ESO Guideline Webinar, 30 October 2023, Q&A

Responses to questions on ESO Guideline on Primary Angiitis of the Central Nervous System (PACNS) by the guideline chair Marialuisa Zedde, AUSL di Reggio Emilia, Italy

- *Where clinical suspicion is high for PACNS, would you start corticosteroids before confirming diagnosis?*

The question is interesting and addresses a common clinical dilemma. The early starting of steroid treatment is not really supported and, in some cases (e.g. SV-PACNS or tumefactive PACNS), one of the potential differential diagnoses is PCNSL and in this case, steroid treatment might be an obstacle for the diagnosis.

- *In a case of suspected PACNS, what would be your first battery of tests?*

It depends on the level of suspicion, because I think it is complex to have a "clinical" suspicion alone without neuroimaging support in the last years, so clinical and neuroimaging data help to preliminary set a range of hypothesis including vasculitis (not necessarily PACNS, but secondary vasculitides too have to be considered). Usually, clinical presentation is a good starting point to suspect or exclude RCVS. Extended autoimmunity testing and (in some circumstances) searching for autoimmune (one of the examples might be neurosarcoidosis, but also ANCA associated vasculitides might have a cerebral involvement and IgG4 diseases not always involve pachymeninges) infective causes is necessary (serological and CSF analysis are standardized, but you should consider VZV arteriopathy a potential cause, looking for anti VZV antibody serum/CSF ratio).

- *Where the PACNS presents with haemorrhage, is there still a role for antiplatelet?*

Usually, antiplatelets have not a significant role in PACNS, but strong data are missing. At the moment, it is hard to consider these drugs in hemorrhagic forms.

- *What is the suggested dosing for corticosteroids in PACNS?*

In different case series, different regimens have been used and for different durations with or without IV administration. Usually, as for other autoimmune diseases, IV methylprednisolone (1000 mg) is considered as starting treatment, for 3-5 days, followed by a very slow tapering off per os (prednisone 1 mg/Kg) lasting several months.