	-	-	-	+
Cutaneous plexiform schwannoma	+	-	+	-	-	-	-	-
Cranial lesions								
VS bilateral	-	-	-	-	+	-	-	+
VS unilateral	-	-	-	-	-	-	-	-
Cranial nerves	-	-	-	-	-	-	-	-
Meningioma	-	-	-	-	-	-	-	-
Cortex dysplasia	-	-	-	-	-	-	-	-
Vascular	-	-	-	-	-	-	-	-
Others	-	-	-	-	-	-	-	-
Spinal lesions								
Ependymoma	-	+	-	-	-	-	-	-
Schwannoma	-	-	-	-	-	-	-	-
Meningioma	-	-	-	-	-	-	-	-
Peripheral abnormalities								
Peripheral nerve schwannoma	+	-	-	-	-	-	+	-
Neuropathy	-	-	-	-	-	-	-	-
Others	-	-	-	-	-	-	-	-

Incidental finding/asymptomatic	-	-	-	+	-	+	-	-
Symptom/Pathology leading to diagnosis (if different to presenting symptom)	-	CAL, multiple subcutaneous peripheral nerve schwannoma	Hypacusis, tinnitus, gait disturbances (VS)	-	-	-	-	-
Patient No								
	108	110	113	114	115	116	121	132
Sex	M	M	M	M	F	M	F	M
Family history	Negative	Negative	Negative	Positive (father)	Negative	Negative	Negative	Positive (mother)
NF2 mutation type	MOS	NA	NA	MIS	NA	NA	NA	MOS
Age at signs of symptoms	22	23	19	37	19	19	15	19
Age at time of diagnosis	29	23	22	37	19	23	36	19
Age at start of treatment (L/R)	31/-	24/-	22/28	46/46	27/32	28 and 39/32	36/-	24/23
Age at time of surgery (L/R)	31/-	24/-	22/28	46/46	27/32	28 and 39/32	36/-	24/23
Age at deafness (L/R)	unknown, unknown	24/unknown	22/-	-/-	27/32	Unknown/unknown	56/-	Unknown/24
BVZ in the following course	No	No	No	No	No	No	No	No
Radiation in the following course (L/R)	No/No	No/No	No/No	No/No	No/Yes	No/No	No/No	No/No
Presenting symptoms	Seizure, papilledema, cutaneous plexiform schwannoma	Hypacusis	Hypacusis, cutaneous plexiform schwannoma	Hypacusis, tinnitus, gait disturbances, facial palsy	Hypacusis, intracranial pressure symptoms	Hypacusis	Sudden hearing loss, Facial palsy	Hypacusis, tinnitus
Presenting feature/pathology								
Ophthalmological findings								
Cataract	-	-	-	-	-	-	-	-
Retinal/Maculopathy	-	-	-	-	-	-	-	-
Strabismus	-	-	-	-	-	-	-	-
Others	Idiopathic papilledema	-	-	-	-	-	-	-
Cutaneous features								
CAL spots	-	-	-	-	-	-	-	-
Cultaneous plexiform schwannoma	+	-	+	-	-	-	-	-
Cranial lesions								
VS bilateral	-	+	+	+	+	+	+	+
VS unilateral	-	-	-	-	-	-	-	-

Cranial nerves	-	-	-	VII schwannoma	-	-	-	-
Meningioma	-	-	-	-	-	-	-	-
Cortex dysplasia	-	-	-	-	-	-	-	-
Vascular	-	-	-	-	-	-	-	-
Others	-	-	-	-	-	-	-	-
Spinal lesions								
Ependymoma	-	-	-	-	-	-	-	-
Schwannoma	-	-	-	-	-	-	-	-
Meningioma	-	-	-	-	-	-	-	-
Peripheral abnormalities								
Peripheral nerve schwannoma	-	-	-	-	-	-	-	-
Neuropathy	-	-	-	-	-	-	-	-
Others	-	-	-	-	-	-	-	-
Incidental finding/asymptomatic	-	-	-	-	-	-	-	-
Symptom/Pathology leading to diagnosis (if different to presenting symptom)	-	-	-	-	-	-	-	-

Patient No								
	138	140	147	148	153	154	157	161
Sex	M	M	F	M	F	M	F	M
Family history	Negative	Negative	Negative	Positive (mother)	Negative	Negative	Negative	Negative
NF2 mutation type	NA	NA	MOS	NA	NS	NA	MOS	No
Age at signs of symptoms	39	18	41	27	26	31	32	22
Age at time of diagnosis	39	27	41	27	26	31	32	22
Age at start of treatment (L/R)	47/45	31/27	-/42	46/46	26/27	-/19	-/32	-/-
Age at time of surgery (L/R)	47/45	31/27	-/42	46/46	26/27	-/40	-/32	-/-
Age at deafness (L/R)	48/48	32/30	42/35	-/-	22/27	-/-	-/-	-/-
BVZ in the following course	No	Yes	No	No	Yes	No	No	No
Radiation in the following course (L/R)	No/No	No/No	No/No	No/No	No/No	No/Yes	No/No	No/No
Presenting symptoms	Tinnitus	Visual impairment	Tinnitus	Hypacusis, balance disorders, headache, double vision	Hypacusis, visual loss,	Sudden hearing loss	Hypacusis	Strabismus
Presenting feature/pathology								
Ophthalmological findings								
Cataract	-	+	-	-	-	-	-	-
Retinal/Maculopathy	-	-	-	-	-	-	-	-
Strabismus	-	-	-	-	-	-	-	Tumor-associated
Others	-	-	-	-	-	-	-	-
Cutaneous features								

CAL spots	-	-	-	-	-	-	-	-
Cultaneous plexiform schwannoma	-	-	-	-	-	-	-	-
Cranial lesions								
VS bilateral	+	-	+	+	+	+	+	-
VS unilateral	-	-	-	-	-	-	-	-
Cranial nerves	-	-	-	-	-	-	-	-
Meningioma	-	-	-	-	Optic nerve sheath	-	-	Temporal horn left lateral ventricle
Cortex dysplasia	-	-	-	-		-	-	
Vascular	-	-	-	-		-	-	
Others	-	-	-	-		-	-	
Spinal lesions								
Ependymoma	-	-	-	-	-	-	-	-
Schwannoma	-	-	-	-	-	-	-	-
Meningioma	-	-	-	-	-	-	-	-
Peripheral abnormalities								
Peripheral nerve schwannoma	-	-	-	-	-	-	-	-
Neuropathy	-	-	-	-	-	-	-	-
Others	-	-	-	-	-	-	-	-
Incidental finding/asymptomatic	-	-	-	-	-	-	-	-
Symptom/Pathology leading to diagnosis (if different to presenting symptom)	-	Hypacusis, dizziness (VS)-	-	-	-	-	-	-

Patient No					
	164	169	170	174	175
Sex	M	F	M	F	M
Family history	Positive (daughter)	Negative	Negative	Negative	Negative
NF2 mutation type	NA	No	NA	NA	No
Age at signs of symptoms	35	24	21	35	39
Age at time of diagnosis	35	26	21	35	39
Age at start of treatment (L/R)	35/-	-/27	24/-	56/-	-/-
Age at time of surgery (L/R)	35/-	-/27	24/-	56/-	-/-
Age at deafness (L/R)	-/-	-/-	-/-	57/59	-/-
BVZ in the following course	no	no	no	no	no
Radiation in the following course (L/R)	no	no	no	no	no
Presenting symptoms	Hypacusis	Hypacusis, facial palsy	Tinnitus	Tinnitus	Radiating pain (C7)
Presenting feature/pathology					
Ophthalmological findings					
Cataract	-	-	-	-	-

Retinal/Maculopathy	-	-	-	-	-
Strabismus	-	-	-	-	-
Others	-	-	-	-	-
Cutaneous features					
CAL spots	-	-	-	-	-
Cutaneous plexiform schwannoma	-	-	-	-	-
Cranial lesions					
VS bilateral	+	+	+	+	-
VS unilateral	-	-	-	-	-
Cranial nerves	-	VII schwannoma	-	-	-
Meningioma	-	-	-	-	-
Cortex dysplasia	-	-	-	-	-
Vascular	-	-	-	-	-
Others	-	-	-	-	-
Spinal lesions					
Ependymoma	-	-	-	-	-
Schwannoma	-	-	-	-	-
Meningioma	-	-	-	-	-
Peripheral abnormalities					
Peripheral nerve schwannoma	-	-	-	-	+
Neuropathy	-	-	-	-	-
Others	-	-	-	-	-
Incidental finding/asymptomatic	-	-	-	-	-
Symptom/Pathology leading to diagnosis (if different to presenting symptom)	-	-	-	-	-



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