

Unusual Presentation of Anca Vasculitis with Otitis Media and CVA

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Abstract

A 42-year-old African female (Kenyan in origin), known to have hypertension that was diagnosed during gestation and persisted; presented with a history of dysphagia for four years. Before her admission, she had presented to the surgical clinic with ear fullness and ear pain with throbbing headaches, and she was diagnosed with bilateral oto-mastoiditis. Her symptoms subsequently, progressed to a multisystemic disease involving the lungs, brain, kidneys, and peripheral nerves.

ANCA vasculitis is a rare disease, the main target organs in granulomatous polyangiitis include the ear, nose throat, and upper respiratory tract, and glomerulonephritis of the kidney and the lungs. Localized disease manifestation can include the nose (rhinorrhea, nasal crusts, septal perforation) and ear (otitis media, hearing loss, sensorineural deficits). The diagnosis of vasculitis from localized disease manifestation can be difficult to diagnose.

Case Presentation

A 42-year-old African female (Kenyan in origin), known to have hypertension that was diagnosed during gestation and persisted; presented with a history of dysphagia, to solids, liquids, and semisolids for four years. Before her admission, she had presented to the surgical clinic with ear fullness and ear pain with throbbing headaches and was diagnosed to have bilateral oto-mastoiditis with a mastoid effusion for which she underwent mastoidectomy after recurrent episodes of ear infection. Cultures obtained grew corynebacterium urealyticum and staphylococcus aureus for which she received antibiotics sensitive to gentamicin, cotrimoxazole, ciprofloxacin, clindamycin, erythromycin, vancomycin and tetracycline, and multiple tests for tuberculosis had been negative including cultures. The HIV test was negative. A few months later she presented with trigeminal neuralgia and received prednisone.

Two weeks before her recent admission in 2019, she developed progressive bilateral lower limb numbness up to the calves. Two days before admission, she experienced sharp bilateral pains radiating to the buttocks, poorly responsive to analgesia. She also reported hoarseness of the voice.

On presentation, the blood pressure was 136/97mmHg, pulse 91/min, respiratory rate 18/min with a temperature of 36.6 degrees centigrade. Chest and abdominal examinations were unremarkable. Neurological examination revealed bilaterally distal lower limb length-dependent polyneuropathy (with altered sensation

to pain, light touch, and vibration sense).

She was thereafter noted to have right-sided hemiparesis, with brisk right bicep and knee-deep tendon reflexes. A repeat MRI done showed a 6mm acute right pontine infarct, and a demonstration of the oto-mastoiditis and left mastoid effusion (Figure 1, Figure 2). She was commenced on aspirin and clopidogrel and atorvastatin. A vasculitis screen was carried out and C-ANCA was positive; this entertained the diagnosis of Anti-neutrophilic associated vasculitis.

Her neuralgia was treated with analgesia, for the laryngeal infection she was started on high-dose cotrimoxazole, based on the need for later prophylaxis against opportunistic infections once treatment for anti-neutrophilic associated vasculitis was commenced, for which she received rituximab.

In 2020 she presented with tracheitis (figure 3, figure 4), and sputum cultures had been positive for Klebsiella oxytoca for which she was treated with antimicrobial therapy and continued her maintenance therapy of prednisone, mycophenolate sodium, cotrimoxazole prophylaxis, alendronate, and her antihypertensive medications. Unfortunately, she developed a massive upper gastrointestinal bleed secondary to multiple vasculitis gastric ulcers evidenced by an upper endoscopy. She kept having a persistent upper gastrointestinal bleed and given the vasculitis, she underwent plasma exchange. Over the next couple of days, she experienced persistent upper gastrointestinal hemorrhage for which she was transfused and a surgical opinion was sought.

However, surgical interventions could not be instituted given her hemodynamic instability. Her condition persistently deteriorated and she suffered a cardiopulmonary arrest for which she was

resuscitated as per the ACLS protocol. She was regrettably pronounced dead.

Imaging

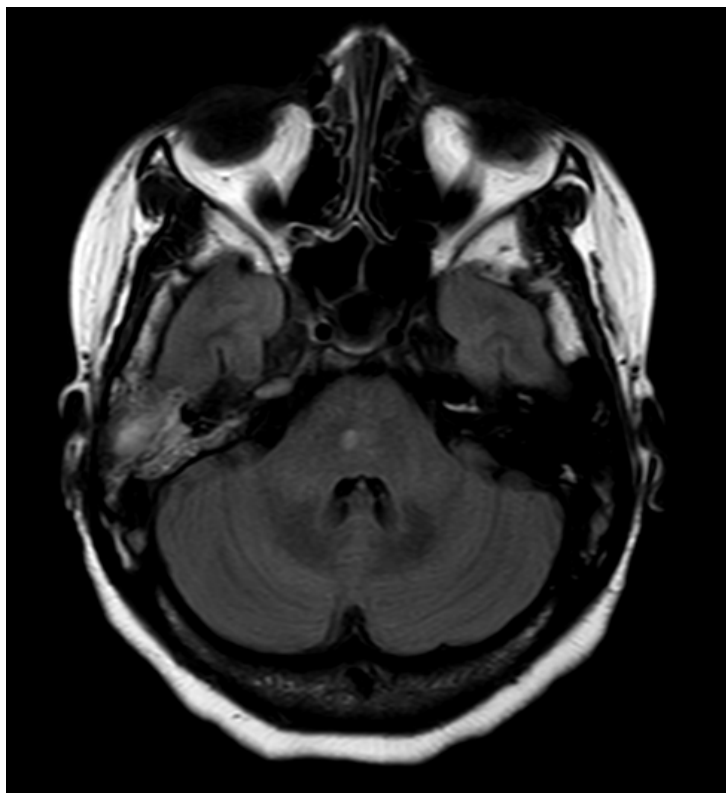


Figure 1: There is a 6mm Flair hyperintense focus with associated true restricted diffusion in the right hemi-pons as shown below:

Axial FLAIR demonstrating the 6mm hyperintense focus in the right hemi-pons. Also seen are hyperintense signals within the right mastoid air cells in keeping with otomastoiditis.

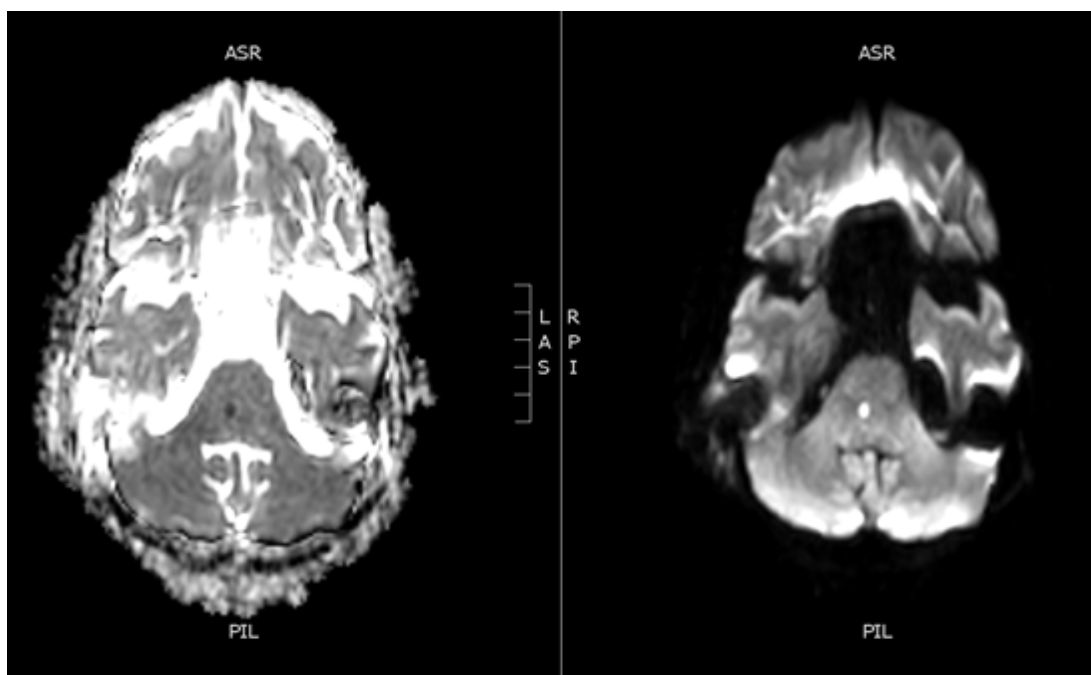


FIGURE 2: Axial DWI and ADC maps demonstrating the restricted diffusion in the right hemi-pons (dark on ADC and bright on DWI).



Figure 3: Sagittal contrast-enhanced CT of the neck demonstrates soft tissue thickening of the posterior wall of the upper trachea (inferior to the posterior cricoid), slightly eccentric to the left, and with a craniocaudal extent of approximately 12.2 mm as shown below:

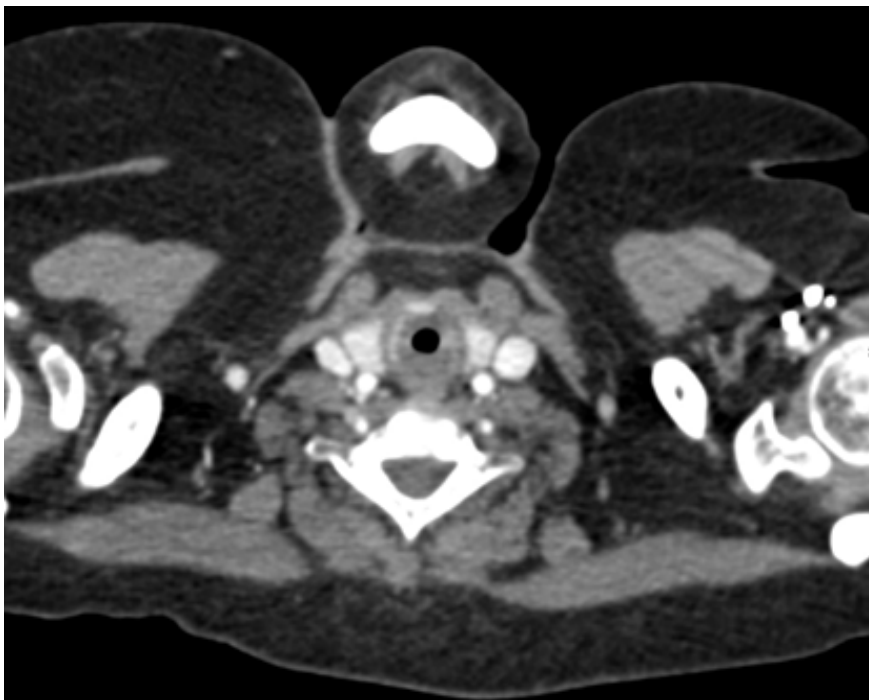


Figure 4: Axial images at the same level demonstrate the narrowing of the adjacent airway by approximately 50%. No invasions into the surrounding soft tissues or cartilaginous structures were seen. The rest of the neck spaces are intact. No enlarged or abnormal neck nodes.

Discussion

ANCA vasculitis is a rare disease characterized by antibodies against neutrophil and monocyte lysosomal enzymes which include myeloperoxidase and proteinase 3 [1]. The pathogenesis for ANCA vasculitis involves numerous immune cells; B lymphocytes produce ANCA, with BAFF encouraging the autoimmunity with B cells, plasmablasts, and plasma cells [2]. These activate neutrophils and result in the generation of C5a via the complete pathway, causing further activation of neutrophils [3]. Activated neutrophils result in the propagation of tissue damage and vascular inflammation.

The main target organs in granulomatous polyangiitis include the ear, nose, throat, and upper respiratory tract, and glomerulonephritis of the kidney, and the lungs. Localized disease manifestation can include the nose (rhinorrhea, nasal crusts, septal perforation) and ear (otitis media, hearing loss, sensorineural deficits). The diagnosis of vasculitis from localized disease manifestation can be difficult [1].

Otological manifestations as seen in our patient as otitis media is very uncommon as an initial presentation. Otitis media is caused by a granulomatous process that occurs in the middle ear and can also involve the mastoid cavity [4]. The otological symptoms occur insidiously and making early diagnosis essential as one has the risk of developing profound hearing loss and the systemic disease can be fatal, hence early diagnosis has become of paramount importance to improve prognosis [5]. The otological symptoms tend to progress despite myringotomy or mastoidectomy [1]. The above patient had undergone a mastoidectomy twice with a progression of symptoms.

Neurological manifestations can occur but are rare in granulomatous polyangiitis. They may present with cranial nerve palsy, peripheral neuropathy cerebrovascular events, or cerebritis [6]. Three main histological frameworks have been identified: CNS vasculitis of the small vessels of the brain and spinal cord, granuloma invasion from the extracranial site, and isolated intracranial granulation [7]. The unusual manifestation in the aforementioned patient was the presentation of otitis media with trigeminal neuralgia, most patients with otitis media tend to present with facial nerve palsies as illustrated by Yasuaki Harabuchi et al facial nerve palsy can be caused by inflammatory granulation that spread through the facial canal, this is not uncommon with Otitis media associated ANCA vasculitis [5]. To our knowledge, this may be one of the first cases where otitis media presents with trigeminal neuralgia in a patient with GPA.

Her disease later progressed to peripheral length-dependent neuropathy after a pontine infarct. Peripheral neuropathy is the most common neurological manifestation as per De Groot K et al; he has a cohort of 128 patients with GPA 46% of the patients had peripheral neuropathy and 4% had cranial nerve palsies (8). Cerebral Vascular accidents are also an uncommon manifestation of GPA though can occur, De luna et al had 35 patients in her cohort of study all of them underwent an MRI of the brain and 43 % of patients had either ischemic or hemorrhagic change in the blood vessels [7].

Treatment should be initiated once the symptoms manifest though many patients that present with otitis media miss the early window on treatment and fall in the curve of high mortality rate. Treatment for ANCA vasculitis is induction of remission via high-dose steroids at 1mg/kg/day, where they normally pulse with steroids for two to three days [2]. The addition of cyclophosphamide tends to improve the remission time in patients with ANCA vasculitis, rituximab and plasma exchange are other possibilities for treatment as well [2].

ANCA vasculitis has a high rate of mortality when not diagnosed efficiently, the prevalence and incidence of ANCA vasculitis are low in Africa and this can be secondary to having low resource centers to carry out tests for confirmation [9]. In 2012 a study had been done in a Tunisian hospital and only 21 patients had GPA out of which 18 had ear, nose, and throat symptoms, and 11 presented with neurological symptoms [10]. Currently to our knowledge, we do not have any studies on ANCA vasculitis in Kenya leave alone the rare manifestation of GPA with Trigeminal neuralgia. This case may be one of the first reportable cases.

Otological symptoms and CNS symptoms are rare complications but can present as atypical symptoms in our case, it is essential to identify these cases early to prevent mortality rates as early treatment is the key to survival and remission.

Conclusion

ANCA vasculitis is a rare disorder unusual presentations include recurrent otitis media with trigeminal neuralgia. Late diagnosis can result in fatal outcomes and can be treatment refractory.

Conflict of Interest

The authors declare that there is no conflict of interest regarding the publication of this paper.

Consent

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

Ethical Statement

In line with our Institutional Ethics and Research Committee (IERC) guidelines, this case report was exempted from full IERC review.

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