

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

most frequent at weekends, so tests early in the week can be more helpful. Hair tests may remain positive for months to years and are due to become available in the near future.

CIE pGPA is a chronic inflammatory process with acute flares-ups accompanied by tissue destruction. Patients should be advised that treatment is possible and may require immunosuppression medication, but cocaine use must be completely avoided for this to be effective and to prevent relapse.

Guidance

In the presence of severe rhinitis and tissue necrosis:

- **A full history and examination is essential.**
- Common complaints include nasal pain (often severe, with tenderness), bleeding, extensive nasal crusting, and excoriation of the skin around the nasal vestibule.
- Cocaine use may be confirmed but is frequently denied, or recounted as historical.
- Nasal endoscopy shows localised severe rhinitis and often necrosis, including bleeding, crusting, and variable extension of tissue destruction (as per imaging described above).
- Patients with potential CIE pGPA must be reassured that their medical care is confidential, and they should understand that regular urine testing is required including potential hair sampling to avoid misdiagnosis and harmful erroneous treatments. Written information can be helpful in this.

Investigations

1. Urine sample for cocaine metabolites: Samples should be standard practice (e.g. every clinic visit) to check for undeclared or continued cocaine use as well as exclude dipstick changes seen in GPA.
2. Any patient refusing urine tests should be discharged back to their GP to consider drug rehabilitation, however, patients can also be in a desperate position and will need our support.
3. Blood tests (vasculitic screen): ANCA +/- ENAs, FBC, U&Es, ACE, ESR, and CRP
4. CT scan of sinuses: important to determine disease extent and tissue destruction.
5. Urgent EUA and biopsy: tissue samples obtained ideally before active medical treatment to help exclude other causes such as GPA, neoplasia and invasive fungal disease.
6. Additional investigations: In cases with any uncertainty, chest imaging, U&Es, and serial bloods (ANCA, ENAs) should be considered to revisit the possibility of vasculitis (usually GPA).

Management

1. Cocaine use must be stopped, and regular urine tests monitored for compliance.
2. Symptomatic treatment should be advised, including saline nasal douches, appropriate analgesia, and topical nasal ointments if required, particularly if *Staphylococcal aureus* colonisation is suspected.
3. Both oral and Intranasal corticosteroids may be advocated for immediate treatment but responses can be variable.

4. Patients with suspected CIE pGPA should be referred urgently to a specialist vasculitis team prior to immunosuppression.
5. In the scenario with active destructive nasal disease whereby cocaine use is denied and urine tests are negative, treatment should be based on individual patient assessment and still referred on for consideration for immunosuppression. Hair sampling, once available, will be very helpful in this situation.
6. Patients actively taking cocaine or in denial should be discharged back to their GP for urgent rehabilitation, albeit challenging and such community pathways are not standardised.
7. Occasionally, a patient with GPA may give an honest history of previous cocaine use that is no longer current. GPA is associated with serious renal and pulmonary disease and in this situation, the diagnosis of GPA should be taken seriously and managed accordingly.

Specialist Management

The role of immunosuppressive treatment is evolving and administered by experienced multidisciplinary vasculitis teams.

Limits of Surgical Treatment

The role of early surgery (beyond biopsies for diagnostic exclusion) is not recommended. It is tempting to offer debridement, restoration of the nasal airway, or sinus ventilation, but such procedures may exacerbate the disease, especially should cocaine use continue. Improving the nasal airway may paradoxically facilitate continued use of cocaine.

Surgical reconstruction is not recommended until there is a collaborative agreement between specialists directly involved in the patient care, or a specific MDT if one exists, that all clinical and biochemical signs of active inflammatory disease have remitted for a reasonable period, such as 18 months. Patients must also be warned before any procedures that surgery may reactivate their vasculitic disease and cause reconstruction to fail.

References

1. **Granulomatous Disease, Vasculitides and the Cocaine nose.**
Andrew C. Swift and Peter Andrews, Chapter 46, 593 – 607, In Contemporary Rhinology: Science and Practice, 2023, Eds. AC Swift, S Carrie, C de Souza, Springer Nature.
2. **The role of ANCA in the management of cocaine-induced midline destructive lesions or ENT pseudo-granulomatosis with polyangiitis: a London multicentre case series.**
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