

# NON-CLASSICAL ORGAN INVOLVEMENT IN GIANT CELL ARTERITIS

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## BACKGROUND

The increasing use of imaging techniques, particularly 18F-FDG PET-CT, has expanded our understanding of giant cell arteritis (GCA). It is now recognized as more than a cranial disease, exhibiting a broader and more heterogeneous clinical spectrum than previously thought. There is growing evidence that a significant number of patients exhibit uncommon manifestations of the disease.

## OBJECTIVE

To examine the frequency and types of non-classical organ involvement in patients with GCA.

## METHODS

This retrospective study analyzed a consecutive cohort of 148 patients with proven GCA diagnosed between 2005 and 2023. The diagnosis was established either by histology (temporal artery biopsy) or imaging (CDUS of the temporal arteries and/or evidence of large vessel vasculitis on 18F-FDG PET-CT, the latter performed in 142 cases). A causal link to GCA was established when manifestations were identified at diagnosis or within six weeks after initiating glucocorticoid treatment.

## RESULTS

The main clinical features and laboratory data of GCA patients are summarized in Table 1. Among the studied group, 31.1% (46/148) exhibited one or more non-classical complications previously described in the literature. The findings included:

- **Cardiac involvement** in 8.1% (12/148), with 1 case of ischemic heart disease and 11 (7.4%) of pericardial disease. Clinical pericarditis was diagnosed in 5 patients (3.3%), with 2 having pure pericarditis and 3 with pleuropericarditis. Aortitis was concurrently detected on PET-CT in these 5 patients, with 2 having a thoracic aortic aneurysm. Pericardial effusion was observed in another 6 patients, 4 of whom also had pleural effusion. These findings, confirmed by echocardiograms, were not linked to conditions like cardiac insufficiency or hypoalbuminemia.
- **Respiratory symptoms** in 14.8% (22/148), including odynophagia (3.3%, one with dysphonia), dry cough (1.3%), pleural disease (7.7%), and interstitial lung disease (1.3%). Four patients (2.7%) presented pulmonary nodules/micronodules at diagnosis, resolving completely with corticosteroid therapy, indicative of an inflammatory origin. PET-CT scans showed pulmonary artery involvement in 2 patients (1.5%), both with concurrent PMR findings.
- **Neurological involvement** excluding strokes, were present in 2.9% of cases (4/148). These included 2 cases of peripheral neuropathy (sural nerve biopsy showing demyelinating neuropathy and axonal degeneration), 1 case of epilepsy, and 1 case of acute spinal cord infarction (from D10-D11 to the conus medullaris, with negative anti-phospholipid antibodies).
- **Renal involvement:** vasculitis of the renal arteries on PET-TC was observed in 3.4% of patients (5/148).
- **Secondary amyloidosis** in 1.3% (2/148)
- **Gastrointestinal involvement** in 6.1% (9/148), including 7 cases of vasculitis of the mesenteric artery and/or celiac trunk on PET-TC, 1 case of mesenteric panniculitis, and 1 case of severely deranged liver function tests indicative of cholestatic hepatitis.
- **Salivary gland involvement** (parotid and/or submandibular) detected via PET-CT in 5.9% (8/148).

Other uncommon manifestations include scalp, tongue or lip involvement/necrosis in 2.7% (4/148) and non-PMR osteoarticular manifestations in 4% (6/148), including 3 cases of seronegative polyarthritis and 3 cases of remitting distal extremity swelling with pitting edema.

Diffuse hypermetabolism of the axial and proximal appendicular skeleton of mild to moderate degree was observed in 17.6% of patients (26/148) on <sup>18</sup>F-FDG PET-CT.

Polyautoimmunity was identified in 12.1% of cases (18/148), encompassing cases of SLE (1), anti-phospholipid positivity (3), collagenous colitis (1), pyoderma gangrenosum (1), axial spondyloarthropathy (1), atrophic gastritis (3) and thyroid disease (8).

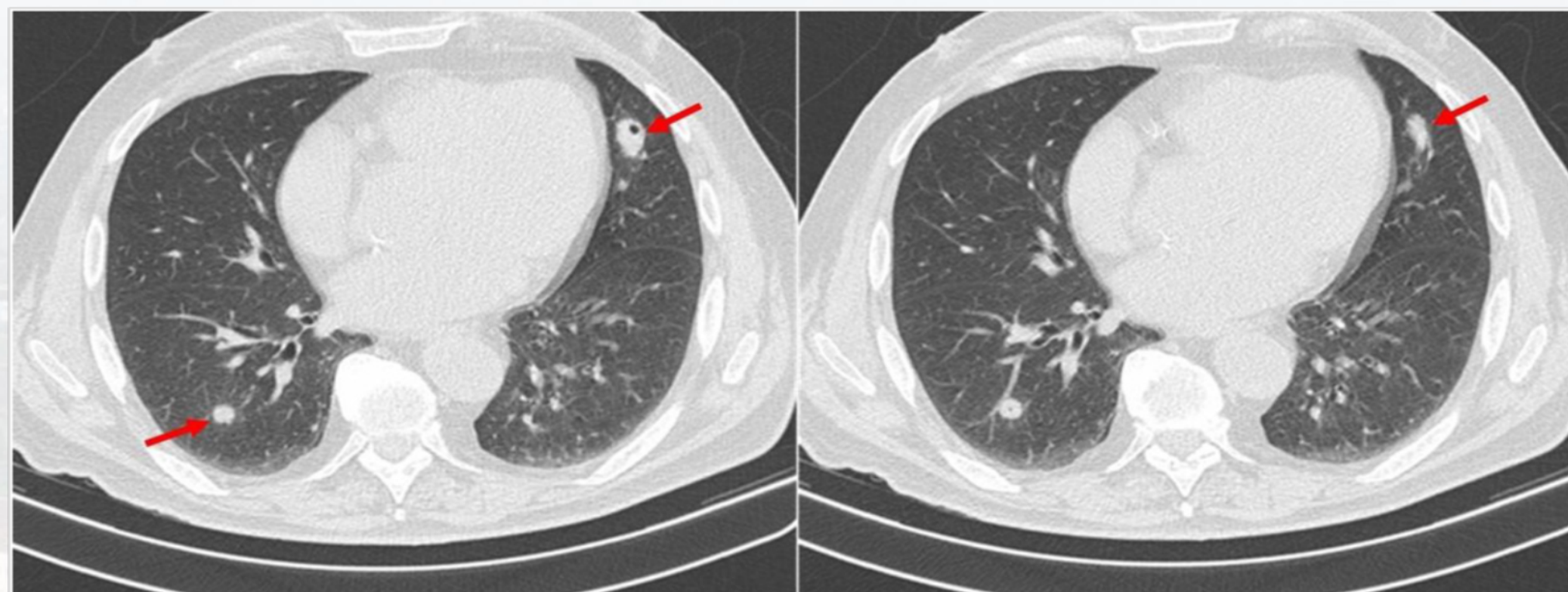


Figure 1 Chest HRCT showing multiple pulmonary nodules (arrows) some of them with small cavitation areas, before corticosteroid treatment.

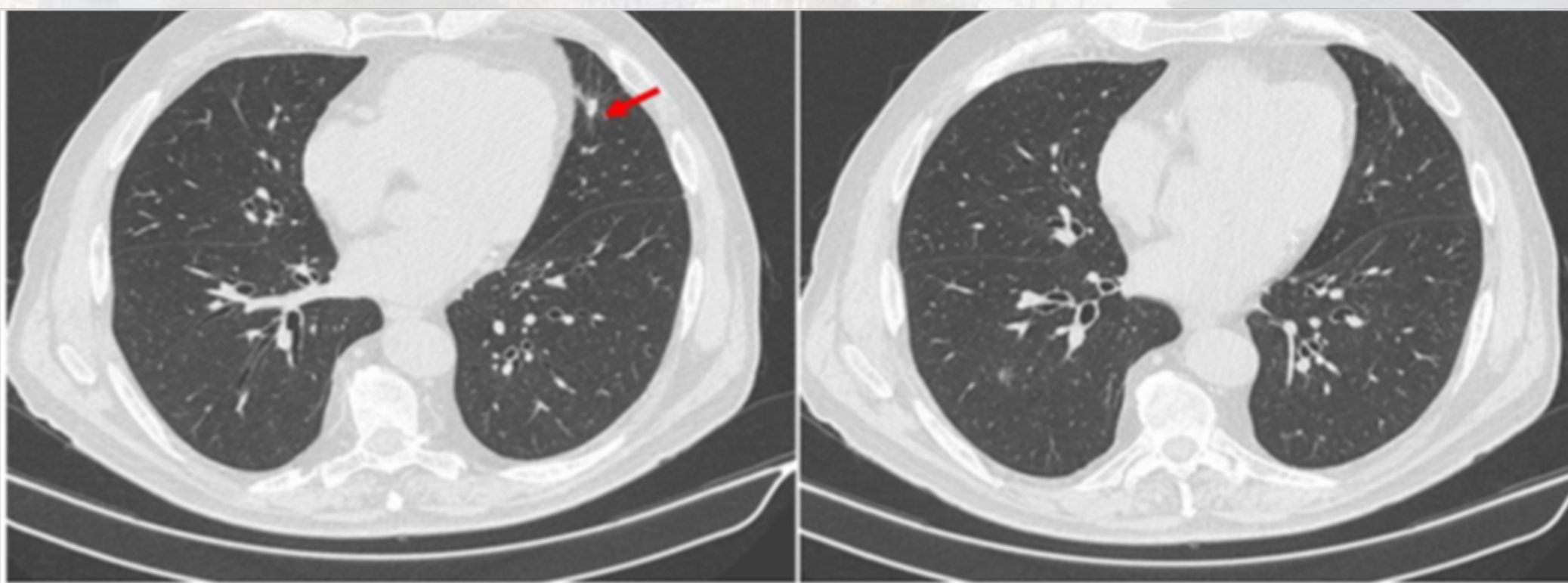
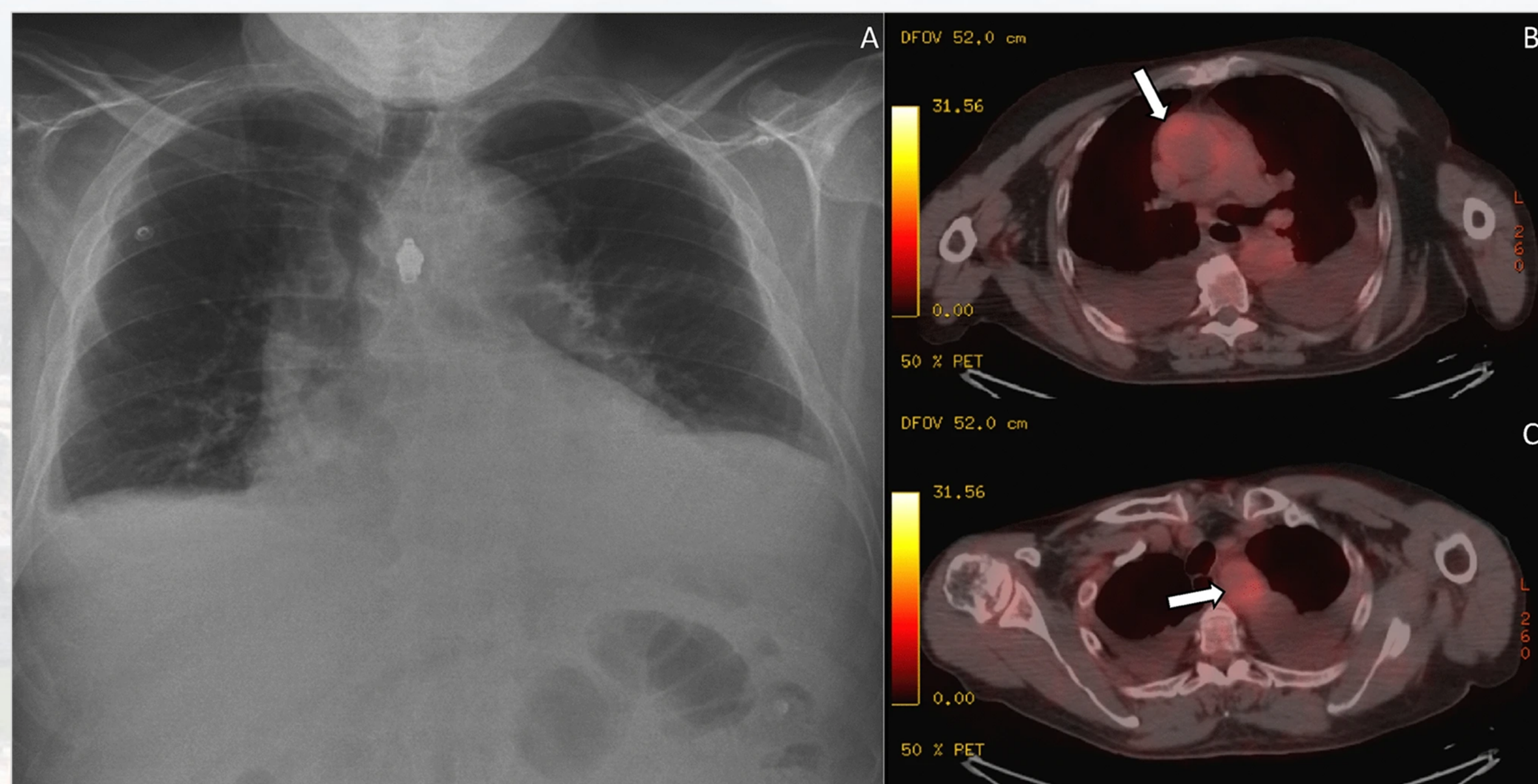
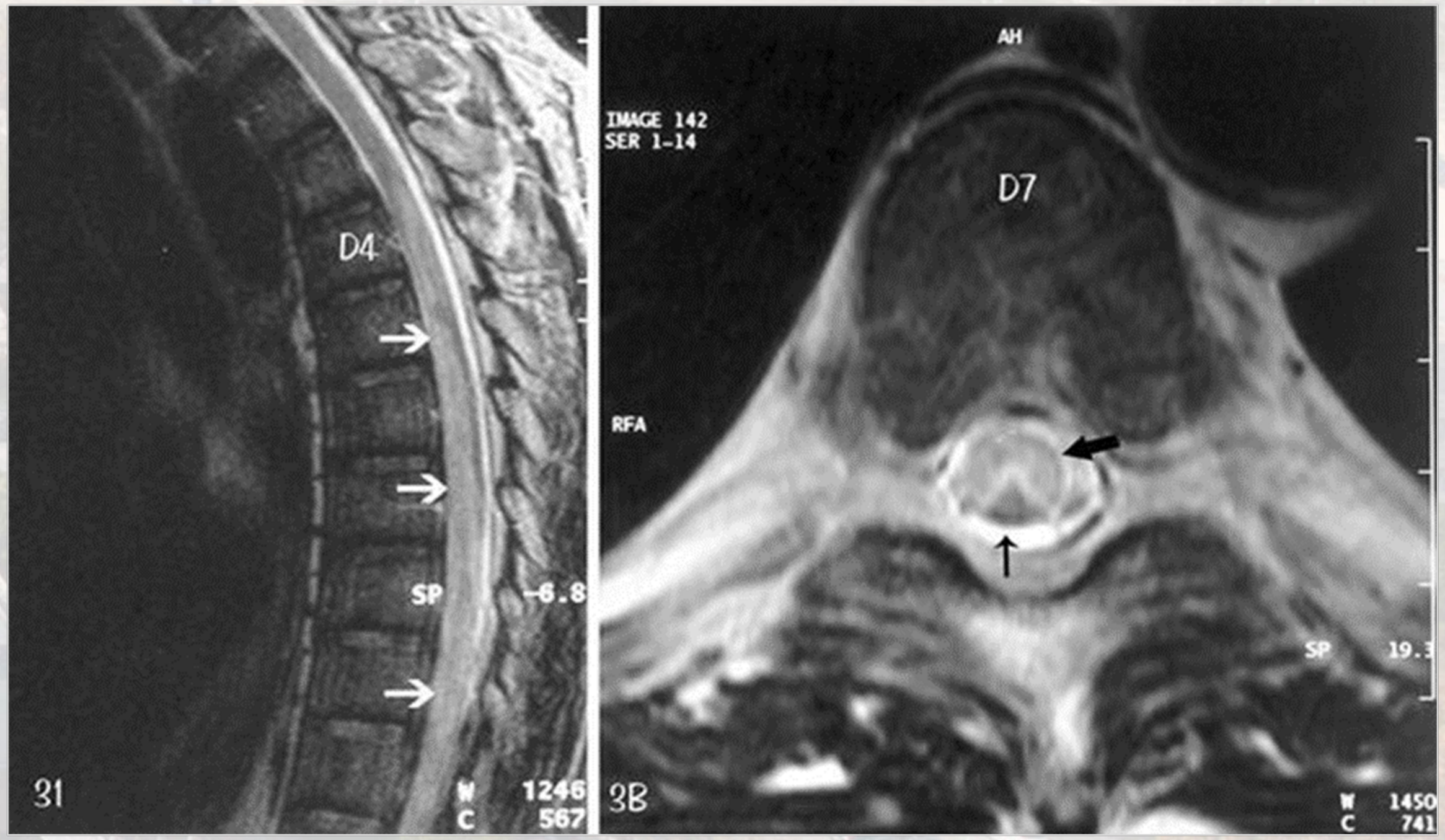


Figure 2. Chest HRCT post treatment: resolution of the nodular lesions and marked fibrotic reaction around the healing residual lesions (red arrow) is evidence of a favourable response to treatment



A Bilateral pleural effusions and increased cardiothoracic index in chest X-ray. Increased FDG activity in the ascending aorta (B) and aortic arch (C) in PET-CT



A T2 weighted sagittal MRI showing high signal abnormality involving the anterior portion of dorsal cord (white arrows) from D4 level and down word, B T2 weighted axial MRI at the level of D7 showing high signal abnormality involving the ventral two thirds (large black arrow) and sparing the posterior third (small black arrow) of the dorsal cord

Table 1. Main clinical and laboratory data of the study cohort.

Number of patients	148
Age (mean $\pm$ SD)	75 $\pm$ 8
Women/men (ratio)	96 (65%) / 52 (35%)
<b>Clinical features</b>	
Headache	93 (63%)
Temporal artery abnormality or scalp tenderness	52 (35%)
Jaw claudication	40 (27%)
Visual manifestations	47 (32%)
Cerebrovascular accidents	12 (8%)
Systemic symptoms*	111 (75%)
Polymyalgia rheumatica	61 (41%)
Limb claudication	5 (3.4%)
<b>Laboratory data</b>	
ESR (mm/h)	68 $\pm$ 29
CRP (mg/L; Ref. value $\leq$ 5)	73 $\pm$ 62
Anemia ( $\leq$ 11 g/dl)	81
Platelets ( $\times 10^9$ cells/mm <sup>3</sup> )	351 $\pm$ 127
Raised ALT/AST	9
Raised alkaline phosphatase*	37
Positive temporal artery biopsy, n= 97	56 (58%)
Positive temporal artery CDUS, n=65	33 (51%)
Positive 18 FDG-PET/CT, n=143	111 (78%)

## CONCLUSION

GCA usually presents with the classic cranial ischemic manifestations, fever and constitutional symptoms, and clinical features related to the extracranial large-vessel (LV) involvement. In addition to this, there has been an increasing knowledge of the occurrence of uncommon or exceptional complications in a significant proportion of patients that could be the presenting symptoms of GCA. These should be considered in the evaluation and follow-up of patients. Early recognition and prompt management are crucial to significantly reduce morbidity and mortality.

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