UNUSUAL PRESENTATION OF ANCA VASCULITIS WITH OTITIS MEDIA AND CVA

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Abstract

A 42-year-old African female (Kenyan Origin), presents with recurrent ear fullness and pain in the ear diagnosed with otitis media there after progressing to having trigeminal neuralgia and multi-systemic disease. ANCA vasculitis is a rare disease of granulomatous polyangiitis targeting multiple organ systems.

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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 since 2016. Prior to 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-mastoditis. 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, 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 and hard 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 since 2016. Prior to 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-mastoditis with a mastoid effusion for which she underwent mastoidectomy after recurrent episodes of ear infection. Cultures obtained grew corrynebacterium urealytiucm and staphylococcus aureus for which she received antibiotics sensitive to gentamicin, cotrimoxazole, ciprofloxacin, clindamycin, erythromycin, vancomycin and tetracycline, and multiple tests or tuberculosis had been negative including cultures. HIV test was negative. A few months later she presented with trigeminal neuralgia, and received prednisone. Two weeks prior to her recent admission in 2019, she developed progressive bilateral lower limb numbness up to the calves. Two days prior to 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 temperature of 36.6 degrees centigrade. Chest and abdominal examination were unremarkable. Neurological examination revealed distal lower limb length dependent polyneuropathy bilaterally (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 remonstration of the oto-mastoidistis 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 ad been positive for Klebsiella oxycota for which she was treated with antimicrobial therapy and continued her maintenance therapy of prednisone, mycophenolate sodium, cotrimazole prophylaxis, alendronate and her antihypertensive medications. Unfortunately, she developed massive upper gastrointestinal bleed secondary to multiple vasculitis gastric ulcers evidenced by an upper endoscopy. She kept having persistent upper gastrointestinal bleed and in view of the vasculitis, she underwent plasma exchange. Over the next couple of days, she experienced persistent upper gastrointestinal haemorrhage for which she was transfused and a surgical opinion was sought. However, surgical interventions could not be instituted in view of 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 shows 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 demonstrates narrowing of the adjacent airway by approximately 50%.No invasion into the surrounding soft tissues or cartilaginous structures was 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, plasma blasts and plasma cells (2). These activate neutrophils and results in generation of C5a via the complete pathway, causing further activation of neutrophils(3). Activated neutrophils results in propagation of tissue damage and vascular inflammation. The main target organs in granulomatous polyangiitis include the ear, nose throat and upper respiratory tract, 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. Granulomatosis polyangiitis can affect the external, inner and middle ear. Otitis media is caused by granulomatous process that occurs in the middle ear and can also involve the mastoid cavity (4). In many cases it is difficult to differentiate from otitis media and granulomatous polyangiitis. 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 paramount importance to improve prognosis (5). The ontological symptoms tend to progress despite myringotomy or mastoidectomy (1). The above patient had undergone an mastoidectomy twice with progression of symptoms. Most cases of otitis media associated vasculitis (OMAAV) present with facial nerve palsy (6). Neurological manifestations can occur but are rare in granulomatous polyangiitis. They may present with cranial nerve palsy, peripheral neuropathy cerebrovascular events or cerebritis (7). Three main histological frameworks have been identified: CNS vasculitis of the small vessels of the brain and spinal cord, granuloma invasion from extracranial site, isolated intracranial granulation(8). The unusual manifestation in the aforementioned patient was the presentation of otitis media with trigeminal neuralgia and her disease progressed to peripheral length dependent neuropathy and there after a pontine infarct. Peripheral neuropathy is the most common neurological manifestation as per De Groot K et la; he has a cohort of 128 patients with GPA 46% of the patients had peripheral neuropathy and 4% had cranial nerve palsies (9). The unusual presentation we had with our patient is the presentation with chronic otitis media and trigeminal neuralgia, most patient with otitis media tend to present with facial never palsies as illustrated by Yasuaki Harabuchi et la 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). Cerebral Vascular accidents is 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 (8). ANCA vasculitis has a high rate of mortality when not diagnosed efficiently, the prevalence and incidence of ANCA vasculitis low in Africa and this can be secondary to having low resource centers to carry out tests for confirmation (10). It is important to treat as early once the symptoms manifest though many patient 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). Addition of cyclophosphamide tends to improve the remission time in patient with ANCA vasculitis, rituximab and plasma exchange are other possibilities for treatment as well (2). Otological symptoms and CNS symptoms are rare complications but can present as atypical symptoms as 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.

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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