Clinical images of ENT involvement in ANCA associated Vasculitis

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Anti-neutrophil cytoplasmic autoantibody associated vasculitis (AAV) are rare necrotizing vasculitides in which ear, nose and throat (ENT) involvement is responsible for the presenting symptoms in most patients, particularly in those with Granulomatosis with Polyangiitis (GPA) and Eosinophilic Granulomatosis with Polyangiitis (EGPA), frequently preceding other manifestations. Symptoms are often non-specific, and patients may be misdiagnosed as having an infectious or allergic condition. However, ENT involvement can be severe and the delay in diagnosis can result in important sequelae, secondary to tissue destruction in the affected areas, compromising function and quality of life. Close collaboration between otorhinolaryngologists and rheumatologists is therefore fundamental for the best management of these patients.

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We present 4 clinical images of 4 patients with GPA and different types of ENT involvement.

The first patient is a 47-year old woman with epistaxis, rhinorrhoea, aural fullness, nasal crusting and saddle nose deformity (Figure 1A) associated with pulmonary, renal and musculoskeletal involvement. Saddle nose deformity is due to progressive destruction of the cartilaginous framework of the nose and occurs in around 23% of GPA patients[1,2].

The second patient is a 45-year old man with long standing GPA and interstitial lung disease. He also presented with chronic sinonasal inflammation and progressive destructive midline lesion (Figure 1B). Nasal septum perforations are usually caused by vasculitis in the Kiesselbach area. This is one of the most frequent

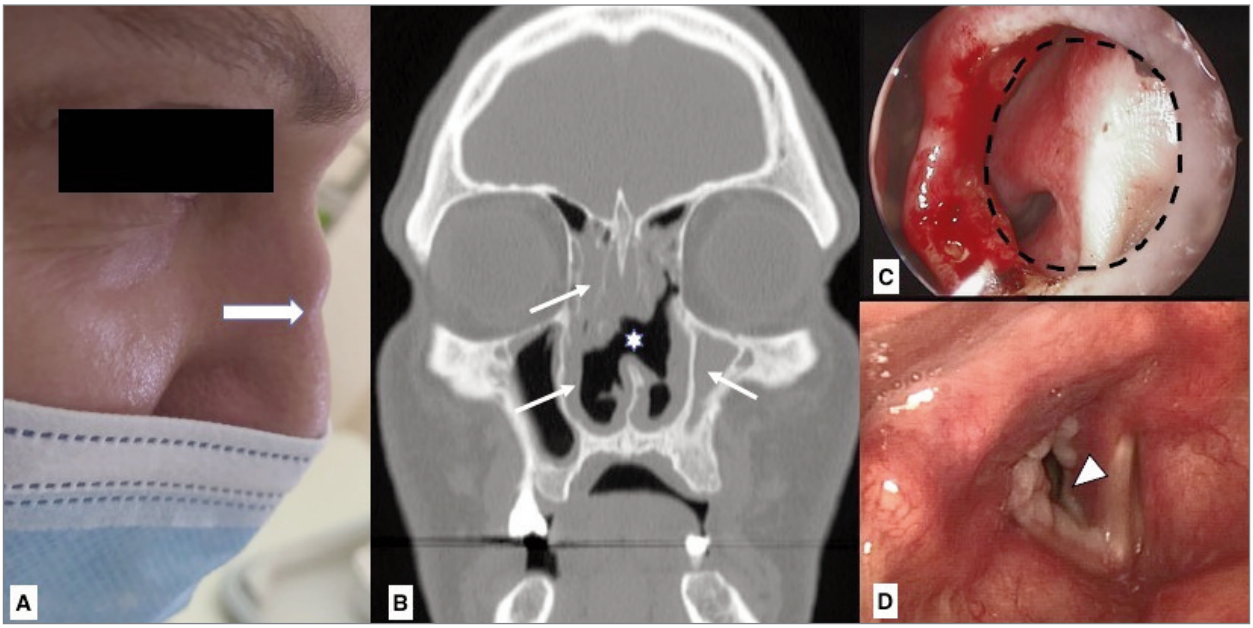

FIGURE 1. Different types of ENT involvement in ANCA associated vasculitis. A – Saddle nose deformity; B – Nasal septum perforation (asterisk) and sinonasal inflammation (arrows); C – Nasal septum perforation on nasal endoscopy; D – Subglottic stenosis (arrow head)

ENT manifestations in GPA, affecting approximately 33% of cases[2].

The third patient is a 46-year old woman with localized GPA, with exclusive ENT involvement. She presented with nasal septum perforation and sinonasal inflammation (Figure 1C). Localized GPA occurs in more than 25% of GPA cases[3]. Nevertheless, most will progress to generalized GPA, with only 5% remaining localized[4].

The fourth patient is a 58-year old woman with otitis media with effusion, nasal crusting and subglottic stenosis (Figure 1D), and multi-organ involvement. Subglottic stenosis is caused by excessive reconstruction that follows destruction of surrounding tissues due to vasculitis. Occurs in around 10-20% of GPA patients[5].

In addition to systemic medication for AAV, patients received local treatments, including nasal douching and hydration, topical steroids (fluticasone nasal puffs and diluted budesonide) and antibiotics (trimethoprim–sulfamethoxazole), which were very useful in improving ENT symptoms and quality of life.

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