Rasmussen's Encephalitis - Management and Treatment choices in Pediatric Patients

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INTRODUCTION

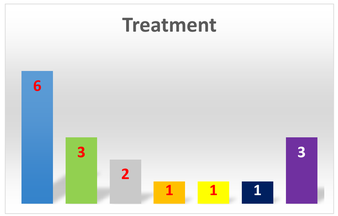
There is a high variability in both clinical picture and medical care in patients with Rasmussen's encephalitis (RE). The aim of this study is to review the clinical phenotypes, treatment used and outcome in patients diagnosed with RE in a single center.

METHODS

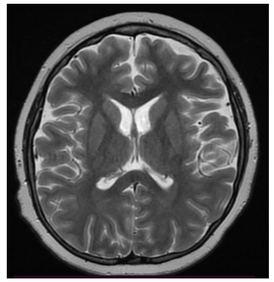
The clinical files of the patients diagnosed with Rasmussen encephalitis between 2007 - 2023 have been analyzed retrospectively (Bien diagnostic criteria 2005). Age of onset, clinical signs at onset and in evolution, seizures characteristics, CSF, MRI, including volumetries in evolution, medications used and outcome were evaluated; correlations have been made. One special cases is presented.

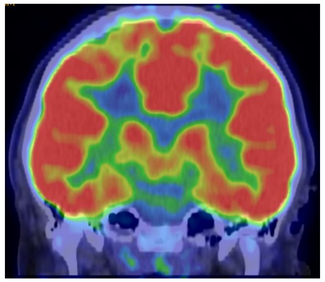
RESULTS

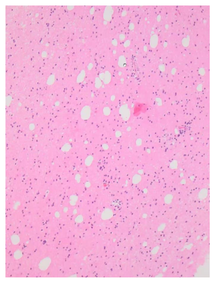
Ten patients with RE have been identified. The mean age of onset was 7.7 (2.8 – 16) years. All patients had focal motor seizures as first symptoms and 8 (80%) developed epilepsia partialis continua. 2 (20%) patients had negative oligoclonal bands and 1 (10%) showed repeatedly negative MRI; cerebral biopsy was performed to clarify the diagnosis. Volumetries have been performed in 6 patients (Table 3). Treatment included corticosteroids and intravenous immunoglobulins (IVIGs) in 6 (60%) patients, immunoglobulin alone in 3 (30%), rituximab associated to IVIGs in 2 (20%), 1 azathioprine, 1 cyclophosphamide, 1 mycophenolate mofetil. 3 (30%) brain surgery. All patients continued to have refractory seizures except 2 with hemispherotomy and 1 with a mild course ab initio.



CASE BT had onset age 9 years, EPC, negative oligoclonal bands, negative CSF antibodies for immune encephalitis, no MRI progression of atrophy after immune-suppressiove therapy, but suggestive PET-CT and cerebral biopsy. He has persistent seizures and motor deficit and no cognitive impairment.







Tabel 1. Clinical data of the study group

Patient	Gender	AH	Age at disease onset years	months	Age [decimal]	First MRI study months after onset	No of MRI studies	EPC	Hemi-paresis	Hemi-anopsia	Cognitive decline	Dysphasia
1	Male	Right	2	8	2.67	7	2	yes	yes	yes	no	
2	Male	Right	9	1	9.08	4	3	yes	yes	no	no	no
3	Male	Left	7	8	7.67	7	3	yes	no	no	no	no
4	Male	Left	10	0	10.00	2	6	yes	yes	no	yes	yes
5	Female	Right	11	0	11.00	9	4	yes	yes	yes	yes	yes
6	Female	Left	5	4	5.33	0	3	yes	yes	yes	yes	yes
7	Female	Right	4	3	4.25	0	3	yes	yes	no	no	no
8	Male	Right	3	10	3.83	10	3	yes	yes	no	yes	yes
9	Female	Right	6	0	6.00	46	1	no?	no	no	no	no
10	Male	Right	16	0	16.00	1	4	no?	yes	no	yes	no
Mean			7	7	7.58	8.6	32					

EPC=Epilepsia partialis continua, AH=Affected Hemisphere

Table 3. Absolute volume changes (cm3/year)

	AH					UH				
Patient	Cerebrum	LV	Putamen	Caudate	Amygdala	Cerebrum	LV	Putamen	Caudate	Amygdala
1	-262.86	15.28	-8.98	-5.94	-1.44	54.92	3.06	-0.52	0.04	-0.20
2	3.61	0.36	-0.29	-0.18	-0.09	5.13	0.60	-0.23	-0.16	-0.13
3	-2.11	-0.16	-0.14	-0.20	0.00	-2.97	-0.15	-0.16	-0.09	0.01
4	-3.6	-0.05	-0.18	0.06	0.03	-2.67	0.6	0.00	0.11	-0.03
5	20.54	-10.81	-3.08	-2.74	-0.76	10.10	25.09	-0.35	-0.70	0.09
6	-59.112	3.648	-0.48	-0.84	-0.072	-3.768	1.392	0.024	-0.264	0.048

LV - Lateral Ventricle; AH - Affected Hemisphere; UH - Unaffected Hemisphere

Tabel 2. Treatment

Patient	Imunnotherapy	Months from onset to immunotherapy	Present ASM	Long term cortico therapy	Mo from onset to cortico therapy	Surgery	Seizure persistence present time	Seizure frequency in present
1	no	-	LEV, LCM	yes	12	yes	yes	20/day
2	IVIG, AZA	24	LEV, LTG, CZP, CBZ	yes	20	no	yes	<10/day
3	IVIG	52	LEV, LTG, CBZ	yes	50	no	yes	1/ month
4	IVIG	10	LEV, VPA	no	11	no	yes	1-5/day
5	IVIG		no	yes		yes	no	0
6	IVIG, Rituximab	13	CBZ,LEV, PB,CLB	yes	12	yes	no	0
7	IVIG, Rituximab	3	PB, CLB, Briva, VPA, TPM, PHT	no	4	no	yes	6/month
8	IVIG, Cyclophosphamide, Mycophenolate	11	VPA, LAM, PB, LEV, DZP	yes	0	no	yes	2/day
9	IVIG	51	CZP, Briva, OXC	no	no	no	yes	3-4/month
10	IVIG	6	PHT iv	yes	4	no	yes	2-4/month
		21.3			14.125			

ASM=anti-seizure medication; IVIG=intravenous immunoglobulin; AZA=azathioprine; LEV=levetiracetam; LCM=lacosamide; LTG=lamotrigine; CZP=clonazepam; CBZ=carbamazepine; VPA=valproic acid; PB=phenobarbital;

CONCLUSIONS

1. Hemispherotomy remains the option of choice with best results. When indicated, it should be performed at an early stage for a better cognitive outcome. 2. Immunotherapies may only slow the progress of the disease with less atrophy and less hemiplegic progression, but persistent seizures. 3. The cognitive outcome is similar in both surgical or immunomodulatory treatment. 4. Volumetry may be a reliable diagnostic tool. 5. Further systematic randomized clinical trials are needed to clarify the efficacy of non-surgical treatment options that might be used to control seizures and preserve neurological function.

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