C-ANCA Associated Vasculitis. Pathway to Diagnosis

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Abstract

Introduction: C-ANCA-associated vasculitis is a rare autoimmune condition primarily affecting the respiratory and renal systems. It often presents with nonspecific symptoms such as fever, fatigue, and respiratory distress, making early diagnosis challenging. Timely detection and treatment are crucial to preventing severe organ damage.

Case Report Description: A 21-year-old female presented with complaints of weakness, a runny nose, headaches, a mild cough, and fever. Symptomatic therapy was initiated. Two weeks after symptom onset, her condition worsened. She reported coughing up blood, and bullous skin lesions appeared on her lower legs. Suspecting atypical pneumonia, she was hospitalized at Liepaja Regional Hospital. A chest CT scan revealed bilateral pneumonia and suspected intrapulmonary hemorrhage. Serologic testing showed C-ANCA levels > 200 U/L, and creatinine was 91 µmol/L. Treatment was initiated with Solu-Medrol, and she was transferred to Pauls Stradiņš Clinical University Hospital for further evaluation and therapy. Upon admission, she continued coughing up blood and was hypoxic, requiring oxygen support. A multidisciplinary consultation was held, leading to pathogenetic therapy with Solu-Medrol and Rituximab. A kidney biopsy revealed crescentic and necrotizing glomerulonephritis. During hospitalization, she had recurrent nosebleeds, and a sinus CT scan revealed nasal septum mucosal ulceration. Her kidney function worsened, with creatinine peaking at 250 µmol/L, requiring continued pathogenetic therapy. Follow-up chest CT showed positive dynamics. C-ANCA levels decreased to 141 IU/mL. Her kidney function improved, and she was discharged on Medrol, antihypertensives, Omeprazole, Biseptol, and Aranesp. For six months post-discharge, she had monthly nephrology follow-ups, later spaced to 3–6 months with glucocorticoid tapering. She completed induction therapy with Rituximab (700 mg × two doses) and maintenance therapy (500 mg × three doses). The latest C-ANCA level is 13.2 IU/mL. At present, she has no symptoms of vasculitis exacerbation, and no additional vasculitis therapy is required. Nephrology follow-ups are currently planned every 8–10 months, and she receives symptomatic therapy as needed.

Conclusion: This case highlights the importance of early recognition and treatment of C-ANCA vasculitis in young patients with nonspecific symptoms. Early diagnosis and targeted therapy are vital for achieving remission and improving prognosis. This case serves as a reminder to consider rare conditions like C-ANCA vasculitis in differential diagnoses, especially when symptoms persist despite initial treatment.

Keywords

C-ANCA-associated vasculitis; Multisystem autoimmune disease; Glomerulonephritis; Intrapulmonary hemorrhage.