



Cyclic neutropenia. Clinical case report

Neutropenia cíclica. Reporte de un caso

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ABSTRACT

Cyclic neutropenia occurs due to the fluctuation of cellular production levels of bone marrow stem cells. This is to say, they change during the cycle, and although numbers are recurrently low, function is normal. Cyclic neutropenia was first described by Leale in 1910 with dominant autosomal character. It manifests approximately every 21 days, with range of 14 to 36 days, lasting 3-6 days per episode. During the time when there are few circulating neutrophils, the patient is susceptible to infections. The clinical picture of this process includes: susceptibility to infection, feverish conditions, oral ulcers, impetigo, increased lymph nodes, periodontitis and stomatitis. It is important to stomatologically handle these patients; after inter-consultation with treating physician, prevent infection in a prophylactic scheme based on amoxicillin 50 mg/per kg weight or clindamycin 20 mg/per kg weight, review recent blood count results (maximum ten days before treatment). In emergency treatments pain and infection will be handled conservatively, with use of 0.12% chlorhexidine mouthwashes and Philadelphia solution when necessary.

Key words: Cyclic neutropenia, odontology, mouth rehabilitation, prevention.

Palabras clave: Neutropenia cíclica, odontología, rehabilitación bucal, prevención.

RESUMEN

La neutropenia cíclica ocurre debido a que los niveles de producción celular por parte de las células madre de la médula ósea fluctúan, es decir, cambian durante el ciclo; aunque el número es recurrentemente bajo, su función es normal. Fue descrita por primera vez por Leale en 1910, con carácter autosómico dominante. Se presenta aproximadamente cada 21 días con un rango de 14 a 36 días durando un total de 3 a 6. Durante el periodo en el que existen pocos neutrófilos circulantes, el paciente es susceptible a las infecciones. Dentro del cuadro clínico se presentan: susceptibilidad a infecciones, cuadros febriles, fatiga, úlceras orales, impétigo, aumento de ganglios linfáticos, periodontitis, estomatitis. Es importante manejar estomatológicamente a estos pacientes, previa interconsulta con médico tratante, prevenir cuadros infecciosos bajo un esquema profiláctico a base de amoxicilina 50 mg/kg peso o clindamicina 20 mg/kg peso, revisar biometría hemática reciente (máximo 10 días previos al tratamiento); en tratamientos de urgencia se manejará de forma conservadora el dolor e infección, uso de enjuagues con clorhexidina al 0.12% y solución Philadelphia en caso de ser necesario.

INTRODUCTION

Patients afflicted with cyclic neutropenia suffer periodic decreases of neutrophil numbers; this is an infrequent disorder and is characterized by habitual fever episodes.

Cyclic neutropenia occurs due to the fluctuation of bone marrow stem cells' cellular production, that is to say, a change during the cycle. Although the number is recurrently low, their function is normal.

Neutrophils are polymorphonuclear granulocytes, they approximately measure 8 to 12 micrometers, they are the most abundant leukocytes present in the blood, they live for 72 hours after having exited the bone marrow, and thus can be considered cells with the following characteristics: they are phagocytic, mobile cells, with gelatinous consistency; they can easily cross blood vessels as well as respond to inflammation and infection stimuli five hours after initiation of process.

Therefore, neutropenia can be considered a significant decrease in the number of polymorphonuclear

neutrophils per mm³ in peripheral blood. It is caused by alterations in cell production, excessive destruction of peripheral tissues, cell non-maturation or a disrupted cell distribution.¹

These patients exhibit certain susceptibility to infections; therefore, neutropenia can be classified in the following manner:

- a) Length: acute, chronic.
- b) State of medullar reserve: preserved or low.

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c) Severity: mild, moderate or severe.

d) Nature or origin:

1) Acquired: medication:

- Anticonvulsant.
- Antihistaminic.
- Barbiturates.
- Chemo-therapeutic drugs.
- Diuretics.
- Sulfonamides/antibiotics.

2) Congenital:

- Kostmann disease.
- Shwachman syndrome.
- Cyclic neutropenia.
- Mielokatesis.

Cyclic neutropenia was first described by Leale in 1910. He depicted it as a condition with autosomal dominant character, a result of the mutation of EIA2 gene, position 13.3 of the short arm of chromosome 19 which codifies neutrophil elastase. Its time span is occurrence within a rank of 14 to 36 days, normally around day 21, with duration of 3 to 6 days. It is more common for infections to appear during the period when there are few circulating neutrophils.²

There is an interruption in cell production of bone marrow stem cells. This can cause from normal production up to severe stagnation of neutrophil maturation. Pharmacological treatment of granulocyte colonies stimulating factor (G-CSF) promotes maturation of neutrophil-precursor cells.

Under normal circumstances, G-CSF is mainly produced in the bone marrow. When there is bacterial infection, G-CSF production increases due to the stimuli produced by certain components of the infectious agent on immune system cells. Final result is increase of neutrophil maturation in the bone marrow, greater release of said neutrophils into the bloodstream as well as activation of said cells functions, that is to say, their ability to destroy pathogen agents.

With respect to incidence it can be said that frequency is 2 cases per million subjects.³

The clinical picture might include the following manifestations: susceptibility to infections, fever, fatigue, impetigo, increase in the size of lymph nodes, periodontitis, cheilitis, stomatitis.

Diagnosis can be emitted when the following can be established: cyclic episodes of 200 total neutrophils per mm³, every three weeks for a period of 3 to 6 days. To achieve confirmation, blood count must be performed 2 to 3 times a week, during 8 weeks.

Treatment is IV administration of «granulocyte colonies stimulation factor» (G-CSF), whose function, as indicated by its name, is to stimulate neutrophil

production and shorten neutropenia duration, which brings along symptom's decrease. In cases when the patient does not react favorably, bone marrow transplant or corticosteroids can be used.

When patients reach adolescence, they experiment symptom decrease and cycles become less noticeable.⁴

PROTOCOL FOR STOMATOGNATHIC ASSESSMENT OF PATIENT WITH CYCLIC NEUTROPENIA CONDUCTED AT THE MEXICO'S CHILDREN'S HOSPITAL «FEDERICO GÓMEZ»

- 1) Condition identification, determination and detection of signs and symptoms characteristic of the condition.
- 2) Inter-consultation with treating pediatric hematologist concerning:
 - a) Actual status of the patient.
 - b) Recent blood counts (total neutrophil count).
 - c) Phase of neutropenic cycle which the patient is presently undergoing.
 - d) Type of pharmacological treatment used by the patient to control his neutropenic cycle.
- 3) Potential problems that can be encountered:
 - a) Formation of infectious processes.
 - b) Periodontitis.
 - c) Herpetic stomatitis.
 - d) Presence of acute pain.
 - e) Thrombocytopenia data when they are acquired by chemo-therapeutic drugs.
- 4) In order to avoid complications it is important to follow:
 - a) Anti-bacterial prophylactic scheme based on amoxicillin 50 mg/kg weight or clindamycin 20/mg/kg weight in case of allergy so as to avoid infectious processes. It can be used as therapeutic scheme (every 8 hours for 7 days) whenever appointments are consecutive, or as a prophylactic scheme (1 hour before initiating dental treatment, single doses) whenever it is an emergency treatment or isolated appointments.
 - b) Revision of total number or neutrophils in blood counts conducted before and after dental treatment.
- 5) Modifications to treatment plan:
 - a) When leukocyte count suddenly drops, antibiotics must be used in order to avoid post-operative infection.
- 6) Emergency treatments:
 - a) Avoid use of non steroid anti-inflammatory drugs, especially acetylsalicylic acid when there are thrombocytopenia data, nevertheless, paracetamol can be used.

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