Compared with patients diagnosed at respiratory medicine, patients diagnosed at rheumatology had a higher frequency of fever (33.3% vs. 0), rash (33.3% vs. 4.5%), arthralgias (19.0% vs. 0) and limb numbness (57.1% vs. 22.7%), but lower frequency of asthma-like symptoms (66.7% vs. 100%; all $P < 0.05$). There were more patients diagnosed at rheumatology received treatment of glucocorticoid (100% vs. 72.7%), immunosuppressive agents (85.7% vs. 4.5%) and intravenous immunoglobulin (28.6% vs. 0) than those diagnosed at respiratory medicine (all $P < 0.01$). Patients diagnosed at rheumatology also received higher initial dose of glucocorticoid than those diagnosed at respiratory medicine [50(50~100) mg/d vs. 28(10~40) mg/d of prednisone dosage, $P < 0.001$].

4) Among the 21 patients diagnosed at rheumatology, 14(66.7%) had asthma-like symptoms, who showed significant longer time required for diagnosis than those without asthma-like symptoms [36(24~120) months vs. 2(2~13) months, $P = 0.020$], but showed no significant difference of onset age between these two groups ($P > 0.05$).

5) The median revisited Five-Factor Score (FFS) was 0(0~1). There were 17(37.8%) patients had FFS ≥ 1 , who had older diagnostic age than those with FFS=0 [65(46~68) years vs. 48(42~53) years, $P < 0.05$], but with no significant difference of onset age or time required for diagnosis. Patients with FFS ≥ 1 had higher level of serum IgE [1280(515~4895) IU/mL vs. 243(72~587) IU/mL] than those with FFS=0 ($P < 0.05$).

Conclusion: Clinical manifestations of EGPA were different between rheumatology and respiratory medicine. Rheumatologist may be more aggressive treating EGPA patients with glucocorticoid and immunosuppressive agents than respiratory physician. Multidisciplinary team is needed for the diagnosis and management of EGPA.

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AB0580

A VISION OF THE CHARACTERISTICS AND COMORBIDITIES OF GIANT CELL ARTERITIS FROM THE HOSPITAL RECORDS OF THE NATIONAL HEALTH SYSTEM

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Background: Giant cell arteritis (GCA) is the most frequent systemic vasculitis in older than 50 years old, with serious repercussions on the quality of life and survival of patients. The low prevalence and heterogeneous presentation make progress in the diagnostic and the knowledge of the disease difficult.

Objectives: The objective of this study has been to gather a large database to analyze the epidemiological characteristics, the comorbidities and the causes of mortality associated with the hospitalizations by ACG in our country.

Methods: Retrospective observational study based on data from the Database of Hospital from the Spanish National Health Service with primary or secondary diagnostic of GCA between January 1st, 2005 and December 31st, 2015. The included variables were sex, age, main reason for admission, secondaries diagnostics, time of stay, costs, comorbidities, temporal artery biopsy and mortality.

Results: The study cohort included 29,576 patients with GCA, 18,568 (62.8%) women, and the mean age was 80 ± 8 years. The number of hospital admissions with this diagnostic has increased between 2008 and 2013 with an annual cumulative increase rate of 3%, passing from 2,487 to 2,997 patients/year. We have found a different disease affection pattern in men and women in terms of distribution by age group and the comorbidities associated. We have found that the mortality risk increases with the age and with the presence of cardiovascular disease OR 1,816 (1,484-2,221), ischemic heart disease OR 1,590 (1,302-1,941), renal disease OR 1,386 (1,225-1,569) and cancer OR 2,428 (2,039-2,891). While, a decrease in the risk of death was observed in patients who associated polymyalgia rheumatica OR 0.682 (0, 5810,801).

Table 1. Comorbidities and mortality in hospitalized with ACG

	Men		Woman		Both sexes	
	N	%	N	%	N	%
DM2 *	2948	26,8%	4669	25,1%	7617	25,8%
PMR *	1474	13,4%	2715	14,6%	4189	14,2%
ICC*	1298	11,8%	2854	15,4%	4152	14%

ENF.		1556	14,1%	2413	13,0%	3969	13,4%
RENAL *							
Cancer		809	7,3%	499	2,7%	1308	4,4%
IHD *		626	5,7%	565	3,0%	1191	4%
ECV *		502	4,6%	651	3,5%	1153	3,9%
EPOC		289	2,6%	559	3,0%	848	2,9%
CCI*	No	4833	43,9%	9174	49,4%	14008	47,4%
	comorbidities						
	≤ 2	5635	51,2%	8697	46,8%	14332	48,5%
	Comorbidities						
	> 2	539	4,9%	697	3,8%	1236	4,2%
	Comorbidities						
Mortality		712	6,5%	1083	5,8%	1795	6,1%

*Statistically significant differences ($P < 0.05$) DM2: Diabetes mellitus Type 2 PMR: Polymyalgia rheumatica ICC: Congestive heart failure IHC: ischemic heart disease ECV: Cerebrovascular disease CCI: Charlson Index of Comorbidities

Conclusion: In recent years the number of GCA hospital admissions has increased. The presence of comorbidities is high and greater in men. An increase in deaths for ischemic cause and a reduction of mortality in patients with PMR associated is observed.

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AB0581

AN ATYPICAL PRESENTATION OF GIANT CELL ARTERITIS WITH SCALP LUMPS: A CASE REPORT

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Background: Giant cell arteritis (GCA) also referred as temporal arteritis is a systemic inflammatory vasculitis of unknown etiology which can result in wide range of systemic, neurologic and ophthalmic complications. It predominantly occurs in older people and may be associated with Polymyalgia Rheumatica. It must be treated urgently as it is associated with significant risk of permanent visual loss, stroke, aneurysm and possible death. Diagnosis of GCA can be based on clinical findings such as headache, scalp tenderness, jaw claudication and temporal artery problems, an elevated ESR and biopsy temporal artery showing typical histopathological findings. Diagnosis of GCA may be delayed when presentation is atypical.

Objectives: We report a case of an elderly lady presenting with scalp lump as an unusual presentation of GCA.

Methods: 70 years old lady presented with multiple painful scalp lumps to the surgeon was referred to the Rheumatologist when the excision biopsy of the lump diagnosed the GCA. (Sub cutis medium sized artery with severe arteritis composed with lymphocytes, neutrophils and occasional Eosinophils). On enquiry she revealed a history of generalized malaise and lethargy at the outset of the illness preceded by jaw pain. She had been initially referred to the dental surgeon which resulted in multiple carious tooth extractions without a relief. She didn't have any eye symptoms or headache at the outset. She lately developed headache and pain on touching the lumps.

On examination she had bilateral hardened thickened palpable non pulsatile temporal arteries. Her scalp was tender with multiple small cystic lumps. Her ESR was highly elevated 141mm/hr. WBC 9.7, Hb 9.5 and PLT 283. Her ANA, liver and renal profiles were normal. Her BL Doppler USS was consistent with BL temporal arteritis.

Results: She was soon started on high dose prednisolone with low dose aspirin which improved her systemic symptoms along with inflammatory markers significantly over a few days. She was referred for eye assessment and the involvement of other systemic larger arteries was ruled out.

Conclusion: Most of the cases the diagnosis of GCA is obvious but in patients with uncommon clinical presentation the diagnosis may be difficult. GCA is classified as disease of large vessel vasculitis but typically also involves medium and small arteries. The granulomatous inflammation in the vessel wall leads to obstruction of the lumen and ischemia in the territory distal to it. GCA involves almost all speciality and it is associated with increased mobility and mortality if not diagnosed and treated early. Awareness of rare presentation of GCA avoids unnecessary investigation and delaying the treatment. If there is no rapid improvement of symptoms within few weeks of starting treatment other diagnosis should be considered.

REFERENCES

- [1] Gunawardena H, Kyle M.V, Atypical presentation s of Giant cell arteritis-A District General Hospital perspective, *Rheumatology*, volume 44, Issue suppl_3, 1 July 2005, pages iii17-iii18
- [2] Weyand CM, Goronzy JJ. Giant cell arteritis and Polymyalgia rheumatica. *N Engl J Med* 2014;371:50-7
- [3] Shambu.S, Suarez. L, Giant cell arteritis: An atypical presentation diagnosed with the use of MRI imaging, *Case Reports in Rheumatology*, volume 2016, article ID8239549

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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.

REFERENCES

- [1] Graf J. Central Nervous System Disease in Antineutrophil Cytoplasmic Antibodies–Associated Vasculitis. *Rheum Dis Clin N Am*. 2017 Nov;43(4):573–8.
- [2] Wolf J, Schmitt V, Palm F, Grau AJ, Bergner R. Peripheral neuropathy as initial manifestation of primary systemic vasculitides. *J Neurol*. 2013 Apr;260(4):1061–70.

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AB0583

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