

Recurrent Periorbital Cellulitis Secondary to Cyclic Neutropenia

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Abstract

Purpose: To present a 1-year-old boy with cyclic neutropenia who presented with multiple episodes of periorbital cellulitis (POC).

Methods: The child presented with three episodes of POC. In the second episode, the cellulitis was associated with nasolacrimal duct obstruction, and in the third episode, a pansinusitis was noted. He underwent a thorough systemic evaluation.

Results: Patient's evaluation revealed the diagnosis of cyclic neutropenia.

Conclusion: This report emphasizes the possibility of an underlying immunodeficiency with recurrent POC, even in an apparently healthy child.

Keywords: Cyclic neutropenia, Dacryocystitis, Orbital cellulitis, Periorbital cellulitis, Sinusitis

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INTRODUCTION

Periorbital cellulitis (POC) is an inflammatory process of the eyelids and periocular soft tissues anterior to the orbital septum. POC can be potentially dangerous because of the possibility of progression to periorbital and orbital abscess, blindness, and intracranial complications. The most common origin of the POC in children is rhinosinusitis, and children with rhinosinusitis suffer from orbital complications more commonly than adults. POC might also be caused by trauma, external ocular infections, local skin inflammations, dental infections, and periorbital malignancies.¹

Recurrent POC is a rare condition, with a few reports in the literature. Herein, the authors present a case of recurrent POC secondary to cyclic neutropenia, associated with the review of the literature.

CASE REPORT

A 1-year-old boy was admitted to our hospital with a 2-day history of right periorbital swelling and erythema [Figure 1c]. The patient had mild rhinorrhea, nasal congestion, and low-grade fever. Extraocular motility was normal, and there were no abnormalities in the pupillary reaction and optic nerves. He had two previous episodes of POC in the left eye when he was 6 and 8 months old which was treated in the ophthalmology department. The two previous episodes improved with intravenous antibiotics within 48 h. In the second admission, he also underwent successful probing for the left nasolacrimal duct obstruction [Figure 1a and b]. Written informed consent has been obtained from parents of the patient for publishing the photographs.

His medical history showed that he was born at 35 weeks of gestation, and he had normal growth and met all developmental milestones. His immunization was up-to-date, and the family history was unremarkable.

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Figure 1: Clinical photography of the patient: (a) First episode, presenting as left preseptal cellulitis in the medial canthal area at 6 months of age. (b) Second episode, presenting as a severe extensive left preseptal cellulitis at 8 months of age which improved after antibiotic therapy and nasolacrimal duct probing. (c) Third episode, presenting as right periorbital cellulitis at 12 months of age

We performed computed tomography scan of the paranasal sinuses that revealed sinusitis in both ethmoid and maxillary sinuses and evidence of inflammation in the right preseptal region [Figure 2].

Diagnostic nasal endoscopy revealed pus in the right middle meatus, with no significant anatomical abnormality or polyps. The nasal discharge was cultured. On the 1st day of admission, complete blood count (CBC) demonstrated a total white blood cell count of 12,100/ml with a 4% neutrophil count. In subsequent CBC recheck, episodic neutropenia was confirmed. Other hematologic indices showed monocytosis, and no morphologic abnormalities of the neutrophils were detected in the blood smear.

Immunologic assessment including immunoglobulins level, antineutrophil antibodies, nitroblue tetrazolium blood test, and other evaluations for systemic disorders showed no abnormal findings. Cystic fibrosis was ruled out by sweat chloride test, and investigations for HIV antibody were negative.

The patient received intravenous antibiotics consisting of ceftriaxone (Jaber Ebne Hayyan Pharmaceutical, Tehran, Iran) and clindamycin (Alborz Darou, Qazvin, Iran) upon admission. The POC improved significantly after 48 h of intravenous administration of antibiotics, and he was discharged subsequently with oral antibiotics. After 6 days, the neutrophil count returned to normal range. We reviewed the medical chart of his admissions in the ophthalmology ward for the prior episodes and noticed similar neutropenia in those attacks, which was overlooked because of prompt response to treatment and early discharge.

The diagnosis of cyclic neutropenia was reconfirmed by serial CBC review (3 times/week for 8 weeks) and excluded acquired causes of neutropenia and immune deficiencies. In the neutropenic phase, neutrophil count dropped $<200/\text{mm}^3$. He was referred to hematologic service and received granulocyte colony-stimulating factor and prophylactic antibiotics. In a 3-year follow-up, he remained asymptomatic, with no recurrence of cellulitis episodes.

DISCUSSION

POC is a relatively common disorder in the pediatric population, and a multidisciplinary approach including an otolaryngologist, ophthalmologist, and pediatrician is necessary for the best management outcome. Although rhinosinusitis is the most prevalent cause of POC, it has rarely been presented as the

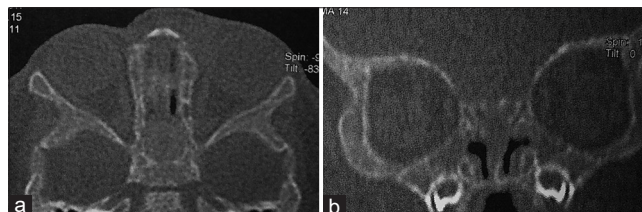


Figure 2: Computed tomography scan of the orbit and paranasal sinus showing sinusitis in underdeveloped sinuses: (a) Axial image. (b) Coronal image

source of recurrent POC. Therefore, other underlying etiologies should also be considered and investigated in these patients.¹

Various etiologies including the anatomical abnormalities have been described in patients with recurrent POC in the literature [Table 1]. Jatana *et al.* reported an abnormal uncinat process in continuity with the ethmoid bulla in a 28-month-old boy with seven episodes of recurrent unilateral POC due to recurrent rhinosinusitis.³ Infections with unusual microorganisms have also been reported as a cause of recurrent POC. Mauriello and Atypical Mycobacterial Study Group presented atypical mycobacterial infections of the periorbital region after periorbital and facial surgery.⁵

Although recurrent POC often present in the same orbit, bilateral POC has rarely been reported.² In our patient, there was intermittent bilateral involvement, which means that the etiology of recurrent POC was not limited to one eye.

Neutropenia can present in several clinical settings. The most common etiologies of neutropenia are acquired, including immune neutropenia, neutropenia associated with sequestration, viral infections, and various medications. Inherited etiologies of neutropenia are less common and are often more profound. Cyclic neutropenia is a rare hereditary disorder with variable presentations. This disorder is characterized by periodic neutropenia lasting for 4–6 days with a 21-day interval. According to the absolute neutrophil count (ANC), neutropenia has been classified as mild (ANC: $1000\text{--}1500/\text{mm}^3$), moderate (ANC: $500\text{--}1000/\text{mm}^3$), and severe (ANC: $<500/\text{mm}^3$).¹⁵ In this patient, clinical characteristics and laboratory investigations revealed the severe form of cyclic neutropenia.

Opportunistic infections can occur during the neutropenic period. The most common sites of infection are skin, oral mucosa, and lungs.¹⁵ Of note, our patient never presented recurrent infections in sites other than orbit and paranasal

Table 1: Etiologies that caused or mimicked recurrent periorbital cellulitis

Studies	Underlying abnormality or disorder
Caulley <i>et al.</i> ²	Bilateral bony dehiscence of the lamina papyracea
Jatana <i>et al.</i> ³	An abnormal uncinat process
Karkos <i>et al.</i> ⁴	A long abnormal uncinat process and an isolated infected cell in anterior ethmoidal air cells
Mauriello and Atypical Mycobacterial Study Group ⁵	Atypical mycobacterial infections of the periocular region
Hoare ⁶	HIV infection
Galindo-Ferreiro <i>et al.</i> ⁷	Orbitopalpebral venous lymphatic malformation
Akçay <i>et al.</i> ⁸	Congenital lacrimal sac diverticulum
Sorin <i>et al.</i> ⁹	Atopic reaction to cosmetics, herpetic infection, environmental allergies and malingering
Rappaport <i>et al.</i> ¹⁰	Plasmacytoma
El-Hakim <i>et al.</i> ¹¹	Non-Hodgkin's lymphoma
Hassan <i>et al.</i> ¹²	Teratoma
Kim and An ¹³	Extranodal natural killer/T-cell lymphoma
Younes <i>et al.</i> ¹⁴	Shwachman-Diamond syndrome

sinuses. In fact, it seems that the neutropenic episodes caused sinusitis and dacryocystitis as the underlying condition for the recurrent POC. In addition, he had no positive family history of the recurrent or unusual infections. In each episode, POC resolved within a short period following administration of intravenous antibiotics, and in the periods with the normal neutrophil count, there were no infectious signs.

In conclusion, we presented a child with recurrent POC secondary to cyclic neutropenia. This report signifies the importance of considering unusual etiologies including the immune system deficiencies when patients with recurrent POC are encountered.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient's parent has given his consent for images and other clinical information to be reported in the journal. The patient's parent understands that his names and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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