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RESULTS OF A SYSTEMATIC LITERATURE REVIEW INFORMING THE 2018 UPDATE OF THE EULAR RECOMMENDATIONS FOR THE MANAGEMENT OF LARGE VESSEL VASCULITIS: EVIDENCE TO GUIDE THE MANAGEMENT OF GIANT CELL ARTERITIS

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Background: The latest EULAR recommendations for the management of large vessel vasculitis (LVV) were published in 2009 (1). Since then, imaging has become a reliable diagnostic tool and new therapeutic options are available for giant cell arteritis (GCA), supporting the need to update the recommendations.

Objectives: To analyse the current evidence for the management (diagnosis/monitoring and treatment) of LVV to inform the 2018 update of the EULAR recommendations.

Methods: A systematic literature review (SLR) dealing with diagnosis/monitoring and treatment strategies for LVV, respectively, was performed. Medline, Embase and Cochrane databases were searched from inception until 31st December 2017. Evidence on imaging was excluded in light of recently published EULAR recommendations (2). We reviewed data relevant to GCA. Level of Evidence (LoE), was assessed in accordance with the 2009 Oxford Centre for Evidence-based Medicine.

Results: We identified 283 papers from the SLR. The implementation of a fast-track approach to diagnosis significantly lowered the risk of permanent visual loss compared to historical cohorts (Level of evidence – LoE 2b). The SLR confirmed the efficacy of prompt initiation of glucocorticoids (GC). There was no high-quality evidence on the most appropriate starting dose, route of administration, tapering and duration of GC (LoE 4). Patients with GCA are at increased risk of dose-dependent GC-related adverse events (LoE 3b). The addition of methotrexate (MTX) (LoE 1a) or tocilizumab (TCZ) reduced relapse rates and GC requirements (LoE 1b). There was no consistent evidence that anti-platelet agents given at the time of GCA diagnosis prevented future ischaemic events (LoE 2a). There was little evidence to guide monitoring of patients with GCA.

Conclusion: Results from a SLR confirmed the importance of a prompt diagnosis and rapid initiation of GC therapy. Patients with GCA are at an increased risk for GC-related comorbidities. MTX and TCZ can reduce GC exposure and relapse rates. There are yet no high-quality data to guide monitoring and duration of treatment in GCA.

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CUTANEOUS POLYARTERITIS NODOSA: A SUBSET OF SYSTEMIC PAN OR A DISEASE APART?

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Background: Cutaneous Polyarteritis Nodosa (cutaneous PAN) is a rare form of necrotizing vasculitis involving predominantly medium and small arteries. It is limited to the skin. The clinical course is mostly chronic, however it has a more favourable prognosis than systemic PAN. Cutaneous PAN may be preceded by infection, drugs intake or may reflect underlying inflammatory or autoimmune disease. The diagnostic criteria are controversial and in some cases overlapping with those of systemic PAN. Cutaneous PAN is limited to the skin, but occasionally patients present extracutaneous symptoms, namely: myalgia, arthralgia or multineuritis

Objectives: To describe the clinical manifestations of cutaneous PAN, emphasizing the extracutaneous manifestations, as well as comorbid associations.

Methods: Retrospective study (1989-2018) of patients with cutaneous PAN seen in a tertiary center: referral area 800.000 inhabitants. All the cases were reviewed regarding epidemiological, clinical, electromyographic, histopathological and laboratory findings. Infections and comorbid diseases were also reviewed.

Results: Twenty-three cases were recruited: 8 male (34.78%) and 15 female (65.22%) patients. Mean age at onset: 47 years (range 18-76). Mean follow-up period :105 months (range 12-380). Cutaneous manifestations included: subcutaneous nodules (95.65%), ulcers (52.17%), livedo *reticularis* (60.87%), livedo *racemosa* (8.70%), *atrophie blanche* (39.13%) and purpura (4.34%). Cutaneous PAN most commonly occurred on the legs (100%), followed by the thighs (47.83%), the upper limbs (39.13%) and the trunk (17.39%). The most frequent extracutaneous symptom were asthenia (78.26%). Extracutaneous manifestations, limited to the same area of cutaneous lesions, were myalgia (60.87%) and paraesthesia (56.52%). An electromyography was performed in 14/23 patients and mononeuritis multiplex was observed in 7 patients. Only one patient progressed to systemic PAN, with development of renal disease 12 month after the diagnosis. The following infections were identified: hepatitis C (2), tuberculosis (1), *Streptococcus pyogenes* (1) and rickettsia (1). Comorbid diseases included: autoimmune hypothyroidism (2), cutaneous psoriasis (1), recidivant polychondritis (1), adult onset Still's disease (1), ulcerative colitis (1), pulmonary idiopathic fibrosis (1) and acute myeloid leukaemia (1). Glucocorticoids were used at medium-high doses (20-40 mg/day) in 20 patients as induction therapy, and NSAIDs at full doses in the remaining 3. Thirteen patients (56.52%) developed relapsing forms. Of this, 12 patients (52%) needed associated immunosuppressive treatment to prevent relapse. The most widely used immunosuppressant was methotrexate, followed by mycophenolate or azathioprine.

Conclusion: A long-term follow-up, with careful anamnesis and examination, including electromyography, is necessary to better characterize the cutaneous and extracutaneous manifestations of cutaneous PAN. The progression to systemic involvement is infrequent and cutaneous PAN has a more benign course from a clinical, and prognostic point of view.