

Dental Management of a Patient with Cold Agglutinin Disease

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Aim

- To present the risk assessment and treatment modifications required for management of a complex haematological disorder, namely cold agglutinin disease



Objectives

- To provide an overview of cold agglutinin disease (CAD)
- To review the currently available literature in relation to the dental implications / management
- To present a case study of a patient with CAD presenting for dental care
- To discuss the risk assessment & treatment modifications in relation to this case

Cold agglutinin disease

- Rare variant of autoimmune haemolytic anaemia
- Destruction of red blood cells by autoantibodies (cold agglutinins)
- Triggered by exposure to cold temperatures

Cold Agglutinin Disease | Genetic And Rare Diseases Information Center (GARD) – An NCATS Program". *Rarediseases.info.nih.gov*. 2017. (Accessed online 25th Apr 2017)

Cold agglutinin disease

- Acquired
- Primary – idiopathic
- Secondary
 - Infections: influenza, HIV
 - Autoimmune conditions: SLE
 - Cancers: lymphoma, chronic lymphocytic leukaemia, multiple myeloma

Cold agglutinin disease

- M = F
- Middle aged and older
- Peak: 7th & 8th decade
- Incidence: 1 in 1 000 000 / year
- Prevalence: 1 in 60 000
- Prognosis: variable

Cold Agglutinin Disease (CAD). *Cold Agglutinin Disease* http://coldagglutinindisease.org/?doing_wp_cron=1493621989.9759299755096435546875. (Accessed online 28th Apr 2017)

Cold agglutinin disease

Signs and symptoms:

- Related to anaemia

General

- Chronic fatigue
- Pallor
- Dizziness
- Dyspnoea
- Irritability

Oral

- Mucosal pallor
- Glossitis
- Angular cheilitis
- Ulceration

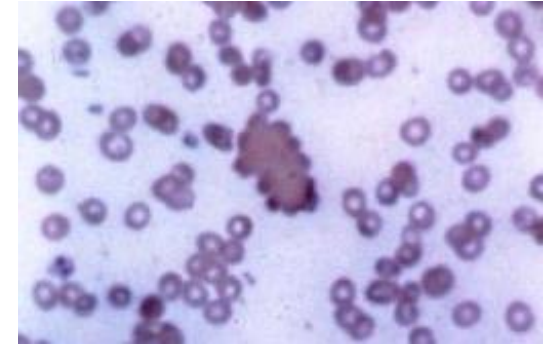
Cold agglutinin disease

Signs and symptoms:

- Acrocyanosis (purplish discolouration) of fingers, toes, tip of the nose, ears
- Ischaemic changes of skin
- Jaundice
- Dark urine
- Heart failure
- Shock

Packman, C. (2015). The clinical pictures of autoimmune hemolytic anemia. *Transfusion medicine and hemotherapy*. 45(5):317-324.

Cold agglutinin disease



Diagnosis:

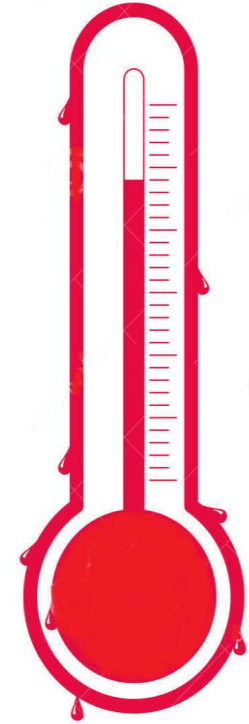
- FBC – abnormal agglutination
- Peripheral blood smear – agglutination
- Direct antiglobulin test (DAT) – positive for IgM
- Cold agglutinin titre
- Urinalysis – haemoglobinuria

Hill, Q., Stamps, R., Massey, E., Grainger, J., Provan, D., Hill, A. (2017). The diagnosis and management of primary autoimmune haemolytic anaemia. *British Journal of Haematology*. 176: 395-411.

Cold agglutinin disease

Treatment:

- Treat underlying cause
- Avoid exposure to cold
- Monoclonal antibodies
- Plasma exchange
- Blood transfusions



Barcellini, W. (2015). Current treatment strategies in autoimmune hemolytic disorders. *Expert Review of Hematology*. 8(5):681-91.

Zanella, A. and Barcellini, W. (2014). Treatment of autoimmune hemolytic anemias. *Haematologica*. 99(10): 1547-1554

Literature review

- One case report

Kerley, T. *et al* (1979)

- 59yo male
- CAD and advanced periodontal disease
- Dental clearance, alveoplasty, gingivoplasty under LA
- Special measures: Warming surgery to 32°C, IV fluids, closure of surgical site

Kerley, T., Schow, C. and Mulherin, D. (1979). Treatment of autoimmune hemolytic anemias. *Oral Surgery Oral Medicine Oral Pathology Oral Radiology*. 48(1): 39-40.

Case Presentation – Patient H

- 75 year old male
- Referred by Haematology
- Dental Haematology Services at UCLH



Presenting Problem

1. Receding gums
2. Intermittent pain from the lower gums



Past Medical History

1. Cold agglutinin disease

- Diagnosed 6 years previously
- Under Haematology
- Management with:
 - Rituximab
 - Eculizumab
 - Blood transfusions
 - Plasma exchanges

Past Medical History cont'd

2. Lymphoplasmacytic lymphoma

- Diagnosed 3 years previously
- Chemotherapy ongoing - Bendamustine
- Close review



Past Medical History cont'd

3. Diabetes
4. Hypothyroidism
5. Vitiligo



Social History

- Married
- Occupation: Film director / writer
- Smoking: Nil
- Alcohol: Rarely



Relevant Dental History

- Registered with GDP
- Hygienist x 2 / year

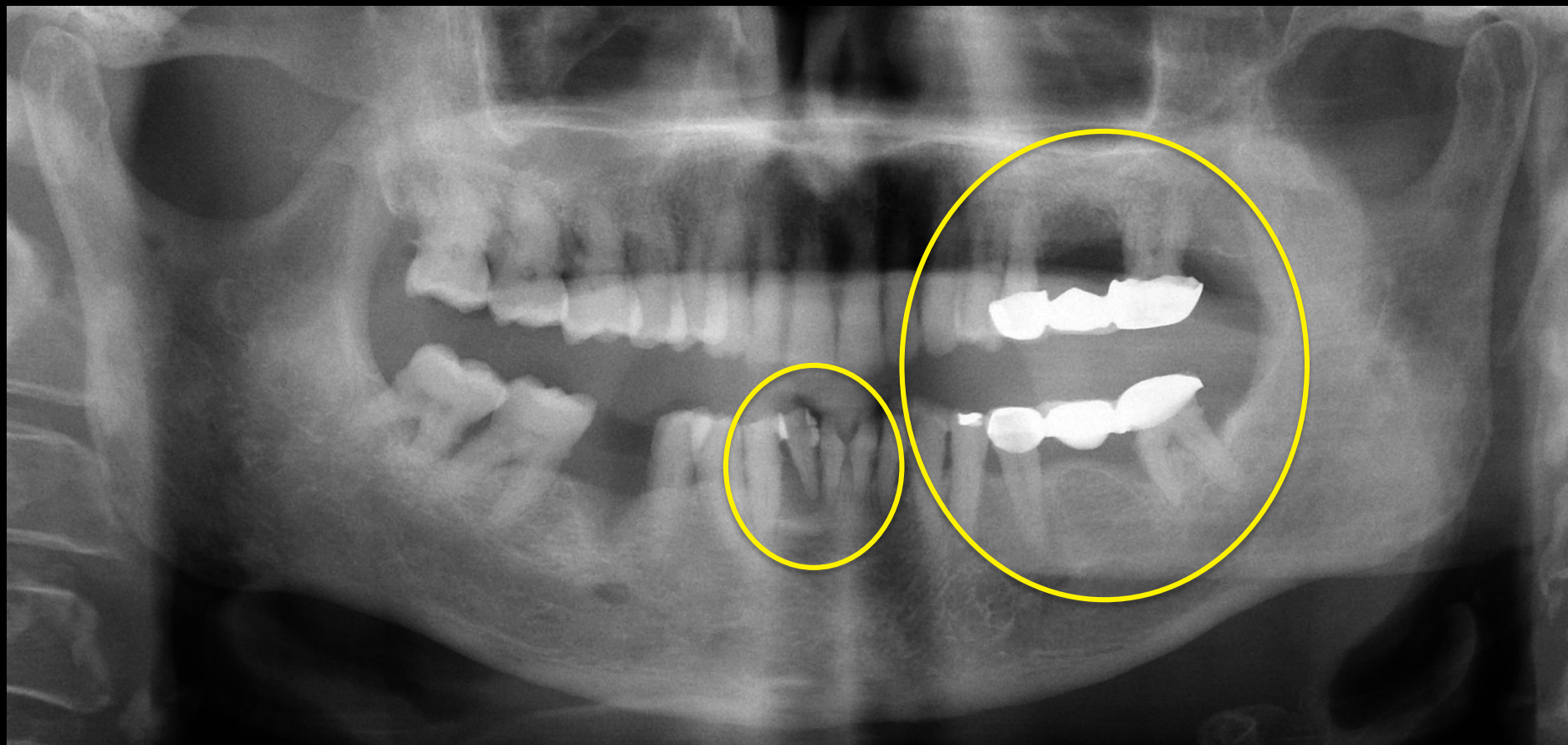


Clinical Examination



- Heavily restored dentition
- Extensive supra and sub gingival calculus deposits
- Mobility:
 - Grade I – LR8
 - Grade II – UR2, LL1, LL2
 - Grade III – LR2

Radiographic Examination



Dental Diagnoses

- Generalised severe chronic periodontitis
- Chronic periodontal abscess of lower right lateral incisor (LR2)

Risk Assessment



Social:

- Ability to attend appointments
- Timing of appointments

Dental:

- Dental setting (secondary care)
- Reduced co-operation (fatigue)
- Reduced manual dexterity
- Increased susceptibility to periodontal disease

Risk Assessment



Medical:

- Risk of acute haemolysis
- Risk of hypoxia
- Risk of bleeding
- Risk of infection
- Risk of delayed healing
- Risk of hypoglycaemia

Treatment Plan

1. Oral hygiene instruction
2. Extraction of lower right lateral incisor
3. Scaling
4. Root surface debridement
5. Review



Treatment Modifications

1. Access
2. Communication
3. Consent
4. Education
5. Surgery
6. Spread of infection

1. Access

- Appropriate setting – secondary care, in case of an acute event
- Haematology team – inform of appointments
- Timing of appointment – morning, consider chemotherapy cycles
- Pre-operative investigation results – blood tests



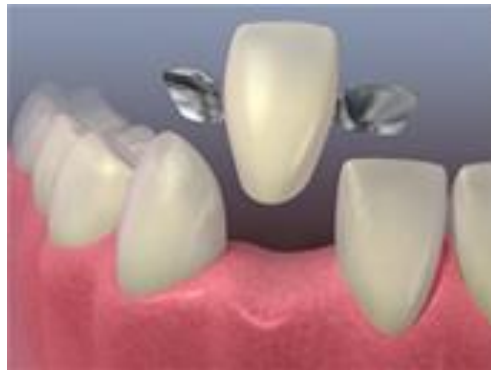
2. Communication

- Patient – consent, special requirements, manage expectations
- Liaison with Haematology team
- Laboratory – blood sample warming requirements



3. Consent

- Informed consent
- Risks of intervention vs no intervention
- Gap replacement options



4. Education

- Periodontal disease aetiology
 - Local – gross plaque and calculus deposits
 - Systemic – immunosuppression, diabetes, fatigue
- Preventative advice
 - Oral hygiene
 - Dietary



5. Surgery

- Pre-operative:
 - Blood test
- Peri-operative:
 - Supplemental oxygen
 - Monitoring vital signs
 - Local haemostatic measures
- Post-operative:
 - Antibiotics



6. Spread of infection

- Standard infection control precautions
- Blood products



Protection against blood borne infections in the workplace: HIV and hepatitis. *Health and Safety Executive.*

Standard Infection Control Precautions (SICP). (2016). *NHS Professionals.* <http://nhsprofessionals.nhs.uk> (Accessed online 28th Apr 2017)

Conclusions

- CAD is a rare haematological disorder
- Multidisciplinary approach when planning dental care
- Appropriate risk assessment and treatment modifications are required to facilitate dental treatment
- Principles are applicable to other haematological disorders

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**Thank you
for
listening**

