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CYCLIC NEUTROPENIA FROM RIYADH, SAUDI ARABIA

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ABSTRACT

Cyclic neutropenia has first been described in the literature in 1910, which discussed a 19 -month -old boy, who presented with periods of recurrent neutropenia, mouth ulcers, and fever[1]. Over the years our understanding of this disease has been refined with the submission of multiple case reports worldwide and attempts to unify the diagnosis and disease course. We present a case of an adult male with cyclic neutropenia, the first of its kind to be described from Saudi Arabia.

KEYWORDS: *Neutropenia; Neutrophils, Eosinophils, Monocytes, Hematocrit And Platelet*

Article History

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INTRODUCTION

Case

A 28 -year -old Saudi male patient presented to an outpatient clinic on 09/01/2018, in a private hospital, in Riyadh, Saudi Arabia, complaining of oral thrush of 2 days durations. It was not associated with any oral ulceration, stomatitis, fever, or other constitutional symptoms. He then described a history of recurrent oral thrush, occurring on 3 occasions in the previous 4 months. The thrush would persist for 2-4 days, then subsequently subside. He tried using miconazole gel on one occasion, which helped shorten the duration of symptoms. During these attacks, he had no other complaints.

His past medical history is unremarkable, with no history of chronic diseases, no use of prescribed, over the counter, or herbal medications. His family history is also unremarkable, but his brother has a history of infertility for 5 years, due to low sperm count. No similar symptoms in first degree relatives, and no known history of genetically inherited diseases.

Physical examination showed a healthy appearing, asymptomatic male, with oral thrush. The rest of the examination was unremarkable, with no organomegaly, no lymphadenopathy, and no genital or oral ulcers or any dermatologic or perianal abnormalities noted. He was afebrile, with an oral temperature of 37°C, and other vital signs were within normal limits.

A serum analysis was done on the same day (09/01/2018), with CBC and chemistry tests, which showed a WBC count of 3.53 K/UL (reference range 4.500-11.000), with isolated neutropenia; neutrophils 0.9 K/UL (reference range 1.700-7.000). Eosinophils, monocytes, hematocrit, and platelet count were within normal ranges. Other serum chemistries

such as liver function tests and renal function tests were within normal limits. There was also no elevation in ESR; 7.00 mm/hr (reference range 0.0-0.15) or CRP; 0.1 mg/dl (reference range 0.0-5.0).

The patient presented again a month later (16/02/2018), complaining of oral ulcers, with otherwise unremarkable history, and physical examination. His CBC and differentials at this time showed a WBC count of 3.50 K/UL (reference range 4.500-11.000 K/UL), and once again, isolated neutropenia; neutrophils count 0.92 K/UL (reference range 1.700-7.000).

He was then brought for a routine CBC with differentials on 2 other occasions, once on 02/03/2018, which showed a normal WBC and neutrophil count; 5.01 K/UL and 2.0 K/UL, respectively, and on 05/09/2018, also a normal set of readings; WBC 3.51 K/UL and NEU 1.1 K/UL. Semen analysis was also done on 05/09/2018, which showed a decrease sperm count; 14.800 *10e6/mL (reference range 20.000-300.000), and a decrease in sperm motility; 35.000 % (reference range 60.000-100.000).

A diagnosis was based on the signs and symptoms of recurrent infections on separate occasions, with supporting evidence of neutropenia during these episodes of infection. The patient was only counseled to use miconazole gel during any episodes of oral thrush.

LITERATURE REVIEW

Cyclic neutropenia is a rare, congenital, primary hematologic disorder, which is characterized by recurrent low number of neutrophils (neutropenia), oscillating at different intervals. During these neutropenic episodes, patients usually present with fevers, oropharyngeal infections, such as gingivitis, pharyngitis, oral ulcers, and thrush. It is also reported that patients may present with perianal cellulitis, abdominal pain from gastrointestinal ulcers, cervical lymphadenopathy, among others[2]. Hematologic studies show neutropenia, and may also elevate platelet counts, elevated monocytes, and decreased hematocrit[2]. Earlier case reports also described the presence of eosinophilia and anemia[3]. Between neutropenic episodes, individuals are generally healthy and asymptomatic[2].

Some studies describe it to have an autosomal dominant pattern of inheritance[3], while others report that only one-fourth of patients to have significant family history[4]. The prevalence of cyclic neutropenia is 1:1,000,000 in the general population including those cases of familial and simplex type[1].

Cyclic neutropenia is usually diagnosed within the first year of life based on approximately three-week intervals of the signs and symptoms mentioned above, with supporting hematologic evidence of neutropenia[2]. Genetic testing is usually supplementary, and studies over the years have pointed to multiple heterogeneous genetic defects in ELANE, GFI1, HAX1, G6PC3, JAGN1, VPS45 or activating mutations in the Wiskott-Aldrich syndrome (WAS) gene, which is all linked to arrests in neutrophil maturation[1].

Treatment of cyclic neutropenia includes targeting infections associated with neutropenic episodes, and reducing the severity of such infections once neutrophil counts normalize. Use of granulocyte colony stimulating factor (G-CSF)[2], has been reported as the mainstay treatment, as well as use of antibiotics, rigorous oral and dental care[6]. Once the neutrophil count returns back to baseline, some studies report using G-CSF to increase resistance to developing infections, and some patients may benefit from hematopoietic stem cell transplantation[2].

DISCUSSIONS

Our patient's symptoms included oral thrush and ulcers, but other symptoms mentioned in the literature, such as fevers, perianal cellulitis, abdominal pain from gastrointestinal ulcers, cervical lymphadenopathy[2], etc. His repeat CBCs with differentials demonstrated only isolated neutropenia, with no changes in platelet, eosinophil, monocyte, or hematocrit count[2]. Interestingly enough, patients with this disorder usually present in early childhood[2], and gradually decrease in severity once they reach adulthood[1], but our patient actually started developing symptoms of recurrent infections in adulthood. His lack of family history also points to a more sporadic, congenital genetic defect, as some of the literature reported an autosomal dominant mode of inheritance[3]. On semen analysis, a decrease sperm count, and motility were discovered, which has been described in a case report of a family with cyclic neutropenia[5], but is otherwise a rare association.

Finally, due to the mild symptoms of oral thrush and oral ulceration during the patient's neutropenic episodes, it was decided that only symptomatic use of oral miconazole gel.

CONCLUSIONS

Cyclic neutropenia has been reported in the literature as a disease that is diagnosed in early infancy, while our patient presented in adulthood. We also would like to point to the fact that, during his neutropenic episodes, only oral thrush would appear, with no other signs and symptoms. This may further expand our knowledge of the discrepancy of the severity of this disease, as our case was very mild. More research is needed to estimate the prevalence of cyclic neutropenia in the Saudi population, as this is the only mentioned case report from this country.

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