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Serum C-reactive protein reflects the proportions of cellular crescent formation and glomerulosclerosis in patients with microscopic polyangiitis

1. Iwakiri1, Y. Matsuura1, A. Yamashita1, S. Uezono2, S. Hisanaga3, T. Tokura4, K. Kitamura5, Y. Asada1, S. Fujimoto6 with MPA may be a good indicator for conduct of aggressive treatment. Cardiac involvement is also associated with crescentic glomeruli. Thus, higher serum CRP in patients with MPA. Elevated serum CRP at diagnosis indicated higher histopathological features in the biopsied kidney specimens. Also we evaluated the renal outcome (eGFR at diagnosis and 1 year, or ESRD during follow up) between H and L group. Results. – The median proportions of normal, cellular crescentic and sclerotic glomeruli are 42.9, 28.6 and 11.1%, respectively. Serum CRP at diagnosis showed positive correlation with the proportion of cellular crescentic glomeruli (R = 0.28, P < 0.01) and negative correlation with the rate of sclerotic glomeruli (R = −0.49, P < 0.0001). eGFR at 1 year in H group were significantly higher than those in L group (38.5 vs. 25.5 ml/min/1.73 m², P < 0.05). In the Kaplan-Meier analysis, H group showed pauciimmune necrotizing glomerulonephritis with global sclerosis in 90% of glomeruli. CYC was given monthly (400 mg/pulse, 6 × ), steroids were tapered and she is currently in remission with maintenance treatment (AZA 50 mg qd) on kidney transplant list. Discussion. – The case is striking as a mild isolated laryngeal manifestation preceded full-blown severe MPA for 11 months. No other causes were found for the vocal cord disease. MPO-ANCA were positive from the start, raising the question of labelling the disease as MPA with an unusual ENT symptom, mostly seen in GPA. Conclusion. – Laryngeal symptoms can be the initial manifestation of MPO-ANCA systemic vasculitides.

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Unilateral vocal cord paralysis as initial manifestation of severe microscopic polyangiitis (MPA)

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Introduction. – To our knowledge, vocal cord paralysis has not been reported in microscopic polyangiitis (MPA). We present a case that progressed to severe disease with rapidly progressive glomerulonephritis, requiring hemodialysis after 11 months of sole laryngeal involvement.

Methods. – Case description.

Results. – 16-years-old female with dry cough and dyspnea since February 2011. Treated elsewhere with suspicion of infection, laryngopharyngeal reflux and allergy (with steroids) without improvement. In our centre, fibronasolaryngoscopy showed vocal cord edema and incomplete left vocal cord abduction. Flexible bronchoscopy: normal below the subglottis. In September, she was seen in the Vasculitides Clinic as ANA were 1:320 (coarse speckled), P-ANCA 1:20, MPO-ANCA 128 U/mL (normal < 128), negative PR3-ANCA, CRP 5.88 mg/dL (normal < 0.8). Other organs, laboratory tests and a brain CT scan were normal. She was labelled as “MPO-ANCA positive vocal cord paralysis” deserving regular follow-up. In January 2012, she consulted for one-week vormit and distal legs edema; besides, she was only pale and normotensive. Serum creatinine-10.6 mg/dL (3 weeks before, 0.7), urine sediment full with dysmorphic erythrocytes and granular casts. Treatment: methylprednisolone pulses, IV-CYC, hemodialysis and plasmapheresis, the latter stopped after five exchanges when renal biopsy showed pauciimmune necrotizing glomerulonephritis with global sclerosis in 90% of glomeruli. CYC was given monthly (400 mg/pulse, 6 × ), steroids were tapered and she is currently in remission with maintenance treatment (AZA 50 mg qd) on kidney transplant list.

Discussion. – The case is striking as a mild isolated laryngeal manifestation preceded full-blown severe MPA for 11 months. No other causes were found for the vocal cord disease. MPO-ANCA were positive from the start, raising the question of labelling the disease as MPA with an unusual ENT symptom, mostly seen in GPA.

Conclusion. – Laryngeal symptoms can be the initial manifestation of MPO-ANCA systemic vasculitides.

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Rapidly progressive glomerulonephritis is not the sole presentation of renal involvement in ANCA associated vasculitides

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Introduction. – Renal involvement is frequent in ANCA-associated vasculitis (AAV) and remains a strong prognostic factor. Although the typical presentation of renal AAV is a rapidly progressive glomerulonephritis (RPGN), other renal presentations have been described.

Methods. – We included 78 patients with biopsy proven ANCA-associated glomerulonephritis. RPGN was defined by the association of hematuria, proteinuria and doubling of baseline serum creatinine within the 3 months preceding admission. Non-RPGN patients were classified as Slowly Progressive Renal Failure (SPRF) or Normal Renal Function (NRF), according to the initial eGFR (MDRD < 60 or > 60 ml/min).

Results. – RPGN was diagnosed in 45 (55%) patients, SPRF in 17 (23%) and NRF in 16 (22%).