ANCA-associated vasculitis causing nasopalatal destruction

S Mumtaz¹, S Navace¹, M Singh²

¹Royal Free London Foundation Trust, UK

²University Hospitals Bristol NHS Foundation Trust, UK

ABSTRACT

Autoimmune disorders and their manifestations can be very problematic to diagnose and manage. The symptoms can be ambiguous and debilitating. Early intervention is necessary to counter the unforgiving consequences of delayed intervention, such as loss of sight or hearing and destruction of affected tissues. Vasculitis in the head and neck region is rare. A case of ANCA-associated vasculitis causing extensive destruction is presented to enhance our understanding of this devastating multisystem disorder.

KEYWORDS

Otolaryngology – Autoimmune disorders – ANCA-associated – Vasculitis – Septal perforation

Accepted 21 February 2020

CORRESPONDENCE TO

Shadaab Mumtaz, E: shadaab@me.com

Case history

A 38-year-old Caucasian woman attended the emergency department complaining of a ‘hole in the palate’. The patient described that she ‘woke up’ with this problem and denied any pain, bleeding or dysphagia preceding the event. She claimed to have lost about ‘two stones’ in weight over the past nine months, which she related to ‘family stress’. No other associated history was noted, apart from intermittent flu-like symptoms and a productive cough six weeks previously, which settled with a course of antibiotics. There was no other significant past medical history. The patient was an ex-smoker (15 pack-years) who stopped smoking 4 weeks ago and consumed minimal amounts of alcohol per week. She denied use of any recreational drugs.

On examination, the patient appeared cachectic and weighed 54kg. She was alert and orientated, and vital observations were within normal limits. No obvious cervical lymphadenopathy or facial swelling was noted. Intraoral examination demonstrated a 1cm perforation on the left side of the hard palate communicating with the nasal cavity (Fig 1). Flexible nasendoscopy demonstrated destruction of the nasal septum with additional crusting and pus discharge. No other obvious pharyngolaryngeal disease was noted. Computed tomography and magnetic resonance imaging showed extensive bony destruction of the nasal septum, the hard palate and the medial walls of both maxillary sinuses (Fig 2). Histopathology confirmed acute and chronic inflammation along with necrosis. There was no evidence of malignancy.

Blood investigations revealed leucocytosis ($13.9 \times 10^9/l$), thrombocytosis ($507 \times 10^9/l$), eosinophilia ($0.83 \times 10^9/l$), elevated erythrocyte sedimentation rate (ESR) (106mm/h) and anaemia (91g/l). Autoimmune serology confirmed the presence of atypical perinuclear antineutrophil cytoplasmic antibodies (p-ANCA) on indirect immunofluorescence, confirming the diagnosis of ANCA-associated vasculitis (AAV). Normal levels of proteinase-3 and myeloperoxidase autoantibodies on enzyme-linked immunosorbent assay testing were noted. The patient was referred urgently to the rheumatology team after multidisciplinary team discussion.

Discussion

AAV is the nomenclature associated with a set of rare, potentially life-threatening autoimmune disorders affecting the small and medium-sized blood vessels, leading to necrosis of associated structures. The disease is classified broadly into three descriptive groups¹: granulomatosis with polyangiitis (GPA), microscopic polyangiitis (MPA), and eosinophilic granulomatosis with polyangiitis (EGPA).

Although any organ system may be involved, GPA and EGPA tend to mainly affect the ear, nose and throat regions. Other organs commonly affected include the kidneys, lungs, skin, eyes and nervous system.² The clinical features may include flu-like symptoms, bloody rhinorrhoea, nasal polyposis, hearing loss, conjunctivitis,

uveitis, anorexia and unexplained weight loss. Blood investigations may show leucocytosis, thrombocytosis, elevated ESR and normocytic normochromic anaemia. Due to non-specific symptomatology, diagnosis is usually by exclusion and autoimmune serology testing.

Management includes use of immunosuppressive medications such as cyclophosphamide and high-dose glucocorticoids for three to six months, depending on the outcome. After induction therapy, a maintenance regimen is required to continue remission. Up to half of treated patients relapse within five years.⁵

Figure 1 Palatal perforation leading to oronasal communication

Figure 2 Computed tomography imaging showing extensive destruction of the nasal cavity and hard palate

In conclusion, early diagnosis and a broad-based multidisciplinary approach are essential to evaluate and manage these sinister presentations. Surgeons and trainees should consider autoimmune disorders in the surgical sieve to systematically evaluate potentially malignant-looking lesions.

References

1. Jennette JC, Falk RJ, Bacon PA *et al.* 2012 revised International Chapel Hill Consensus Conference Nomenclature of Vasculitides. *Arthritis Rheum* 2013; **65**: 1–11.
2. Jennette JC, Falk RJ. Small-vessel vasculitis. *N Engl J Med* 1997; **337**: 1512–1523.
3. Berden A, Göçeroglu A, Jayne D *et al.* Diagnosis and management of ANCA associated vasculitis. *BMJ* 2012; **344**: e26.