

Area postrema Syndrome as first NMOSD attack, a single center retrospective study

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Introduction

Neuromyelitis Optica Spectrum disorders (NMOSD) is an autoimmune astrocytopathy marked by demyelinating lesions in regions rich in Aquaporin-4 proteins¹. Area Postrema Syndrome (APS) is a clinical syndrome marked by persistent nausea, vomiting and hiccups lasting more than 48H after exclusion of more common causes². These symptoms are usually associated to dorsal medulla oblongata lesions in Magnetic Resonance Imaging (MRI), APS is a core clinical manifestation of NMOSD that is usually misdiagnosed and has delayed treatment^{2,3}. APS is associated to a higher relapse ratio, higher number of lesions at diagnosis and a higher chance of myelitis as further relapses, but usually are milder than other clinical syndromes and have good improvement with proper treatment if recognized³. The literature is still ambiguous about whether APS is associated to greater disability and there is a lack of data on the Brazilian population.

Objectives

To analyze the diagnostic challenges faced by patients living with NMOSD and to measure if there are differences among patients with APS as first attack in terms of diagnostic delay and proper treatment in this population in Northeast Brazil.

Methods

Data was collected from patients’ records and interviews about their first symptoms. NMOSD diagnosis was made following the 2015 criteria¹. Area Postrema Syndrome was defined based on the 2018 proposed criteria². The patients were divided in two groups: Group APS, including those with isolated APS symptoms as first clinical attacks, and Group Non-APS including those with other clinical syndromes or APS combined with other syndromes as first manifestation. Data was then analyzed using R software to compare the two groups.

Results

The number of patients was 74, with average age of onset 36 years. Median time to diagnosis was 7 months. 51 patients were Aquaporin-4 antibody positive (68.9%). APS was the first syndrome in 12 patients (16.2%) and 62 were in the Non-APS group. The APS had similar hospitalization rate (41.6%) compared to Non-APS group (64.5%) (p=0.24). We found no difference in EDSS disability scale at last follow-up, median 3.5 in APS group and 5 in Non-APS group (p=0.18). All APS patients were only referred to neurology or underwent MRI after relapses. The most common professional searched by APS patients was gastroenterology (50%) and 58.3% were submitted to endoscopy, while the ophthalmologist was the most between Non-APS group.

	Group APS (n 12)	Group Non-APS (n 62)	APS vs Non-APS p [*]
Days to first clinic visit, median (min-max)	7 (1-90)	4.5 (1-2922)	0.87
Days to first Neurology visit, median (min-max)	232.5 (60-2922)	25 (1-4383)	0.001
Days to diagnosis, median (min-max)	344 (109-6726)	182 (12-4730)	0.09
Days to first DMD median (min-max)	344 (109-6726)	164 (12-4766)	0.051
Attacks before diagnosis, median (min-max)	3 (2-5)	2 (1-5)	0.001
Acute Treatment, n (%)	0 (0)	45 (72.5)	<0.001

Journey of patient to diagnosis and treatment at first attack. DMD, disease-modifying drugs. #Wilcoxon-rank test used to numerical variables and Chi-square test to categorical variables

Conclusion

The Area postrema syndrome as first clinical attack was associated to longer delay to neurologist visit, to higher relapse ratio before diagnosis and to lower chance of having acute treatment in the first attack. We also found a tendency to delay to diagnosis, but no difference in disability scales at follow-up. This clinical manifestation can be an invisible warning sign to relapses and need awareness to be recognized by physicians.

References

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