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Response to treatment of inflammatory pseudotumours in granulomatosis with polyangiitis based on case series

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Introduction: Granulomatosis with polyangiitis (GPA) is a rare anti-neutrophil cytoplasmic antibody-associated vasculitis, characterised by necrotising inflammation of small- and medium-sized vessels. It typically involves the upper and lower respiratory tract and kidneys; however, virtually any organ may be affected. According to the 2022 European Alliance of Associations for Rheumatology (EULAR) recommendations, remission induction consists of glucocorticosteroids combined with rituximab or cyclophosphamide.
The study aims to evaluate the effectiveness of pharmacological treatment of solid inflammatory lesions located at atypical sites in patients with GPA.

Material and methods: Four hospitalised patients with GPA were included in this analysis. The diagnosis of GPA was established according to current EULAR classification criteria. The inclusion criterion was the presence of solid inflammatory lesions at atypical sites, despite typical respiratory tract involvement in GPA. Atypical lesions were located in the orbit, posterior mediastinum, and prostate and presented as a frontal sinus fistula. All patients received standard-of-care therapy in accordance with current guidelines, including rituximab (3 patients) or cyclophosphamide (1 patient). Treatment response was assessed at baseline and after 6 months based on inflammatory laboratory parameters, the Birmingham Vasculitis Activity Score (BVAS 3.0), and radiological evaluation of the lesions using computed tomography (CT).

Results: After 6 months of treatment, normalisation of C-reactive protein and decrease in disease activity within all BVAS 3.0 domains were observed (Table I). In all patients, we have observed a resolution of inflammatory changes in the respiratory tract and lungs in CT scans. Despite reaching clinical remission in all 4 cases, no radiological regression of atypically located lesions was noted, and all patients continued to present signs of local organ compression.

Discussion: All patients presented with typical involvement of the upper respiratory tract, and three also had lower respiratory tract involvement. The treatment outcomes observed in the studied patients are consistent with previous reports, indicating that solid lesions in GPA often demonstrate limited or no response to standard immunosuppressive therapy. Consequently, surgical intervention is currently considered in selected cases.

Conclusions: The heterogeneous localisation and treatment response observed among patients may reflect differences in underlying signaling pathways and pathogenetic mechanisms, warranting further research.

Table I. Characteristics and treatment of patients with GPA

Variable	Number			
	1	2	3	4
Age	65	39	59	48
Sex	Male	Female	Male	Male
CRP Before After	 66 < 1	 70 11	 146 2	 4 1
BVAS 3.0 Before After	 13 2	 16 1	 N/A 1	 8 1
Organ involvement	URT, LRT, CNS	URT, LRT, J, GIT	URT, LRT, REN	URT
Localisation of atypical lesions	Orbit	Posterior mediastinum	Prostate	Frontal sinus fistula
Atypical lesions response	No regression	No regression	No regression	No regression
Treatment	RTX	RTX	CYC	RTX

BVAS – Birmingham Vasculitis Activity Tract, CRP – C-reactive protein, CNS – central nervous system, CYC – cyclophosphamide, J – joints, GIT – gastrointestinal tract, LRT – lower respiratory tract, REN – renal involvement, RTX – rituximab, URT – upper respiratory tract.