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AB0582

NEUROLOGIC INVOLVEMENT IN ANTINEUTROPHIL CYTOPLASMIC ANTIBODY (ANCA)-ASSOCIATED VASCULITIS –EXPERIENCE FROM A PORTUGUESE CENTER

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Background: Involvement of peripheral (PNS) and central nervous system (CNS) in antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis has been reported in 15-60% and 5-15% of patients (pts), respectively. PNS involvement occurs mainly in eosinophilic granulomatosis with polyangiitis (EGPA) and CNS involvement has been almost exclusively described in granulomatosis with polyangiitis (GPA). Neurologic involvement is associated with a higher morbidity and implies more immunosuppressive treatment in most pts.

Objectives: Evaluate prevalence and clinical features of PNS/CNS involvement in pts with ANCA-vasculitis in a Portuguese center.

Methods: Retrospective analysis of ANCA-vasculitis pts with neurologic involvement followed in our department from January 2016 to December 2018. Data was collected from the Portuguese database REUMA.PT and included demographic data, vasculitis subtype, date of diagnosis, neurologic manifestations and treatment approach.

Results: In total, 11 pts were identified, mostly female (7/11) with mean age of 61±13.3 years (yrs). Five pts had EGPA, 5 GPA and 1 microscopic polyangiitis (MPA). Neurologic involvement was part of disease presentation in 8 pts (5 EGPA, 3 GPA). The other 3 pts had neurologic involvement 7±2.2 yrs after vasculitis diagnosis. At the time of neurologic involvement diagnosis, Birmingham Vasculitis Activity Score was 8.8±4.3. PNS involvement occurred in 9 pts, all presenting mononeuritis multiplex (MM). From these, 5 had EGPA, 3 GPA and 1 MPA.

CNS involvement was reported in 3 pts, 2 with GPA and 1 with EGPA. Pachymeningitis was diagnosed in 1 GPA pt with persistent headache, refractory to analgesics, and thickening and enhancement of the dura mater on postcontrast magnetic resonance imaging. Two pts had cranial mononeuropathy, 1 with EGPA and MM who developed VI palsy 2 yrs after vasculitis diagnosis and 1 pt with GPA and VII palsy at disease presentation who developed XII palsy 8 yrs later.

All pts were treated with prednisolone (1mg/kg/day), mostly in combination with other immunosuppressive drugs. The choice of the treatment was based on the age of the pt and other comorbidities.

Detailed treatment of these pts and subsequent responses are shown in table 1.

Table 1 – Detailed data on patients with neurologic involvement in relation to ANCA-vasculitis

Patient	Gender/Age	ANCA-vasculitis	Interval from vasculitis onset to neurologic manifestations (years)	Type of neurologic involvement	Evolution
1	Male / 54	GPA	0	Mononeuritis multiplex	Improvement with CYC; disease progression after RTX + MMF (IVig as bridging therapy until RTX retreatment)
2	Female / 69	GPA	0	Mononeuritis multiplex	Improvement with RTX + IVig
3	Male / 43	GPA	10	Mononeuritis multiplex	Less than 6 months of treatment
4	Male / 78	GPA	6	Pachymeningitis	Improvement with RTX
5	Female / 36	GPA	0	VII palsy	Resolution with CYC
6	Female / 73	MPA	5	Mononeuritis multiplex	No response to RTX; improvement after 3 months on CYC
7	Female / 71	EGPA	0	Mononeuritis multiplex	Resolution with PDN
8	Male / 47	EGPA	0	Mononeuritis multiplex	Less than 6 months of treatment
9	Female / 61	EGPA	0	Mononeuritis multiplex	Improvement with CYC, followed by maintenance with AZA
10	Female / 69	EGPA	0	Mononeuritis multiplex	Improvement with IVig followed by AZA
11	Female / 70	EGPA	0	Mononeuritis multiplex	Improvement with IVig
			2	VI palsy	Resolution with RTX

Legend – GPA – granulomatosis with polyangiitis; MPA – microscopic polyangiitis; EGPA – eosinophilic granulomatosis with polyangiitis; CYC – cyclophosphamide; RTX – rituximab; MMF – mycophenolate mofetil; IVig – intravenous human immunoglobulin; PDN – prednisolone; AZA – azathioprine

Figure 1

In our cohort there were 2 pts with serious infections: 1 with oropharyngeal and esophageal candidiasis and another with bacteraemia to *Pseudomonas aeruginosa*. Both pts were under immunosuppressive agents including corticosteroids and rituximab or cyclophosphamide.

Conclusion: Neurologic involvement was part of disease presentation in most pts and the commonest manifestation was mononeuritis multiplex. PDN was prescribed to all pts, in most cases in association with other immunosuppressive drugs. Cyclophosphamide and rituximab (RTX) were used as induction treatment, and mycophenolate mofetil, azathioprine and RTX as maintenance. Intravenous human immunoglobulin was used in pts colonized by multiresistant microorganisms/severe infection with immunosuppression and as a bridging therapy to further immunosuppression. Most pts achieved clinical improvement, documented in electromyography.

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100 GCA REFERRALS TO A TERTIARY CARE CENTRE: INFLAMMATION, HEADACHES AND CPAP MASKS

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Background: Giant cell arteritis (GCA) is a large vessel vasculitis typically affecting patients above the age of 50 years. It is the most common cause of vasculitis affecting adults with a UK prevalence of approximately 0.25% of the population above the age of 55 years [1]. Whilst its clinical presentation can often be non-specific, if left undiagnosed it can lead to devastating visual impairment. However, this has to be balanced with the potential for corticosteroid related adverse effects. The threshold for suspected diagnosis and referral to rheumatology centres from the community and acute medical services is therefore low, leading to a relatively high number of referrals with a wide range of clinical features and potential differential diagnoses.

Objectives: To assess the referral characteristics, demographics, management, corticosteroid adverse effects and differential diagnosis of 100 consecutive patients referred with suspected GCA to a tertiary rheumatology centre.

Methods: The notes of 100 consecutive patients referred to the rheumatology department at Addenbrookes hospital between August 2016 and January 2018 with suspected GCA were reviewed. Information on patient demographics, comorbidities, presenting symptoms, availability of up to date inflammatory markers, dates of treatment and clinic review, temporal artery biopsy (TAB) date and result, diagnosis and corticosteroid adverse effects were retrieved.

Results: Seventy three female and twenty seven male patients (average age 72.3 years) were referred within the time period. General practice referred 69% of the cases and common comorbidities included hypertension and type II diabetes. The most common symptoms were headache (97%), scalp tenderness (79%), jaw claudication (35%) and polymyalgia symptoms (34%). Up to date inflammatory markers were available for 91% of referrals and 92% were commenced on corticosteroids before or at the time of referral with an average starting dose of 43.9mg oral prednisolone. A TAB was performed on 85% of patients, with an average time of 9 days between referral and TAB and 40 days between referral and clinic appointment. TAB was positive for GCA in 13/85 patients. Temporal artery ultrasound (TAU) was not available at our centre. The American College of Rheumatology (ACR) classification criteria for GCA were met by 85% of the patients, but only 69% were given a diagnosis of GCA. Other diagnoses included trigeminal neuralgia, temporo-mandibular dysfunction, toothache and a poorly fitted CPAP mask. Corticosteroid adverse effects were experienced by 45% of patients including weight gain, poorly controlled diabetes, mood and sleep disturbance, and 4% suffered severe complications requiring hospital admission (pneumonia, disseminated nocardia infection and two episodes of upper gastrointestinal bleed).

Conclusion: Suspected GCA is a common referral to rheumatology but the non-specific symptoms, quality of referral and potential for visual loss can lead to over-diagnosis. We have found the majority of our referrals to have been appropriate and complete with 85% meeting ACR criteria for GCA and 92% having up to date inflammatory markers. Despite this, 31% of patients received an alternative diagnosis. Increasing the availability of