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FRI0287

EFFICACY OF EOSINOPHILIC GRANULOMATOSIS WITH POLYANGIITIS TREATMENTS ACCORDING TO THE TYPE OF MANIFESTATIONS BASED ON ANALYSIS OF 636 PATIENTS

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Background: Eosinophilic granulomatosis with polyangiitis (EGPA) (Churg–Strauss) is a small-vessel necrotizing vasculitis characterized by blood and tissue eosinophilia and asthma. Glucocorticoids (GCs) represent the treatment cornerstone. So far, EGPA management has been based on conventional immunosuppressants, but GC-dependence remains frequent. Recently, therapies targeting B cells and interleukin-5 have been prescribed, but data on large cohorts are lacking.

Objectives: This study aimed to describe therapeutic management and efficacy of treatments in EGPA patients.

Methods: We set up a multicenter European cohort that included 636 EGPA patients. Treatments used, complete remission rates and vasculitis relapse-free survival were recorded. Complete remission was defined as absence of vasculitis relapse and prednisone dose <5 mg/d at last follow-up. Efficacy to treat GC-dependent asthma/ENT signs was defined as the absence of asthma/ENT symptoms and prednisone dose ≤7.5mg/d within the 6 months after initiation.

Results: For induction, cyclophosphamide (CYC) was the most frequently prescribed immunosuppressant (36.2%), more often in patients with FFS ≥1 (P<0.0001). GCs alone were used in 37.3%, azathioprine (AZA) in 14.4% and methotrexate (MTX) in 6.2%. No difference was found in the 10-years overall survival between patients with FFS=0, FFS=1 and FFS≥2.

Complete remission rates were similar between conventional immunosuppressants (CYC, AZA or MTX) and GCs alone. Vasculitis relapse-free survival was also similar between CYC, AZA or MTX and GCs alone. Similar results were observed for first vasculitis relapse treatments.

During follow-up, GC-dependent asthma and/or ENT manifestations were treated with AZA (40%), MTX (25%), mycophenolate mofetil (16%), rituximab (RTX) (21%), CYC (19%), cyclosporine (6%), omalizumab (5.9%) and mepolizumab (5.5%), allowing GC-tapering ≤7.5mg/in 23%, 31%, 17%, 43%, 5%, 71%, 25% and 50%, respectively. Conventional immunosuppressants were mostly used in first and second line, while eosinophil-targeted biotherapies were used in 4th or 5th lines.

Conclusion: In EGPA patients, the response to conventional immunosuppressants, in addition to GCs, is often disappointing compared to GCs alone, without clear benefit on complete remission rates and relapse-free

survival. In contrast, notwithstanding a small number of treated patients, eosinophil-targeted therapies seemed promising to treat asthma and/or ENT manifestations.

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OPHTHALMOLOGICAL MANIFESTATIONS AND ENDOTHELIN-1 PLASMA LEVELS IN PATIENTS WITH SYSTEMIC NECROTIZING VASCULITIS (SNV)

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Background: Inflammatory eye disease is described in 50% to 60% of patients with ANCA-positive vasculitis and in 10–20% of patients with polyarteritis nodosa, and for 8% to 16% of patients it is an initial manifestation [1,2]. Endothelin-1 (ET-1) as a potential participant in the local regulation of intraocular pressure, ocular blood vessel tone, and iris smooth muscle tone, suggesting that it may be an important mediator in the development of ocular pathologic conditions [3]. The lower ET-1 plasma levels were found in the optic neuropathy [4].

Objectives: to provide a more complete description of the ocular disease in patients with systemic necrotizing vasculitis (SNV), to evaluate the serum level of ET-1 in patients with SNV with and without eye involvement.

Methods: The study included 36 patients with SNV (polyarteritis nodosa - 8, ANCA - associated vasculitis - 28) and healthy controls (n=26). The 17 patients with SNV had ophthalmological manifestations. Clinical activities of patients were calculated according to the Birmingham Vasculitis Activity Score (BVAS). The serum levels of ET-1 (pmol/L) were determined by immunoassay analysis using the kits of Biomedica. The outcomes of this study were the differences in marker levels between SNV patients with and without eye involvement and healthy controls estimated by analysis of the absolute changes in marker levels and the areas under receiver operating characteristic (ROC) curves (AUC).

Results: The ocular manifestation of patient with SNV included episcleritis (n=10), anterior uveitis (n=3), ischaemic optic neuritis (n=3) and occlusive retinal vasculitis (n=1). All patients had active disease (BVAS>11). There were no significant differences of BVAS, ESR and CRP between SNV patients with and without eye involvement. In 14% patients with SNV eye involvement was an initial manifestation. The level of ET-1 (M ± σ) in group of SNV patients with eye involvement (n=17) was 0.28 ± 0.13 and did not differ significantly from the control group (0.27 ± 0.10, p> 0.05). At the same time, in patients without eye involvement (n=19), it was significantly elevated (0.36 ± 0.34) compared with control group and with group of SNV patients with eye involvement (p < 0.001). ROC analysis showed that the AUC for ET-1 is 0.5±0.10 (p=0.98), which indicates not acceptable capacity for ET-1 differentiate groups of patients with ocular involvement and patients without ocular involvement (sensitivity - 59%, specificity - 57%).