A retrospective study of central nervous vasculitis patients

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CENTRAL NERVOUS SYSTEM VASCULITIS

The purpose of our study was to find relevant clinical data predicting progressive course in patients with central nervous system vasculitis (CNV).

We retrospectively studied the data of 18 patients treated with CNV between 1998 and 2022.

56% of patients were male, 44% were female.

Average age of onset was 8,6 years, (range 2,4 – 15 yrs).

17 patients had primary CNVs, and one patient had secondary CNV as part of a systematic vessel disease (SLE).

6 patients had non-progressive, 10 patients had progressive form, and 2 patients had negative MRI or MR angiography.

In the non-progressive form one patent had a post-varicella focal cerebellar arteriopathy.

Average diagnostic delay was 89 days, with a median of 29 days. Patients presenting with stroke had early diagnosis, while those with recurrent headache and transient neurological deficit usually had a longer delay, up to 375 days.

Criteria for non-progressive disease were monophasic course with unilateral and proximal vessel (ACA, ACM and ICA) involvement on MRI. We considered progressive form in patients with episodic disease course, bilateral, proximal and distal involvement on MRA and especially those with for borderzone lesions.

In the non-progressive (NP) form, presenting symptom was stroke, while TIAs were more common in the progressive (P) group.

DISCLOSURES

Authors have no objectives or conflicts of interest.

STUDY

Inclusion criteria: All patients between 1998-2022, with diagnosis of central nervous vasculitis.

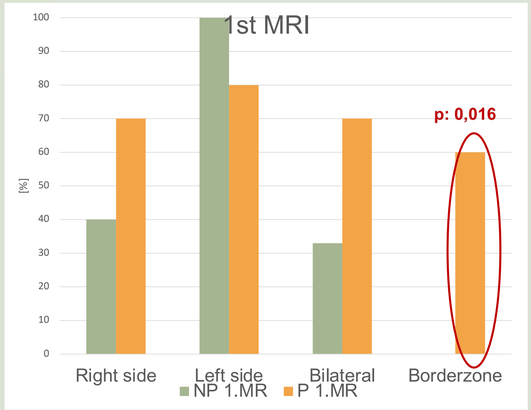
Goal: 1.) Comparison of non-progressive (NP) w progressive vasculitis (P) (symptoms, presenting neurologic status, laboratory findings, MRI, therapy and outcome). 2.) Finding data predictive of progressive vasulitis (P) at first presentation

RESULTS

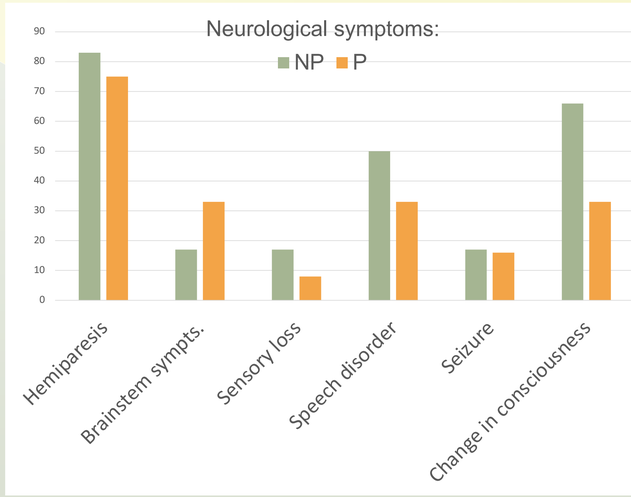
Symptomes	NP	%	P	%	p: 0,01
Headache	4	66	8	80	0,55
Stroke	5	83	5	50	0,18
Hemiparesis	4	66	8	80	0,05
TIAs	1	17	8	80	**0,013**
Repeated TIA-s	1	17	6	60	0,09

Laboratory findings	NP	%	P	%	p: 0,01
WBC ↑	1	17	3	30	0,55
PLT ↑	1	17	2	20	0,86
INR/APTI ↑	2	33	1	10	0,29
D-dimer/fibrinogen ↑	1	17	1	10	0,69
ESR ↑	2	33	3	30	0,55
CRP ↑	3	50	1	10	0,07

CSF	NP	%	P	%	p: 0,01
Cell count ↑	1	17	4	40	0,32
Total protein ↑	0	0	1	10	0,42
Abnormal CSF result	1	17	4	40	0,31



Combination of symptomes	NP	%	P	%	p: 0,01
Headache+stroke	3	50	2	20	0,21
Headaches+TIAs	0	0	8	80	**0,0019**
Headache+stroke/TIA	3	50	8	80	0,21
Headache+hemiparesis	2	33	6	60	0,3
Only headache	1	16	0	0	0,18



Immune therapy	cases	NP	P
escalation	8	0	8
HDMP	10	1	9
Per os steroids	14	4	10
CP	8	(1 -SLE)	7
AZA	2	0	2
MMF	1	0	1
IVIg	1	0	1
PLEX	1	1	0

Outcome: sequele: 7 (NP:4, P:3)

- Hemiparesis: 7 (tetraparesis: 1)
- Epilepsy: 1
- Mental retradation: 1
- Speech disorder: 1
- Behaviour disorder: 1

CONCLUSIONS

- Comparing non-progressive and progressive forms of vasculitis we found TIAs in the case history to be significantly more frequent in the progressive group. This significance became more marked if both TIAs and headaches were encountered.
- Borderzone lesions on the first MRI were also predictive of progressive vasculitis.

Etiological findings:

- 1 case had CSF HSV PCR positivity.
- 1 case w basilar artery stroke: Fabry disease is likely with low alfa galactosidase A activity and mutation c.937G>T(p.(Asp313Tyr)
- 1 case w MCA stroke and family history of aortic dissection: mutation in MYH11 gene, possibly resulting in dissection.
- 3 patients are still in consideration for other etiologies of stroke: 1 for thrombophilia and 2 for systemic vascular disease, because of extracerebral arterial involvement on MRA (external carotid artery).

If MRI shows ischemic lesions central nervous vasculitis should be considered. MRA and ”black blood” sequence images might help to differentiate vasculitis from other causes.

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