Re: Atypical Clinical Course of Antineutrophil Cytoplasmic autoantibodies-associated Vasculitis

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Dear Editor,

We read with great interest the recently published article by Tayebi-Khosroshahi and colleagues entitled "Atypical clinical course of antineutrophil cytoplasmic autoantibodies-associated vasculitis," in the esteemed *Iranian Journal of Kidney Diseases*. They reported a report 16-year-old boy who presented with some constitutional symptoms, purpura, additive arthritis, dysentery and rapidly progressive kidney failure. Patient also had retropharyngeal abscess. After appropriate evaluation consisting kidney biopsy and a positive cytoplasmic antineutrophil cytoplasmic antibodies (ANCA), the diagnosis of ANCA-associated vasculitis was confirmed, they concluded that, their case was the first patient in which the ANCA-associated vasculitis was presented with retropharyngeal abscess.[1] While the article had many educational notes, we would like to add a few points.

In 1936, Friedrich Wegener explained a granulomatous disease involving the upper and lower respiratory tract along with glomerulonephritis.[2] However recently, the term granulomatosis with polyangiitis has been widely accepted and established as an alternative name for Wegener granulomatosis.[3-7] Granulomatosis with polyangiitis is an autoimmune necrotizing vasculitis of small vessels which mainly affects the paranasal sinuses, lungs and kidneys. Although other rare, presentations has also been reported.[4,5,7] A short look to the recently published paper on the evaluation of 10-year experience of Wegener's granulomatosis in Iranian children, conducted by Tahghighi and colleagues on 11 patients (5 females and 6 males, with mean age of 11 years), found, the most common organ system involvement was upper and lower respiratory tract involvement (81.8% and 63.9%, respectively). Other common manifestations were renal (36.3), skin (27.2%), and eye involvement (18.2%). None of their patients, had presented with retropharyngeal abscess; thus, we agree with Tayebi-Khosroshahi and colleagues that they reported a rare presentation of granulomatosis with polyangiitis in a 16-year-old patient.[8] In the renal biopsy, they reported, of 9 glomeruli, 5 of which had cellular crescents and 1 was sclerotic. To better reach for a modality of the treatment, nowadays, it is necessary to apply the histopathologic classification of ANCA-associated glomerulonephritis, published in 2010. According to this classification, we can accommodate this patient in the crescentic class.[9] Also, it is necessary to report, the proportion of globally sclerotic glomeruli,[10-12] and the status of interstitial vessels of their patient too.[13-15] Indeed, ANCA-associated vasculitis is the most common cause of rapidly progressive glomerulonephritis around the world,[3,4,6,9] and it is evident that, this pathologic classification will aid in the prognostication of patients at the time of diagnosis and facilitate uniform reporting between centers.[9] This classification at some point might also provide means to guide therapy too. In this patient therefore, the class of crescentic ANCA-associated glomerulonephritis referred to highly active kidney disease and severely reduced kidney function but stand a good chance for kidney function recovery.[9] In fact, the proportion of active crescentic lesions, especially cellular crescents as in this case, is related to recovery of renal function independent of baseline renal function.[9-12] Contrariwise, the percentage of fibrous crescents adversely affects long-term renal outcome.[9,17] In this patient, successful treatment leaded to serum creatinine level reduction during the treatment and the patients was discharged with good clinical condition as also predicted by the class of this classification.

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