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Distinguishing Wegener's Granulomatosis From Necrotizing Community Acquired Pneumonia: A Case Report and Comparison of Radiographic Findings

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Summary

Wegener's granulomatosis (WG) is a necrotizing granulomatous vasculitis, affecting medium to small vessels in the respiratory and renal vasculature. Patients with WG may present with clinical and radiographic features similar to community-acquired pneumonia (CAP), which may delay life-saving immunosuppressive therapy. We report a 14-year-old female originally diagnosed with recalcitrant, necrotizing CAP complicated by massive pulmonary cavitations eventually proven to be WG. We also compare the radiographic features of WG and necrotizing CAP.

Keywords

Wegener's granulomatosis; pneumonia; computed tomography

INTRODUCTION

Wegener's granulomatosis (WG) is a rare necrotizing vasculitis affecting the renal and respiratory vasculature. Pulmonary radiographic findings in WG, such as infiltrates, nodules, and necrosis with subsequent cavitation, may be mistaken for other parenchymal processes more common in childhood like complications from community-acquired pneumonia (CAP).^{1,2} We present the case of a 14-year-old female patient who presented with findings consistent with complicated CAP but was eventually proven to have WG.

CASE REPORT

A healthy, 14-year-old Caucasian female was admitted after experiencing 3 weeks of fever, cough, and hemoptysis. She failed to improve on outpatient antibiotics and computed tomography (CT) of the chest revealed cavitory disease in the left upper lobe. Her presumptive diagnosis was necrotizing CAP and she was started on broad-spectrum antibiotics. Respiratory cultures, blood cultures, and sputum for acid-fast bacilli were negative. HIV testing and PPD were negative. A subsequent chest CT on hospital day 14 demonstrated progression of disease and prompted placement of a thoracostomy tube.

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Gram-stain and cultures of the pleural fluid were negative and she remained on broad-spectrum antibiotics. The patient improved over 2 weeks and was eventually discharged. Her fevers and hemoptysis returned 3 weeks after discontinuing antibiotics. A chest CT revealed new subpleural nodules and cavitary lesions in both lungs (Fig. 1). Extensive testing for infectious etiologies was negative. An open lung biopsy demonstrated necrotizing granulomas of the small arterioles throughout the lung parenchyma. Additional history revealed nasal congestion and epistaxis starting several months prior to the onset of her pulmonary symptoms. There was no saddle nose deformity, arthritis, or skin rash. Her laboratory examination was significant for leukocytosis (17.3×10^9 cells/L; reference: $5\text{--}15 \times 10^9$ cells/L) with 79% neutrophils (reference: 31%–61%) and thrombocytosis (614×10^9 cells/L; reference: $156\text{--}369 \times 10^9$ cells/L). Her hemoglobin and hematocrit were both decreased at 11.9 g/dl (reference: 12–16 g/dl) and 35.8% (reference: 36%–46%), respectively. Red blood cell indices were normal. Erythrocyte sedimentation rate (Westergren method) was 51 mm/hr (reference: 0–13 mm/hr) and C-reactive protein 1.11 mg/dl (reference: <0.6 mg/dl). Urinalysis was negative for blood and protein. Her anti-neutrophil cytoplasmic antibody titer was 1:640 (reference: <1:40) with cytoplasmic staining, positive anti-proteinase 3 (45 U/ml; reference: <6 U/ml) and negative anti-myeloperoxidase antibodies. Imaging of the head and neck revealed no sinus or tracheal disease. Ophthalmologic examination was normal. She was diagnosed with Wegener's granulomatosis and treated with 6 months of intravenous pulse cyclophosphamide and 8 months of oral prednisolone. Her lung disease stabilized with radiographic and functional improvement and no renal disease has developed.

DISCUSSION

Differentiating Wegener's from necrotizing CAP is challenging. As in this patient, the constitutional and pulmonary signs and symptoms observed in complicated CAP are also frequently observed in WG.^{1,2} Abnormalities on chest CT are found in patients with both conditions although important differences may be observed (Table 1). The patient in this report was unusual in that she presented with a large, solitary cavitary lesion sans nodular changes whereas most cavitary lesions in WG typically occur in the presence of nodular disease and are multifocal.^{3–5} Cavitary changes may be occasionally found in complicated CAP.^{6–9} In addition, cavitary necrosis in CAP is regularly accompanied by pleural effusions while pleural effusions are uncommon in pediatric WG and should be indicative of another process.^{3–9} Perhaps the most important clinical observation that can be made to differentiate WG from complicated CAP is that almost all patients with necrotizing pneumonia respond to aggressive medical and surgical therapy.¹⁰

CONCLUSION

WG may initially present with signs and symptoms similar to CAP. Chest CT may be useful in differentiating WG from necrotizing CAP. WG should be considered in all cases of refractory necrotizing CAP.

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Fig. 1. Massive cavitory lesion in Wegener's granulomatosis. High-resolution chest CT demonstrates large cavitory lesion (C) in left upper volume. Parenchymal and classic subpleural nodules (N) are evident throughout the right lung.

TABLE 1

Frequency of Computed Tomography Findings in Wegener's Granulomatosis (WG) and Necrotizing Community Acquired Pneumonia (CAP)

	Wegener's granulomatosis (%)¹	Necrotizing CAP (%)²	Fisher exact <i>P</i>-value
Nodules	91/101 (90)	2/11 (18)	<0.001
Cavitation	42/101 (42)	42/66 (64)	0.007
Airspace consolidation or infiltrate	34/101 (34)	66/66 (100)	<0.001
Pleural effusion	15/101 (15)	52/66 (79)	<0.001
Hilar or mediastinal lymphadenopathy	18/101 (18)	3/11 (27)	0.43

¹Data abstracted from references 3–5.

²Data abstracted from references 6–9.