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Granulomatosis with polyangiitis

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Case description

A 30 year old male presented to the emergency department with a 6 month history of hemoptysis and epistaxis. He was otherwise healthy with no known medical conditions, no prior surgery, and was taking no medications. His vital signs were normal and his oxygen saturation was 99% SaO, on room air. On physical examination he had faint rales within the right upper lobe. Computed tomography scans of the maxillofacial bones and thorax were performed showing perforation of the nasal septum and a right upper lobe cavitary pulmonary lesion.

On laboratory investigations, his leucocyte level was 13.3x10⁹/L (normal 4-11), neutrophils 9.9x10⁹ (2-7.5) monocytes 1.06x109 (0.2-0.8) with a normal lymphocyte level. There was also an elevated ESR of 10 mm/hr (1-7) with a normal CRP. He had a positive serum C-ANCA. The patient was subsequently diagnosed with Granulomatosis with Polyangiitis and started on treatment with cyclophosphamide and glucocorticoids.

Discussion

Granulomatosis with Polyangiitis (GPA) is an autoimmune medium and small-vessel vasculitis characterised by necrosis and granulomatous inflammation [1]. It anti-neutrophil cytoplasmic antibody (ANCA)-associated which involves both the kidneys respiratory tracts [2]. It is believed to be autoimmune in origin and triggered by environmental events. It predominantly affects Caucasians but occurs in all racial groups [1]. The mean age at diagnosis is 40 but affects a wide age range (8-99 years) [1]. Diagnosis for GPA is based on clinical that is suggestive of vasculitis, positive c-ANCA serology and histological evidence of necrotizing vasculitis, granulomatous inflammation from a biopsy of kidneys, skin or lung, or necrotizing glomerulonephritis [1]. Due to the predominance of thoracic involvement, chest radiology findings frequently suggest diagnosis first. For diagnosis and surveillance of the disease, CT is the choice imaging modality [3]. Findings on chest CT include consolidation, ground-glass opacities, cavitations, masses and nodules [3]. The standard treatment of GPA is a combination of glucocorticoids and cyclophosphamide.

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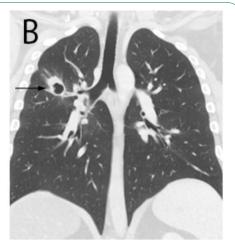


Figure 1: A) Axial Computed Tomography (bone algorithm) of the maxillofacial bones demonstrating perforation of the nasal septum. **B)** Coronal Computed Tomography (lung window) of the thorax showing right upper lobe thick-walled cavitary lesion.

Ethical approval: Written informed consent was obtained from the patient described in this article.

The manuscript has been read and approved by all the authors, and the requirements for authorship in this document have been met, and that each author believes that the manuscript represents honest work.

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