A 13-year-old boy presented with lesions around his eyes since two months. He had been having repeated episodes of similar lesions around eyes, mouth, and perianal region since eight months of age for which he was taking treatment at nearby hospitals. He had recurrent respiratory tract infections. He was born out of second-degree consanguineous marriage. There was no similar history in the family. A detailed physical examination revealed yellow granulomatous nodules and furuncles around the eyes, nose, and oral cavity [Figure 1]. There was growth retardation, but mental development was normal for his age. Acanthosis nigricans was present over both axillae. Postinflammatory hyperpigmentation was present throughout the body. Systemic examination did not reveal hepatosplenomegaly. Laboratory investigations such as complete hemogram and HIV status were within normal limits. Ophthalmic examination revealed 2 × 2 cm furuncle over the lateral canthi with purulent discharge and healed corneal ulcer with leukomatous corneal opacity. Left eye showed ankyloblepharon with lid edema [Figure 2]. Fundus examination was within normal limits. The features were suggestive of chronic granulomatous disease. Nitroblue tetrazolium test was not performed as it was unavailable. The patient was started on oral cotrimoxazole and itraconazole and is presently under follow up.

Chronic granulomatous disease (CGD) is a congenital immunodeficiency disorder of phagocyte bactericidal function with X-linked recessive inheritance, characterized clinically by granulomatous lesions in the skin, lymph nodes, lung, liver, gastrointestinal tract, and bones. The main pathology in this disease is a defect in the subunits of nicotinamide adenine dinucleotide phosphate oxidase, whose normal functioning is essential for killing phagocytosed bacteria by neutrophils using the myeloperoxide-halide system. They develop recurrent bacterial infections by Staphylococcus aureus, Chromobacterium violaceum, Nocardia species, Serratia marcescens, legionella, and atypical mycobacteria. Fungal infection by aspergillus is also common. The nitroblue tetrazolium test is used to confirm CGD.[1]

The disease may become apparent in the first two years of life or may be delayed into the second decade of life. Neonatal pustulosis may be the initial manifestation. Impetiginized periorificial rash is highly characteristic and most commonly seen around the nostrils, ears, mouth, and eyes. Firm, translucent, papular lesions may be found around the eyes, nose, lips, and on the cheeks. Other

Figure 1: Granulomatous nodules around eyes, nose, and mouth

Figure 2: Left eye with lid edema, ankyloblepharon, and granulomatous nodule on the lateral canthus
frequent pyogenic infections include liver abscesses, osteomyelitis, arthritis, pneumonia, skin sepsis, and perianal abscesses.\(^{[2]}\)

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