

Supplementary material

Supplementary table S1. Comorbidities in AAV patients with ENT involvement

Comorbidities	N (%)
myocardial infarction	15 (7.2)
congestive heart failure	11 (5.3)
peripheral vascular disease	15 (7.2)
cerebrovascular disease	12 (5.8)
hemiplegia	1 (0.5)
dementia	5 (2.4)
chronic pulmonary disease	53 (25.4)
peptic ulcer disease	6 (2.9)
diabetes without complications	36 (17.2)
diabetes with complications	1 (0.5)
renal disease	45 (21.5)
malignancies including lymphoma	35 (16.7)
metastatic solid tumor	7 (3.3)
mild liver disease	2 (1.0)
moderate/severe liver disease	0 (0.0)
AIDS/HIV	0 (0.0)

AIDS: Acquired Immunodeficiency Syndrome; HIV: human immunodeficiency virus;

Supplementary table S2. laboratory values at diagnosis and last visit in AAV patients with ENT involvement

Laboratory values	At diagnosis	At last visit
ANCA titer, median (IQR)	67.0 (21.0-207.5)	5.9 (1.3-23.0)
CRP, median (IQR)	34.0 (10.5-92.0)	3.0 (1.0-11.0)
ESR, median (IQR)	60.5 (25.5-96.8)	13.0 (4.0-21.0)
Leukocyte count, median (IQR)	10.7 (8.4-14.8)	7.8 (6.2-10.4)
eGFR >60, N (%)	63 (71.6)	138 (69.0)
Serum creatinine, median (IQR)	83.0 (69.0-134.0)	83.0 (68.0-113.0)
Proteinuria, N (%)	48 (38.4)	46 (24.2)

ANCA: Anti-neutrophil cytoplasmic antibody; CRP: C-reactive protein; eGFR: estimated glomerular filtration rate; ESR: erythrocyte sedimentation rate; IQR: interquartile range;

Supplementary table S3. Different types of ENT involvement at diagnosis and last visit

ENT involvement	at diagnosis N (%)	at last visit N (%)
Nasal bloody discharge	74 (49.0)	17 (9.3)
Nasal crusts	86 (57.0)	35 (19.0)
Nasal ulcers	6 (4.2)	1 (0.5)
Nasal granulomata	8 (5.6)	1 (0.5)
Paranasal sinus involvement	56 (36.4)	6 (3.3)
Subglottic stenosis	16 (10.3)	17 (9.3)
Conductive hearing loss	19 (12.7)	4 (2.2)
Sensoneurinal hearing loss	8 (5.3)	7 (3.9)
Saddle nose deformity	11 (6.9)	24 (12.9)

Supplementary table S4. History of prescribed medication of AAV patients with ENT involvement

Medication		N=209
Steroid maximum daily dose median (IQR)		60.0 (60.0-60.0)
Duration steroid therapy in weeks, median (IQR)		256.0 (85.3-368.5)
Cumulative cyclophosphamide dose in mg, median (IQR)		9510 (6730-13073)
Induction therapy, N (%)	Methylprednisolone pulse therapy	58 (27.8)
	Cyclophosphamide	137 (65.6)
	Plasmapheresis	7 (3.3)
	Mofetil mycophenolate	4 (1.9)
	Azathioprine	1 (0.5)
	Methotrexate	19 (9.1)
	Rituximab	55 (26.3)
	IVIG	1 (0.5)
	Cyclosporine	1 (0.5)
	Mepolizumab	1 (0.5)
	Omalizumab	1 (0.5)
Maintenance therapy, N (%)	Cyclophosphamide	1 (0.5)
	Mofetil mycophenolate	33 (15.8)
	Azathioprine	150 (71.8)
	Methotrexate	22 (10.5)
	Rituximab	32 (15.3)
	Hydroxychloroquine	1 (0.5)
	Cyclosporine	2 (1.0)
	Mepolizumab	4 (1.9)

IQR: interquartile range; IVIG: intravenous immune globulin,