Cocaine Induced ENT pseudo-GPA (CIE pGPA)

British Society of Facial Plastic Surgery and British Rhinology Society Guidelines

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Cocaine-induced vasculitis is a drug-induced vasculitis that causes severe rhinitis with nasal tissue necrosis1. Once a rarity, it is becoming increasingly prevalent within ENT outpatient clinics. This is the first ENT-UK guideline on the management of this disease. Agreement on terminology for this entity is still evolving with recent evidence favouring 'cocaine induced pseudo-Granulomatosis with Polyangiitis (GPA)', which better encompasses its pathogenesis and correctly prompts anti-inflammatory treatment with immunosuppression as well as immediate cessation of cocaine. Older terminology such as 'cocaine-induced midline destructive lesion' (CIMDL) falls short in recognising its pathogenesis and treatment2.

Background to Guidelines

Nasal exposure to cocaine, especially if mixed with levamisole powder induces (in certain individuals) a severe localised inflammatory reaction, leading to severe rhinitis and tissue necrosis. Little is understood about the mechanisms behind the onset and severity of the disease (e.g. dose exposure vs. individual genetic or other sensitisations), but there is increasing recognition of its fundamentally inflammatory nature and that treatment should include immunosuppression in addition to strict cessation of cocaine and symptomatic measures.

Diagnosis

The definitive diagnosis of cocaine-induced ENT pseudo-GPA is based on the clinical finding of a localised nasal vasculitis-like presentation (destructive/necrotic appearance) commonly associated with severe pain in the context of cocaine use, having excluded other systemic diseases such as GPA and neoplasia. In 70% of cocaine-induced ENT pseudo cases, a positive pANCA is demonstrated with a PR3 preponderance. In Cocaine Induced ENT pGPA histology is important (to exclude neoplasia and invasive fungal disease) but often shows chronic inflammation only, unlike true GPA which typically shows granulomatous lesions. In cocaine-induced ENT pseudo-GPA, there is also a lack of general malaise, cANCA testing is negative and urine dipsticks, U&Es and chest imaging are normal which can help exclude Granulomatosis with Polyangiitis (true GPA). Imaging can demonstrate the extent of localised tissue necrosis - typically involving the nasal septum with variable extension towards (or even through) the columella, palate, lateral nasal wall, orbit, or skullbase.

Cocaine Screening

Cocaine Screening is essential - Cocaine use is often denied or claimed as historical. Urine tests remain positive for 3-14 days (depending on regularity and dose used). Cocaine use is