ADEM, para-infectious and post-vaccination optic neuritis. Analysis of a large cohort.

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Introduction:

Subacute loss of vision may occur as the opening symptom of several immune-mediated demyelinating diseases of the central nervous system (DD-SNC), including ADEM, para-infectious and post-vaccination optic neuritis (ON). Published data on ON characteristics in these conditions are scarce.

Objective:

To study the characteristics of patients with ADEM-ON, post-infection ON (PI-ON), and post-vaccination ON (PV-ON) in a large cohort.

Methods:

We selected patients with ON who met Krupp 2013 diagnostic criteria for ADEM, Rappoport diagnostic criteria for para-infectious ON, and Karussis diagnostic criteria for post-vaccination ON. Demographic, clinical, laboratory and imaging characteristics were analyzed. Kurtzke Visual Function System Score (KVFSS) and Wingerchuk Optic Nerve Impairment Score (WONIS) were used to evaluate outcome.

Results:

Out of 955 patients with DD-SNC seen between 2010 and 2019, 311 presented ON at disease onset. 40 patients were excluded. Out of 271 patients, ADEM-ON, PI-ON and PV-ON comprised 28 (10.3 %) patients - 7 (2.6%) ADEM-ON; 7 (2.6%) PI-ON; and 14 (5.2%) PV-ON. Median age at onset was 23 (1.3-6.1) years; 19 (68%) were females, and 16 (59%) non-whites. In 17 (61%) ON was bilateral and associated with serum autoantibodies in 5/23 (21.7%%). Specific-CSF oligoclonal bands were found in 1/16 patients (6.3%), and longitudinally extensive optic nerve lesion was seen on MRI in 10/14 (71.4%) patients. Enhancing of the ON lesion was found in 63.6%. Median KVFSS was 2 (1-4) and median WONIS 1 (0-3).

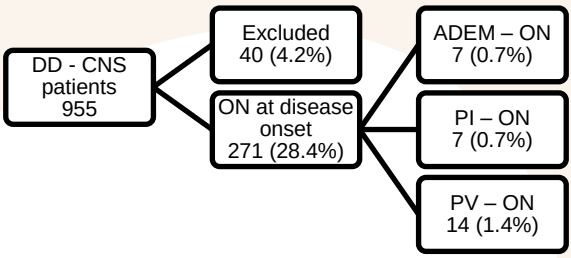

Figure 1. Clinical characteristics of study patients (and overall percentage of incidence).

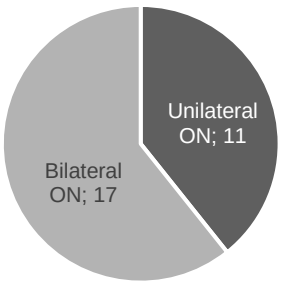

Figure 2. ON characterization of patients who had ADEM-, PI- or PV-type ON.

ADEM-ON, PI-ON and PV-ON are rare forms of ON which share similar pathophysiological mechanisms. Taken together they predominantly occur in children and young adults, are more frequently bilateral and associated with gadolinium-enhancing longitudinally extensive optic nerve lesion. The visual outcome is usually good.

Conclusions:

Identification of ADEM-ON, PI-ON and PV-ON among other forms of ON at onset of DD-SNC may have predictive value for favorable visual outcome.

References:

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