A unique presentation of small vessel vasculitis

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Background

Introduction:

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While ENT manifestations are common in several small vessel vasculitides (more common in GPA w/90% occurrence vs MPA w/~35%), there are no documented cases of uvular swelling as the presenting clinical finding of the disease. Far more common upper airway findings include sinusitis, otitis media, rhinorrhea, and in severe GPA cases, cartilage destruction. These cases also frequently accompanied by skin, lung and kidney complications, all of which were also present in this case.

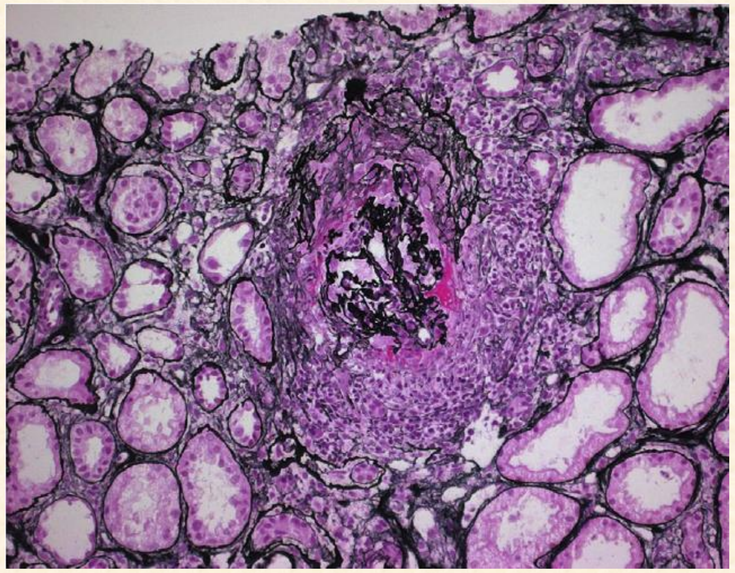
Case Report

Case presentation:
43-year-old man with non-contributory past medical history who presented to ED with a hoarse voice and difficulty swallowing over a five-day period. He also noted uvular swelling that progressed over the same time period. In addition, the patient developed a red, raised rash over his extremities and abdomen of unclear duration.

•Patient was hypertensive (150/100) and tachycardic (120) on arrival.
•Labs were notable for Cr 2.3 (baseline 1), D dimer 10,171, CRP 171 and urinalysis positive for blood.
•CT chest/abdomen/pelvis demonstrated destructive soft tissue thickening centered in the right maxillary sinus with extension into the ethmoidal sinus with a diffusely enlarged and edematous uvula, enlarged cervical chain lymph nodes, and persistent consolidative mass-like opacities in the bilateral upper lobes.
•At this time, the differential was broad and included infection, malignancy and autoimmune.

Clinical Data:

After discharge, he completed a renal biopsy which was consistent with PR3+ vasculitis. Fig 1 below is an example of ANCA, PR3+ vasculitis seen on Kidney biopsy



Case Continued

During his initial admission, extensive infectious workup was negative. Additional labs notable for positive c-anca and elevated proteinase 3. He underwent skin biopsy which demonstrated leukocytoclastic vasculitis. Lung biopsy with necrotic lung parenchyma, no granulomatous inflammation, no fungi, mycobacteria or definite bacteria seen on GMS, PAS-D, AFB and gram stain, however sample was too small to properly evaluate for vasculitis. Additionally, nasal debridement was performed by ENT. After discharge, he completed a renal biopsy which was consistent with PR3+ vasculitis.

Discussion

- Overall, what I took away from this case, and my further reading about vasculitis, is that there can be many different presenting signs and symptoms of vasculitis. While some constellations of symptoms are classic for certain vasculitides, patients often present with a less clear picture. Autoimmune causes should always be on the differential in patients with new multi organ disease, particularly when it comes to ENT, lung and kidneys. Early diagnosis and treatment is critical in these patients. This case exemplifies the wide array of clinical signs that can appear with c-Anca positive vasculitis.

Follow up

He was treated with steroids which ultimately led to improvement, unfortunately the patient's illness led to several readmissions due to complications of his vasculitis.

Selected References

1Hilhorst, Marc & Paassen, Pieter & Cohen Tervaert, Jan Willem. (2015). Proteinase 3-ANCA Vasculitis versus Myeloperoxidase-ANCA Vasculitis. Journal of the American Society of Nephrology : JASN. 26. 10.1681/ASN.2014090903.

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