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Clinical vignette: Subarachnoid hemorrhage the initial manifestation of granulomatosis with polyangiitis flare

Jairon Johnson

Katie Sanchez

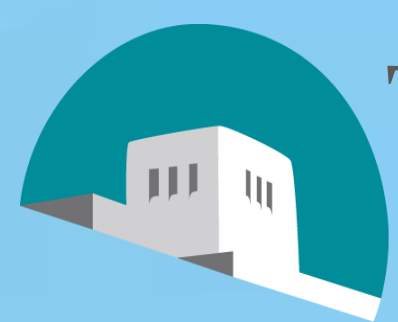
Jennifer Jernigan

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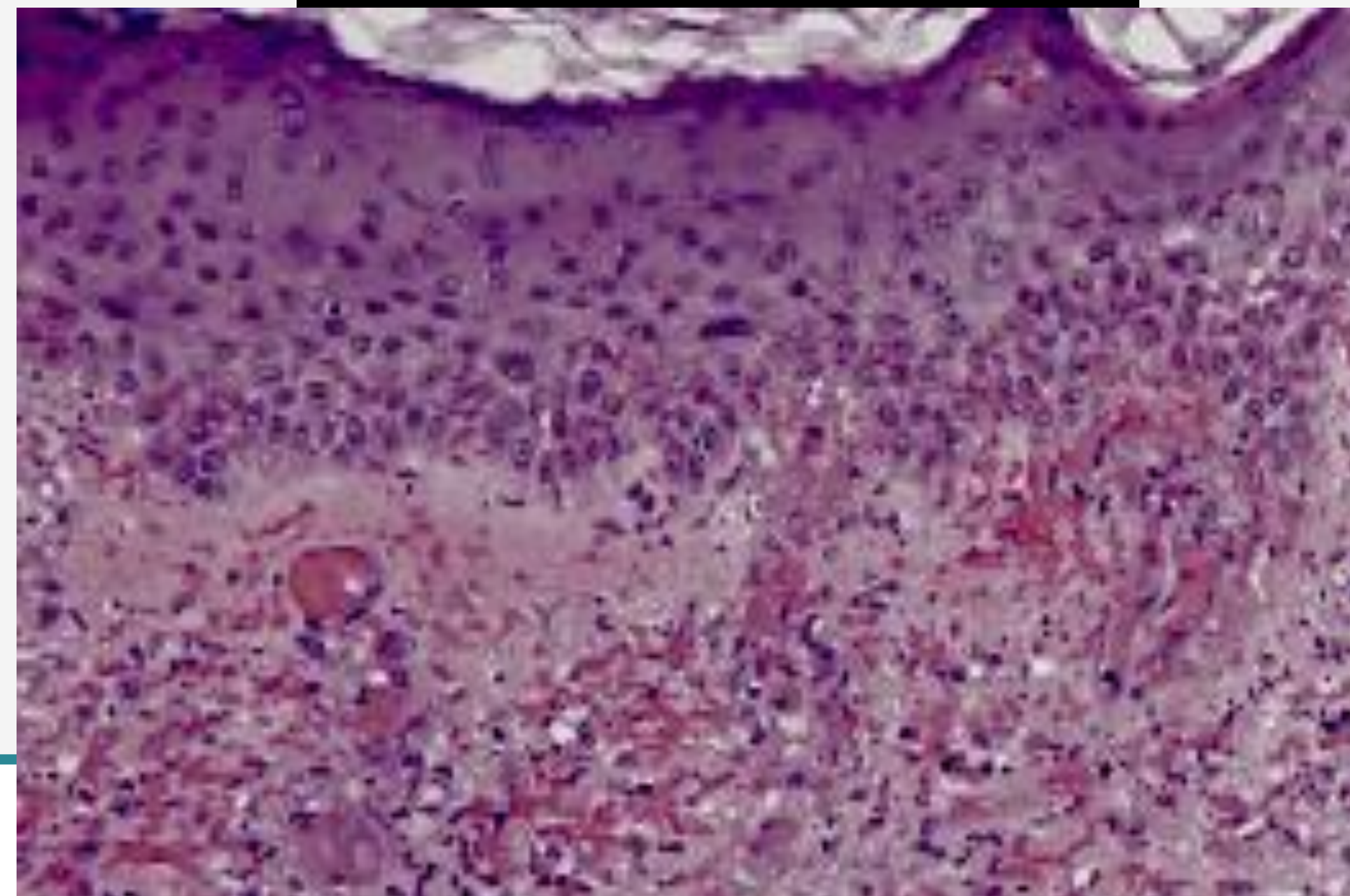
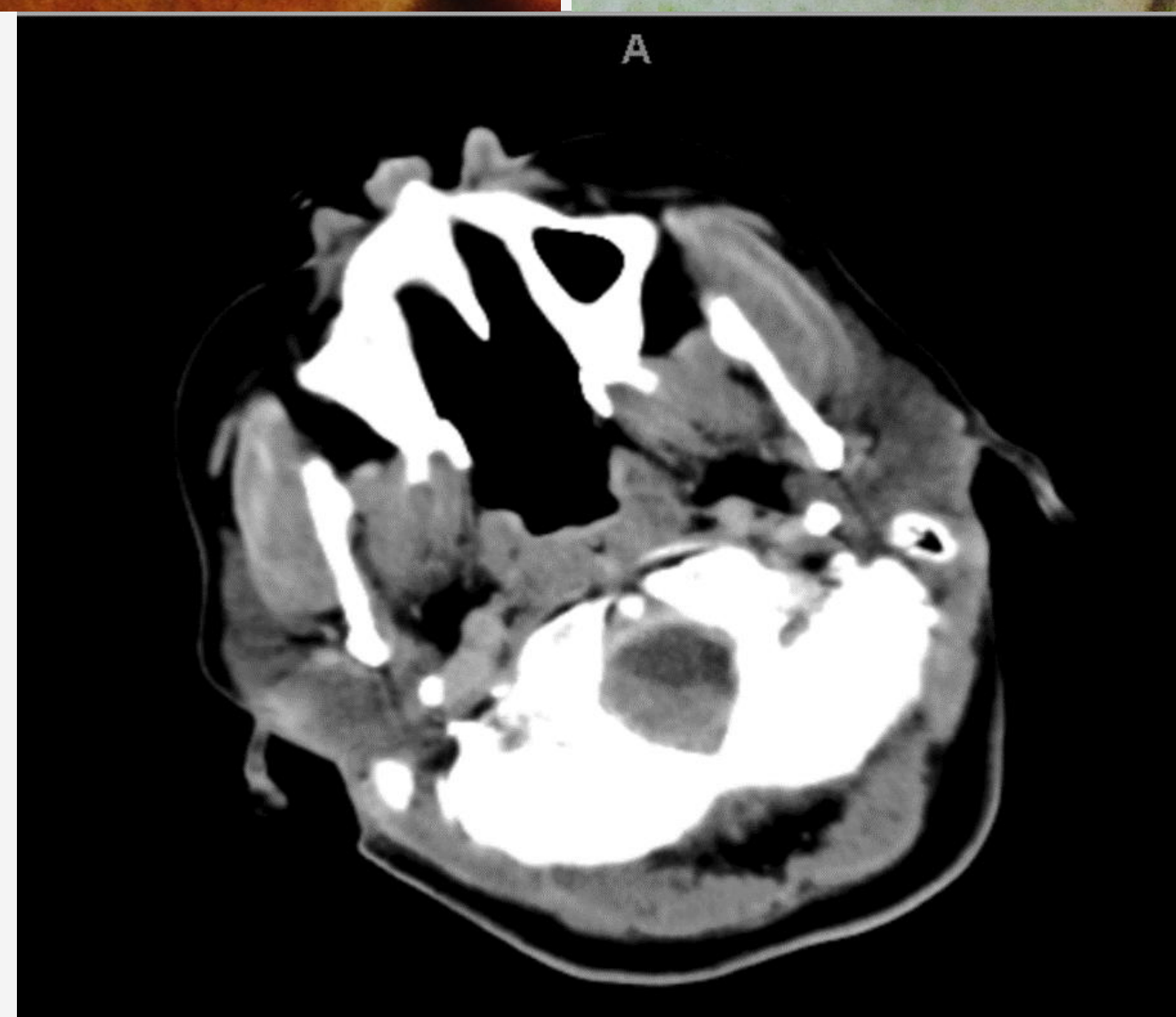


Subarachnoid Hemorrhage the Initial Manifestation of Granulomatosis with Polyangiitis Flare

Jairon Johnson, DO; Katie Sanchez; Jennifer Jernigan, MD
Albuquerque, NM

Introduction

Granulomatosis with polyangiitis (GPA), historically known as Wegner's granulomatosis, is an uncommon disease that is characterized by a constellation of findings including granulomatous inflammation of the respiratory tract, small and medium size vessel vasculitis, and focal segmental glomerulonephritis. Although these are the most common findings, all other organ systems may be affected. The nervous system is affected in 25- 50% of cases, primarily by peripheral neuritis, but less commonly involving the CNS. Very rarely, subarachnoid hemorrhage (SAH) has been reported, and is thought to result from vasculitis of the vessels compromising their integrity. Other manifestations include retinitis, myositis, cutaneous lesions, and constitutional symptoms. Here we report a rare case of chronic GPA, with past fulminant renal failure, who presented with subarachnoid hemorrhage and subsequently developed a cutaneous eruption after stopping immunosuppression for a failed kidney transplant.



Top: Cutaneous lesions erupting on the face and hand on hospital day 6.
Middle: CT image showing SAH in the foramen magnum.
Bottom: Histopathology Skin, Rt Hand (punch biopsy): Leukocytoclastic Vasculitis with Medium-Sized Vessel Involvement

Hospital Course

She was admitted to the NSICU and was treated for hypertensive urgency and had frequent neuro checks. She had severe headache, nausea and vomiting, but no progression of SAH on CT imaging and no need for surgical intervention.

On hospital day 5, she developed swelling and new skin lesions on her face. These lesions progressed to involve the extremities, face, and trunk; becoming ulcerated and painful. A rheumatologic work-up was completed and showed:

Labs:

ANCA screen: negative, ANA: negative
C3: 73 (normal 90-180), C4: 19 (normal 10-40)
PR3: negative, MPO: positive

Skin biopsy was performed and showed leukocytoclastic vasculitis

Presentation

History:

This is a 42 year old female with a past medical history of GPA resulting in renal failure, post renal transplant in 1999 with recent nephrectomy after rejection, hypertension, and hypothyroidism. She was diagnosed with GPA 15 years prior by renal biopsy and was ANCA +. She was treated with immunosuppression after the renal transplant, but had discontinued medications after having a nephrectomy 3 weeks prior to presentation.

She presented to an outside hospital with severe headache and confusion in the setting of systolic blood pressures >200. CT scan showed a subarachnoid hemorrhage involving the basal cisterns and filling the fourth ventricle.

Diagnostic angiogram on presentation to our facility was normal with no aneurysms, AVMs or other vascular abnormalities.

Discussion:

In patients with a history of GPA a subarachnoid hemorrhage can be the initial presentation of a vasculitis flare. In GPA 22-54% of patients can present with neurological involvement, most commonly mononeuritis multiplex and polyneuritis while CNS manifestation occurs in only 7-11% of patients. Of those, SAH or ICH are very uncommon. There have only been 8 reported cases of SAH associated with GPA in the literature and only 2 of those cases reported full recovery.

SAH in patients with GPA is clinically different than in the general population. All but 1 of the cases of SAH in patients with GPA were non-aneurysmal, while 85% of spontaneous SAH in the general population is caused by ruptured cerebral aneurysms. Another interesting difference is that SAH in the general population has a female preponderance, while only 1 of the 8 previously reported cases in GPA was female.

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